NEVUS OF OTA: PRESENTATION OF A CASE WITH ORAL PIGMENTATION AND CODICIL TO TANINO’S CLASSIFICATION

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Keywords: Bluish hyperpigmentation, Palatal pigmentation, trigeminal nerve, congenital melanocytosis, melanocytic hamartoma

Abstract
Nevus of Ota is a rare pigmented disorder affecting the skin along the distribution of trigeminal nerve. Oral involvement of this pigmented lesion is very rare. Classification of Nevus of Ota given by Tanino represents the skin manifestations well but does not include the extra cutaneous manifestations. Hence an addition to this classification is being proposed through this case report which describes the 17th case of palatal involvement in a male patient in a female dominant Nevus of Ota.

Introduction
Congenital or acquired melanocytoses are benign pigmented lesions of the skin or mucosa1. Nevus of Ota also known as nevus fuscocaeruleus ophthalmomaxillaris is a congenital dermal melanocytosis first described by Ota in 1939 as a melanocytic hamartoma. It clinically presents as bluish hyperpigmentation along the branches of the trigeminal nerve2.

Case Report
A 22-year old male patient visited for a routine dental check up. His medical, surgical, drug and personal histories were non contributory. Examination of the facial skin revealed a brownish grey pigmentation of the left eyebrow region of 1-1.2 cm size and associated slate-grey pigmentation of the left conjunctiva (Figure 1). Examination of the ipsilateral and contralateral side of face revealed no additional pigmentation. On oral examination, the hard palate on the left side showed irregular greyish macule on the left side with a few surrounding scattered similar coloured macules (Figure 2). The rest of the oral mucosa appeared normal. Upon eliciting the history of the pigmentation, patient was unaware of the oral lesions but the eye pigmentation was present since birth. Evaluation by an ophthalmologist and otolaryngologist did not reveal any pigmentation in the nasal or aural mucosa and no eye complications were present. Based on the history and clinical appearance a diagnosis of Nevus of Ota was arrived at. An attempt to classify was futile as an effective classification is unavailable and hence a codicil is being proposed to the existing Tanino’s classification.

Discussion
Although Hulke in 1861 first described oculodermal melanosis, in 1939 Ota M coined the explanatory name for this condition as nevus fuscocaeruleus ophthalmomaxillaris. Subsequently the condition is known as nevus of Ota3.

The exact etiology of this disorder is yet to be determined. But it is suggested that amelocytic nevoid cells present at birth become pigmented in the teenage life or later due to many triggering factors such as ultraviolet light exposure, female hormones, infection or trauma4,5. Another suggestion is that it may be due to non migration of melanocytes from the neural crest to the epidermis during the embryonic stage. Other theories postulated is an active production by intradermal melanocytes6,7. Although most patients have a negative family history, rarely familial cases have been described, representing heredity as a possible aetiology8.

The original classification by Tanino in 1939 has remained the most useful clinical classification, although mucosal pigmentation or extra cutaneous manifestations are not included. Tanino’s classification is summarized in table 13.
**Table 1**

<table>
<thead>
<tr>
<th>Type</th>
<th>Subtypes</th>
<th>Areas involved</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type-I</td>
<td>IA</td>
<td>Mild orbital type - distribution over the upper and lower eyelids, periocular and temple region.</td>
</tr>
<tr>
<td></td>
<td>IB</td>
<td>Mild zygomatic type - infrapalpebral fold, nasolabial fold and zygomatic regions are affected</td>
</tr>
<tr>
<td></td>
<td>IC</td>
<td>Mild forehead type - only forehead is affected.</td>
</tr>
<tr>
<td></td>
<td>ID</td>
<td>Ala nasi alone is affected.</td>
</tr>
<tr>
<td>Type-II</td>
<td></td>
<td>Moderate type-The lesions affect upper and lower eyelids, periocular, zygomatic, cheek and temple regions</td>
</tr>
<tr>
<td>Type-III</td>
<td></td>
<td>The condition is distributed over the scalp, forehead, eyebrows and nose.</td>
</tr>
<tr>
<td>Type IV</td>
<td></td>
<td>Bilateral type</td>
</tr>
</tbody>
</table>

**Figure 1** Pigmentation on the left conjunctiva and left eye brow region.

**Figure 2** Palatal pigmentation.
A modification has been suggested to include the presence of Nevus of Ito in association with Ota as Type V and Type VI\(^3\). It is suggested that the letter “E” may be suffixed when nevus is associated with extra cutaneous manifestations\(^3\). However, another modification has been suggested for oral lesions as IE\(^5\). As the former uses letter E to represent extra cutaneous manifestations the latter uses the letter E to represent oral lesions resulting in much confusion and ineffective representation of mucosal pigmentation. Therefore a comprehensive classification is proposed, which is as below:

- **Type I to IV:** As suggested by Tanino
- **Type V:** Nevus of Ota with Nevus of Ito
- **Type VI:** Nevus of Ota with extra-cutaneous manifestations
  - VI A: Ocular
  - VI B: Oral - Palate, Buccal mucosa or other areas
  - VI C: Nasal
  - VI D: Tympanic
  - VI E: Leptomeninges

The reported incidence is about 0.2% to 1% in the Japanese population\(^9\). The exact prevalence in Indians is not known but the male to female sex ratio is 1:4.8\(^2,10\). Nevus of Ota is most common in Asians and blacks and rare cases affecting whites have also been reported. Onset is usually at birth but may appear at adolescence in some and during pregnancy in a few women\(^3,7\).

Clinically, it appears as blue-grey macular pigmentation on the skin with the borders being irregular. Classically, it is noted on the skin supplied by the branches of the trigeminal nerve. In 95% of cases, it is unilateral and bilateral in the rest\(^11\). A similar pigmentation may be found on the tympanic membrane, oral cavity, eye and leptomeninges\(^5\). Oral lesions appear as blue-grey macules or plaques with irregular borders\(^5\). Associated abnormalities include congenital glaucoma, Duane’s syndrome and melanoma\(^5,7\). Only 16 cases with palatal pigmentation has been reported, and this is the 17th.

It is important to differentiate N. of Ota from other conditions. Acquired, bilateral nevus of Ota-like macules (ABNOM) or Hori’s nevus is located bilaterally on the face, appears later in life, is blue-brown or slate-grey in color and commonly seen in middle-aged women of Asian descent. It is not accompanied by macules on the ocular and mucosal membranes\(^12,13\).

Mongolian spots (MS) are birthmarks that are present at birth and their most common location is sacrococcygeal or lumbar area. Aberrant MS over occiput, temple, mandibular area, shoulders and limbs have been reported\(^14\). Nevus of Ito occurs in the territory supplied by the acromioclavicular nerve\(^1\), and hence found on the shoulder, neck, upper arm and supra clavicular area\(^2\).

Histopathology of the affected skin shows the presence of dendritic cells containing melanin in the dermis\(^11\).

Different modalities have been proposed for the management of Nevus of Ota. This includes selective photothermolysis with Q switched ruby laser or Nd:YAG laser which is considered to be a safe and effective treatment\(^4,15\), dermabrasion, cryotherapy and historically surgical excision\(^2\).

Malignant transformation of the melanocytoses, though very rare, has been reported including nevus of Ota\(^1\).

In conclusion, Nevus of Ota is a hamartomatous proliferation of melanocytes. This entity is still a rarity in India especially the extra-cutaneous, oral mucosal manifestation. The importance of recognizing this entity needs to be emphasized, as a magnitude of pigmented lesions is noted on the oral mucosa. Also, long term follow-up is essential as very rarely dermal melanocytosis can undergo malignant transformation.
References


GENERAL DERMATOLOGY - Short Case

CLASSIC DERMATOMYOSITIS FOLLOWING BREAST CANCER: A CASE REPORT

Chee Yong Chuang¹, Choon Siew Eng, FRCP²

Keywords: paraneoplastic sign, ductal carcinoma, proximal myopathy

Introduction
Dermatomyositis and polymyositis are idiopathic inflammatory myopathies characterized by symmetrical proximal muscle weakness and distinctive skin manifestations¹-⁴. It is a rare disease with an estimated combined annual incidence of 2 per 100,000 population⁵. Population-based studies from Sweden, Australia, and Scotland showed that dermatomyositis, and to a lesser extent polymyositis, carried an increased risk of malignancies⁶-⁸. Cancers such as adenocarcinomas of the lung, pancreas, cervix and stomach as well as haematological cancers including Hodgkin's and non-Hodgkin's lymphoma accounted for approximately 70% of the cancers associated with inflammatory myopathies⁹. Types of malignancy associated with inflammatory myopathies may depict local prevalence of certain forms of malignancy. For instance, nasopharyngeal carcinoma is commonly reported in Southeast Asian patients with dermatomyositis¹⁰. Breast cancer is the most common accompanying malignancy among women with dermatomyositis/polymyositis in Korea and Taiwan. The onset of dermatomyositis/polymyositis may precede, coincide with or follow the diagnosis of breast cancer¹¹,¹². We report a case of classic dermatomyositis that developed 3 months after the discovery of breast cancer.

Case report
A 25 year old nulliparous Vietnamese lady, married to a Malaysian, was referred to us for the evaluation of a rash that developed 3 months after the discovery of a breast lump by self examination. She had no family history of malignancy and was otherwise well before the breast lump was removed by a wide local excision in a private hospital. A week post-procedure, she noticed asymptomatic rashes that started over the forehead and gradually spreading in a descending fashion to involve the eyes, face, chest, neck, extensors of the upper limb, dorsum of the hands and the inner thighs.

It was associated with symmetrical muscle weakness affecting the proximal muscles of both upper and lower limbs. The weakness was profound as she found herself unable to carry out activities of daily living. She had difficulty sitting up from a lying position, climbing stairs or rising from a seated or squatting position. She also found it difficult to perform activities that require holding the arms up like washing her hair or reaching into overhead cupboards. She also experienced neck weakness, where she had difficulty raising her head from a pillow or holding it up while standing. She also complained of difficulty swallowing, nasal regurgitation and hoarseness of voice. Her weakness deteriorated rapidly till she was bed bound.

Physical examination revealed profound proximal muscle weakness and classic skin lesions. She had bilateral violaceous erythematous rash with periorbital and eyelid edema consistent with heliotrope rash (Figure 1). Photo-distributed erythema with V-neck sign and shawl sign were noted over the anterior chest and upper back respectively. Gottron's sign characterized by elevated violaceous papules and plaques over bony prominences were also present in addition to nail fold changes namely periungual erythema, telangiectasia and ragged cuticles (Figure 2).
Figure 1  Violaceous erythematous macules and patches in sun-exposed areas exhibiting classic Heliotrope and V-neck signs.

Figure 2  Gottron’s papules, periungual erythema and ragged cuticles.
Myositis was confirmed by elevated muscle enzymes (creatine kinase: 10720 U/L, lactate dehydrogenase: 840 U/L, alanine aminotransferase: 149 U/L, aspartate aminotransferase: 491 U/L), electromyography and muscle biopsy. Both erythrocyte sedimentation rate (42 mm/hour) and C-reactive protein (12.1 mg/L) were elevated. Anti-nuclear antibody was positive with a titre of 1280 demonstrating a speckled pattern. Anti-double-stranded DNA, anti-Jo1 and anti-topoisomerase antibodies were all negative. Histology of her breast lump revealed poorly differentiated infiltrating ductal carcinoma ([Bloom and Richardson's Grade III, Clinical Staging: T2N1M0, Stage II A].

In spite of the institution of high dose prednisolone, she developed increasing dyspnoea with acute hypoxemic respiratory failure complicated by nosocomial pneumonia. She was ventilated and treated with multiple intravenous antibiotics, intravenous methyprednisolone and subsequently intravenous immunoglobulin. Aggressive care allowed weaning her off the ventilator support but patient declined an urgent mastectomy and axillary clearance which was scheduled with the hope of alleviating her myositis. She opted for treatment back in Vietnam and requested to be discharged from hospital while still on prednisolone 40 mg daily. She succumbed to the disease in Vietnam one month after discharge before any surgical intervention.

Discussion

Idiopathic inflammatory myopathies are a mixed group of disorders characterized by symmetrical proximal muscle weakness and evidence of inflammation involving striated skeletal muscles. Patients with dermatomyositis fulfil criteria for polymyositis in addition to having characteristic cutaneous manifestations.

Dermatomyositis is usually an idiopathic autoimmune connective tissue disease but about 15-30% of adult-onset dermatomyositis have underlying malignancies. In our patient, dermatomyositis was diagnosed based on the “revised criteria for the diagnosis of inflammatory myositis” proposed by Bohan and Peters. She had paraneoplastic dermatomyositis following breast cancer that had metastasized to axillary nodes. The relationship between dermatomyositis and malignancy has been established in various studies. Malignancy may be diagnosed before, simultaneously or following the development of dermatomyositis. The majority of the diagnosis of cancer was made within two years pre- or post-diagnosis of the inflammatory myopathy.

In Finland, Denmark and Sweden, the common cancer associated with dermatomyositis are ovarian, breast, lung, pancreatic, stomach and colorectal cancer. Whereas in Southeast Asian countries like Singapore, Korea and Taiwan, breast, stomach and nasopharyngeal cancer (NPC) predominated. Breast cancer is the top cancer in women both in the developed and the developing world, comprising 16% of all female cancers. Thus, it is not surprising, that it is also a common cancer seen in patients with dermatomyositis. Breast cancer accounted for 21.3% of malignancies in 89 Taiwanese women with paraneoplastic dermatomyositis / polymyositis. The mean age of patients with either adult onset dermatomyositis or breast cancer is usually more than 50 years old. Here, we report a rare case of a young patient with rapidly progressive and fatal dermatomyositis following a newly diagnosed breast cancer.

References


CLINICAL CHARACTERISTICS OF PATIENTS WITH LEPROSY IN HOSPITAL KUALA LUMPUR

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Abstract

Background: Nearly half of the new leprosy cases reported in Malaysia were foreigners from neighbouring Asian countries.

Objectives: To determine the clinical characteristics of leprosy and its difference, among the Malaysian and the foreign patients

Methods: This is a 4-year retrospective analysis of 75 leprosy patients who attended Hospital Kuala Lumpur Hansen’s Clinic. Variables included the disease clinical characteristics, clinical severity and the complications.

Results: Foreigners accounted for 51% of total patients with mean age of 35.8 years. Malaysians presented at mean age of 40.8 years. The gender ratio (male: female) was 2.7:1 in the former and 2.3:1 in the latter. The Malaysians tend to present later (average after three years) to the clinic. The clinical presentations in both groups of patients didn’t significantly differ.

Conclusion: In Malaysia. Leprosy shouldn’t be labelled as the disease of the immigrants. Social awareness on the disease should be equally highlighted to both locals and foreigners, especially among females.

Keywords: granulomatous disease, Hansen’s disease, Malaysia

Introduction

Leprosy is a chronic granulomatous infection caused by Mycobacterium leprae (M leprae), principally affecting the skin and peripheral nervous system. M leprae is an obligate acid-fast bacillus, and humans are its principal reservoir.

In Malaysia, leprosy remains to be a disease burden despite a tremendous reduction in its prevalence for the past 20 years. Leprosy prevalence rate in Malaysia from year 2005 to 2007 was 0.3 per 10 000 populations in three consecutive years. The incident rate in 2007 was 0.7 per 100 000, a decline from 1.1 per 100,000 in 2005.

The proportion of foreigners diagnosed with leprosy from the total new cases in Malaysia increased from 24.5% in year 2000 to 44.0% in 2007. In year 2007, foreigners’ contribution to all new leprosy cases in Malaysia had showed an increasing trend of 13% from the previous year.

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In developed country like the United States, 200-300 cases are reported each year, mainly from the states with large immigrant populations (eg, California, New York, Florida)\(^2,3\). The disease spreads by aerosolized droplets from lepromatous patients and, less commonly, through direct skin contact.

Leprosy can have a protracted and insidious onset. After exposure, person who is susceptible to leprosy may develop a single skin lesion after an incubation period averaging 5 to 7 years (range from 3 months to 40 years). Therefore clinical signs may not manifest until after the immigration process is complete. Given that the majority of new migrants with leprosy manifested the disease within 1 year of immigration, it is postulated that stress or socio-economic factors associated with migration may have pushed the disease from being quiescent to symptomatic. Host immunity, particularly the cell-mediated immune response, plays an important role in its tissue damage. The severity of deformities caused by nerve tissue damage was influenced by the type and extent of bacillary spread and multiplication, also the occurrence of immunologic complications i.e. lepra reactions.

Malaysia as one of the fastest growing developing countries in Asia is facing the same issue as in developed country, with the high influx of foreign workers attributing to the higher number of medical cases. Given that leprosy is still prevalent in certain countries in Asia like Cambodia and Nepal\(^4\), where it is not easily treated due to poor socio-economic conditions, the unprecedented mobility of these foreigners crossing the Malaysian borders has increased the number of leprosy patients. Thus, it is important for Malaysian health care providers to be familiar with the clinical characteristics of leprosy locally.

Objective
We sought to determine the clinical characteristics of patients with leprosy, and the differences in clinical characteristics between local Malaysian patients and foreign patients.

Methods

Study Design and patients
A retrospective analysis was performed. Study population included patients diagnosed with leprosy in Hospital Kuala Lumpur, a tertiary referral hospital, between the year 2006 and 2009. This review focused on demography, classification, deformities, lepra reactions and drug resistance.

A total of 75 patients (52 male and 23 female), aged between 10 and 75 years, were analyzed according to group; Malaysians and foreigner. Foreign patients were further subdivided into permanent resident or not. Patients who had incomplete medical record and who defaulted follow-up were excluded from the study.

All patients diagnosed with leprosy had slit skin smears taken from the suspected sites of skin lesions and stained with Fite method. Skin biopsy was performed to those who had negative finding from slit skin smears, those who had bacillary Index of more than 3, and to those who presented only with nerve lesion.

Data Collection
All data were derived retrospectively from the medical records and were recorded in data sheets in the computer. The variables used for comparison in clinical presentations included the type or severity of leprosy in accordance to Ridley-Jopling Classification, drug resistance to one or more drugs using mouse foot pad culture and disease complications including lepra reactions and neurological complications.

Statistical analysis
Statistical analysis was performed using Fisher’s exact test for the comparison of proportions. A value of P less than 0.05 was considered statistically significant. All analyses were performed using SPSS (Statistical Package for Social Science) programme version 14. Comparisons of frequencies between local and foreign patients were conducted with independent sample T test or Mann U Whitney Test.

Results
A total of 75 patients (aged from 10 to 75 years) were reviewed. Thirty eight of the 75 patients (50.7%) were foreigners. The average age in both groups fell on the second bimodal age distribution, with peaks at the age of 35-44 years. The nationalities of foreign patients were shown in Figure 1.

The ethnic distribution of local patients was demonstrated in Figure 2. Malay and Chinese ethnic accounted for 38% each of all Malaysian patients foreigners, mostly Indonesians, accounted for 51% of total patients. Twelve of them, all Indonesians, were permanent residents.
Multibacillary leprosy predominated in each group, 86% among Malaysians and 95% in foreigners, as shown in Table 1. The Malaysian patients had higher prevalence (35.1%) of lepromatous leprosy (LL) whereas foreign patients presented with more cases (34.2%) of borderline lepromatous (BL) as shown in Table 2.

The overall mean duration of symptoms before diagnosis was 2 years (8 months-5 years). Malaysians tend to present later, with a mean of 3 years after onset of symptoms, as compared to the foreigners (Figure 3).

![Figure 1](distribution_of_foreign_patients_by_nationality.png)  
**Figure 1** Distribution of foreign patients by nationality

![Figure 2](distribution_of_malaysian_patients_by_race.png)  
**Figure 2** Distribution of Malaysian patients by race

<table>
<thead>
<tr>
<th>Patient's groups</th>
<th>Paucibacillary</th>
<th>Multibacillary</th>
<th>Neural</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malaysian</td>
<td>4 (11.0%)</td>
<td>32 (86.0%)</td>
<td>1 (3.0%)</td>
<td>37</td>
</tr>
<tr>
<td>Foreigner</td>
<td>1 (2.5%)</td>
<td>36 (95.0%)</td>
<td>1 (2.5%)</td>
<td>38</td>
</tr>
<tr>
<td>Total /No (%)</td>
<td>5 (7.0%)</td>
<td>68 (91.0%)</td>
<td>2 (2.0%)</td>
<td>75</td>
</tr>
</tbody>
</table>

**Table 1** Patients distributed into WHO leprosy classification.

<table>
<thead>
<tr>
<th></th>
<th>Malaysian</th>
<th>Foreigner</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neural</td>
<td>2.7%</td>
<td>2.6%</td>
</tr>
<tr>
<td>Indeterminate</td>
<td>5.4%</td>
<td>0.0%</td>
</tr>
<tr>
<td>Tuberculoid</td>
<td>8.1%</td>
<td>2.6%</td>
</tr>
<tr>
<td>Borderline Tuberculoid</td>
<td>24.3%</td>
<td>21.1%</td>
</tr>
<tr>
<td>Borderline</td>
<td>2.7%</td>
<td>10.5%</td>
</tr>
<tr>
<td>Borderline Lepromatous</td>
<td>21.6%</td>
<td>34.2%</td>
</tr>
<tr>
<td>Lepromatous</td>
<td>35.1%</td>
<td>28.9%</td>
</tr>
</tbody>
</table>

**Table 2** Patients distributed into Ridley-Jopling classification of leprosy.
Fifty percent of Malaysian patients were diagnosed with grade 1 deformity and 8% were diagnosed with grade 3 deformity (Figure 4). Among the foreigners, twenty-one percent presented with grade 1 deformity and 8% with grade 3 deformity. Half of the foreign patients (50%) had already had grade 1 deformity at presentation (Figure 4).

Among all patients who developed lepra reactions, 22 patients were diagnosed with type 1 (reversal) reaction, 27 patients with type 2 reaction (erythema nodosum leprosum) and 2 patients with Lucio’s reaction. Full dapsone resistance was confirmed by mouth foot-pad culture in one Malaysian patient.

Type 2 lepra reaction (ENL) occurred in 39.5% of Malaysians and in 32.4% of foreign patients. The occurrence of lepra reactions, deformities and drug resistance did not differ significantly between the two groups of patients. (Table 3)

Figure 3 Duration (year) from onset of symptom to time of diagnosis, of study patients.
Table 3 Clinical presentations in both groups of patient.

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Frequency (%)</th>
<th>Malaysians N = 37</th>
<th>Foreigners N = 38</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoaesthetic/anaesthetic skin</td>
<td>80</td>
<td>45</td>
<td>35</td>
</tr>
<tr>
<td>Nerve thickening</td>
<td>79</td>
<td>36</td>
<td>43</td>
</tr>
<tr>
<td>Peripheral Neuropathy</td>
<td>73</td>
<td>33</td>
<td>40</td>
</tr>
<tr>
<td>Positive slit-skin smears</td>
<td>87</td>
<td>40</td>
<td>47</td>
</tr>
<tr>
<td><strong>Mean + SD</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bacillary index (BI)</td>
<td>3.5 ± 1.7</td>
<td>3.4 ± 1.7</td>
<td>3.6 ± 1.7</td>
</tr>
<tr>
<td>Morphology index (MI)</td>
<td>3.1 ± 2.2</td>
<td>3.7 ± 2.7</td>
<td>2.5 ± 1.5</td>
</tr>
<tr>
<td>Partial Dapsone Resistance</td>
<td>2 (5.4%)</td>
<td>8 (10.7%)</td>
<td></td>
</tr>
<tr>
<td>Full Dapsone Resistance</td>
<td>1 (2.7%)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td><strong>Frequency</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sensitive to all MDT</td>
<td>4</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Drug sensitivity not done</td>
<td>11</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Mouse foot pad culture result not</td>
<td>19</td>
<td>19</td>
<td></td>
</tr>
<tr>
<td>available</td>
<td></td>
<td></td>
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</table>

*MDT: Multidrug Therapy*

Figure 4 Frequency of neurological deformity in both groups of patients.
Discussion
In this case series, there was a male predominance with a male-to-female ratio of 2.3 to 1. This is consistent with other studies done in other states of Malaysia and also most part of the world excluding Africa, Thailand and Japan. In the past decade, Malaysian women received equal education as the man but most Asian women had higher social embarrassment that leads to higher social stigma on leprosy. The possibility of the disease being under diagnosed in females of this country has never been investigated although; it is believed that the difference between the genders in post pubertal age is real. However, the male: female ratio in this review was accentuated (baseline: a male to female ratio of 1.5-2.0 to 1), probably due to delayed presentation by the female patients. There was no evidence of racial predilection in this disease.

The main complication of leprosy is disability caused by nerve damage, a consequence from Schwann cell invasion by Mycobacterium leprae and the patient’s immune response. Previous studies showed that prevalence of neurological impairment increases with late presentation. Other risk factors for chronic or recurrent neuropathy include leprosy classification (both WHO and Ridley-Jopling) and Lepra reactions. The disease severity, and occurrence of deformity, reaction and drug resistance among the Malaysian and foreigners was comparable. Surprisingly most Malaysian presented later than 1 year from onset of symptom, hence may contribute to higher percentage of Malaysian patient found to have grade 3 deformities at presentation.

A study done in Malaysia between 1985-1990 showed a high percentage of dapsone resistance (22.4%). This study reported 10 partial and 1 full dapsone resistance out of 21 patients with MFP culture results (Table 3), hence no conclusive statistically significant data on dapsone drug resistance can be made on this study population.

Within a short period of less than 4 years (Jan 2006 until October 2010), our study cohort of leprosy diagnosed in Hospital Kuala Lumpur (HKL) consists of more (50.7%) foreigners than that was seen in studies conducted in a longer study period (10 to 14 years) in Penang (22.2%) and Johor Bahru (33%). Kuala Lumpur being the centre of Malaysian economic activities has more foreign workers, hence more imported cases being diagnosed. Most of the foreigners were from the neighbouring Asian countries. This is not surprising as the World Health Organization in 2001 reported that Indonesian and Myanmar were still leprosy endemic countries. All foreign patients presented within 4 years after the onset of symptoms. As most of them had passed the routine medical screening before working in Malaysia, more study should be done to obtain information on triggering factors of symptomatic disease onset. Leprosy transmission may have occurred in their country of origin as Mycobacterium leprae has a long incubation period. It is also alarming to know that subclinical infection was found in up to 5% of healthy Indonesian in their country.

It is also important to highlight whether leprosy cases among the locals in Kuala Lumpur had remained under diagnosed. Most Malaysian patients presented within 3 years of symptom onset. However, 13% of them presented later than 4 years, attributing to significant morbidity at the time of diagnosis. There are many reasons of the delayed diagnosis which includes, patients soughing healthcare advice only after symptoms caused inconvenience, patients preferred traditional treatment at early stage of disease, early disease undiagnosed by primary healthcare practitioners and lastly, patients delayed treatment due to social, geographical or financial reasons. There is an urgent need for Malaysians to be continuously educated about leprosy, as early treatment would significantly improve morbidity and reduced the country economic burden.

Conclusion
Clinical characteristics of leprosy in terms of disease severity, prevalence of lepra reactions, neurological deformity and frequency of drug resistance were comparable between local and foreign patients. Hence in Malaysia, leprosy should no longer be labelled as a disease of the immigrant or imported disease. Public education on leprosy is very important, particularly among the orang asli, bumiputera Sabah and Sarawak.

Compared to foreign patients, local patients tend to present later after the onset of symptoms, attributing to higher non-reversible neurological disability or deformity. ‘Social stigma’ on leprosy among Malaysian population should be replaced by awareness that the disease is curable and that the resultant neuropathy is treatable if treated early.

Acknowledgement
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DERMATOLOGY INFECTION - Short Case

PERIANAL TUBERCULOSIS: A CASE REPORT AND LITERATURE REVIEW

Selvarani S1, Tang JJ1, Norain K2

Keywords: anal tuberculosis, periorificial tuberculosis, ano-perianal tuberculosis, extrapulmonary tuberculosis

Introduction
Periorificial tuberculosis is a rare manifestation of cutaneous tuberculosis which is characterized by painful ulcerative lesions affecting oral, genital, anal mucosa and adjacent skin of the orifices. It occurs following auto-inoculation of mycobacteria from pulmonary, gastrointestinal or genitourinary foci that affect traumatized mucocutaneous areas around the orifices1. Rarely, haematogenous, lymphatic or direct spread from neighbouring organs can lead to this condition. The tongue is the most frequently affected site, but the perianal area can also be affected2. Herein, we report a rare case of perianal tuberculosis presented with non-healing perianal ulcer for 2 years and to emphasize the importance of considering tubercular etiology in the work up of persistent perianal ulcer.

Case report
A 54 year-old Chinese man presented with persistent perianal ulcer of 2 years. He was seen by many doctors and was treated with multiple topical and oral antibiotics without much improvement. The perianal ulcer was getting bigger and associated with pain and serous discharge. He had persistent low grade fever, night sweat with significant loss of appetite and loss of weight. There was also history of intermittent constipation for 2 years which was associated with few episodes of per rectum bleeding. He underwent colonoscopy in 2010 but was essentially normal. There was no history of chronic cough or haemoptysis.

He has no family history of tuberculosis and he denied contact with any patients with tuberculosis. He was married with 3 children and worked as a lorry driver. He denied history of anal intercourse, sexual promiscuity or homosexuality. He was an ex smoker and did not consume alcohol. On examination, he was afebrile and pink. Perianal examination revealed an ulcer extending from base of scrotum to an area of 3 cm around the perianal region (Figure 1). The ulcer was tender with hyperpigmented margin and covered with mucopurulent discharge. There was no inguinal lymphadenopathy. Examination of other systems including respiratory system was essentially normal.

Our initial differential diagnosis included perianal Crohn’s disease, perianal Tuberculosis, Chancroid, Hailey-Hailey disease, Langerhans cell histiocytosis and neoplastic conditions such as extramammary Paget’s disease and Marjolin ulcer. Laboratory findings of complete blood count, renal profile, liver function tests and urinalysis were normal, except for ESR which was raised at 52mm/hr. The VDRL, TPHA, Hepatitis B, C and HIV serology were negative. Chest X-ray revealed scattered microgranular opacity in both upper zone of lung. Sputum for Acid Fast Bacilli was however negative. A skin biopsy taken from the margin of an ulcer showed granulomatous lesion with presence of Langerhans-type multinucleated giant cells and Ziehl Neelson stain showed abundant acid fast bacilli within the tissue (Figure 3 and 4). Skin biopsy was positive for Mycobacterium tuberculosis culture. The patient was also referred to surgical team for colonoscopy but it was not performed due to the risk of bowel perforation.

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These findings established a diagnosis of perianal tuberculosis. He was started on antituberculosis drug with Isoniazid 300mg od, Rifampicin 600mg od, Pyrazinamide 150mg od and Ethambutol 1200mg od. The patient showed a tremendous improvement with antituberculous therapy. The perianal ulcer completely healed following two months of intensive anti-tuberculous treatment (Figure 2). There was also significant improvement of lung radiological findings. He is currently on maintenance phase of antituberculous therapy with Isoniazid and Rifampicin. There was no relapse of lesion and he is planned for a total of 1 year antituberculous therapy.
Discussion

Tuberculosis is a major health problem worldwide. Although there was a significant decrease in morbidity and mortality of tuberculosis due to development of antituberculosis therapy, the number of cases of tuberculosis has increased in recent years in Malaysia due to rising number of AIDS cases and the increased of immigrants from countries that have a higher prevalence of tuberculosis.

Extra-pulmonary tuberculosis is responsible for 15% of all cases of tuberculosis. Extra-pulmonary spread were to the pleura (26%), lymph nodes (17%), genitourinary tract (15%), bones and joints (14%), meninges (6%), peritoneum (4%), miliary TB (8%) and gastrointestinal tract (1%)³. Periorificial tuberculosis is a rare form of extra-pulmonary tuberculosis and is clinically characterized by ulcerative lesions affecting oral, genital, or anal mucosa and surrounding cutaneous areas, as a result of infectious spread from a primary focus in the respiratory, urinary, or gastrointestinal tracts respectively⁴. Perianal tuberculosis is an uncommon manifestation of periorificial tuberculosis which comprises less than 10% of all perianal diseases and 0.7% of all tuberculosis cases⁵. This condition is more frequently seen in men than women (4:1 ratio) and more commonly occurs in the 4th decade of life⁶. The most widely accepted mechanism for perianal tuberculosis is that skin lesions are a consequence of autoinoculation from swallowed sputum containing bacilli that cause secondary digestive lesion⁴. Other mechanism through lymphatic, haematogenous or direct spread from neighboring organs are rare¹. Perianal tuberculosis has also been reported without any presence of gastrointestinal or pulmonary tuberculosis⁷.

Perianal tuberculosis may manifest as an ulcerative, verrucous, lupoid, military and fissure form¹⁻². The most common type is the ulcerative lesion which tends to be painful with well-defined, undermined boundaries and covered by mucopurulent discharge or pseudomembranous material¹⁻². The verrucous type tends to extend into the anal passage from the perianal region with a development pattern similar to that of a wart⁴. However, it may appear as a haemorrhoidal nodule, perianal abscess or anal fistula⁵. In our case, the patient presented with persistent perianal ulceration as his initial manifestation of tuberculosis. Even though his chest x-ray showed significant radiological changes, he did not show any symptoms of pulmonary tuberculosis. In addition to that, his sputum for Acid fast Bacilli was also negative and this may be due to poor specimen collection as the patient was totally asymptomatic for pulmonary tuberculosis. We believe he may have acquired pulmonary tuberculosis initially and then due to the long duration of untreated pulmonary tuberculosis, tuberculosis bacilli could have disseminated to the perianal region via ingestion of contaminated sputum or autoinoculation.

Diagnosis of perianal tuberculosis is difficult and needs a high index of suspicion, especially in patients with perianal involvement as the first presentation of tuberculosis. The diagnosis of perianal tuberculosis is made based on at least one of the following criteria: (i) positive acid-fast bacilli (AFB) stain from the tissue biopsies; (ii) histopathological demonstration of typically caseating granulomatous necrosis; (iii) positive polymerase chain reaction (PCR) for M. tuberculosis on tissue specimen and (iv) tissue biopsy culture positive for M. tuberculosis⁸. The diagnosis is further supported by the clinical response to antituberculosis therapy.

Differential diagnosis of perianal tuberculosis that should be considered are Crohn's disease, ulcerative colitis, pyoderma gangrenosum, syphilis, lymphogranuloma venereum, granuloma inguinale, chancroid, leishmaniasis, deep mycoses, amoebiasis herpes, HSV, CMV, HIV, varicella zoster virus, Cryptococcus neoformans, Mycobacterium avium/intracellulare, sarcoidosis and neoplasias⁹.

Perianal tuberculosis required antituberculosis therapy for at least 6 to 12 months⁸. Those patients with complicated fistulae will need longer antituberculosis therapy for 9 to 18 months⁸. Prognosis is usually good if this condition is recognized early. Complete remission of the perianal lesion is achieved with intensive antituberculosis drugs. Our patient showed complete recovery of perianal ulceration after 2 months of intensive antituberculosis therapy and he was planned to complete 1 year antituberculosis therapy.
**Conclusion**
Perianal tuberculosis is an extremely rare aetiological cause in patients with perianal discharge and ulceration. Perianal tuberculosis should be kept in mind for all patients with any intractable perianal lesion as diagnosis based on the clinical picture is often difficult. Diagnosis is even more difficult to make when there is no previous or active pulmonary infection. Histopathological and microbiological investigations are mandatory in order to confirm this condition since treatment with antituberculosis regime provides complete recovery. Diagnosis of perianal tuberculosis requires high index of suspicion and early recognition will improve the outcome of the disease.

**References**
Obituary

Professor Unandar Budimulja
MD, SpKK, Ph.D, FAADV
12 August 1930 - 2nd April 2013

Professor Unandar Budimulja passed away peacefully on Tuesday, April 2, 2013 at 10.35 pm.

He had dedicated himself selflessly to the development of dermatology in Indonesia and the region. His efforts as a founder council member of the League of Asean Dermatological Societies resulted in the signing of the Bali Declaration of LADS in 1984. As the Founder President, his support and guidance was instrumental in the formation of the Asian Academy of Dermatology and Venereology in 2009.

His involvement and love for teaching spanned many decades since 1962 and he was the Professor of Dermato-Venereology, University of Indonesia since 1991. He was a great leader in clinical dermatology and was a constant figure in all the Regional Conferences of Dermatology (Asian-Australasian) since its inception in 1974. His passion for medical mycology as the Founder and Chairman of Indonesian Medical & Animal Mycology Organization was truly legendary. He continued to serve as the Advisor of the Indonesian Medical Human & Animal Mycology Organization since 1996 and Chairman of the Study Group of Dermatomycosis Indonesia Perdoski.

Professor Unandar was steadfast in the advocacy of high standards in postgraduate dermatologic training in his capacity as the Promoter, Examiner and Assessor of the Faculty of Medicine, University of Indonesia and several other universities in Indonesia since 1986.

He served on the Editorial Board of Media Dermato-Venereology Indonesia and the International Editorial Advisory Board of the Asian Dermatological News and the Indian Journal of Dermatology. He was the Peer Reviewer of the Indonesian Journal of Pharmacology and Therapeutics.

In recognition of his achievements he had received numerous awards including the Satya Lencana Karya Satya Nasional Medal from the President of the Republic of Indonesia in 1990, Lencana Adi Satya Utama Medal from Indonesian Doctors Association in 1997 and the Lencana Satya Bakti Wira Krida Medal from Perdoski in 1999.

In his passing, our dermatology fraternities in Asia have lost an illustrious figure and a friend. He leaves behind a beloved wife and three children and three grandchildren.

Steven Chow
Obituary

Dr. Sivasundram s/o Kathirgamu Alagar
1939 - 17th March 2013

Dr Sivasundram s/o Kathirgamu Alagar was born in Kuala Pilah, Negeri Sembilan in 1939 and studied medicine at the University of Ceylon where he qualified in 1965.

Dr. Siva returned to Malaysia in January 1966 after his housemanship and served as a medical officer in General Hospitals in the states of Selangor and Kelantan. In 1975 he went on to train in dermatology at St John's Hospital for diseases of the skin in London. On his return he worked in Kota Bharu, Kelantan and later in the dermatology department in Hospital Besar Kuala Lumpur where he was gazetted as a Consultant Dermatologist in 1985.

Dr. Siva was then posted to Hospital Tengku Ampuan Rahimah in Klang, Selangor in 1985. He was the Head and Senior Consultant Dermatologist in Klang Hospital from 1985 till March 2004. In Klang, he worked zealously and aided by Dr Saras a senior doctor in the department, went on to establish dermatological services for the state of Selangor. As the only State Dermatologist in Selangor (before the opening of Selayang Hospital), despite his busy schedule in his Department, without fail he used to visit other district hospitals on a monthly basis in Selangor, such as in Banting, Kajang, Tanjung Karang and Kuala Kubu Baru.

Dr. Siva dedicated his whole working life to the service of his patients, especially those less fortunate. Throughout his years of service, he treated his patients to the best of his ability and knowledge. He has been noted to be kind, caring, friendly and sympathetic to his patients.

To the junior doctors who worked and were trained by him, he was regarded as a good mentor, gentle, kind, helpful, easily approachable, knowledgeable, and willing to teach and share his knowledge and ideas and was like a father-figure.

He got along well with his Department staff and worked together as a team. He contributed immensely to the organization and development of his Department and paid special attention to the career prospects of his staff. He was found to be very friendly with a good sense of humour well loved by all. He had a good rapport with other departments in the hospital and worked along well with their consultants.

He had a special interest in dermatological therapeutics and regularly attended the PDM meetings to keep abreast of the recent advances in dermatology.

He retired in 2004 and subsequently served as a visiting Consultant Dermatologist in Klang Hospital on a part time basis till 2006. After retiring from Government Service in 2006, he entered private practice and served as a visiting Consultant Dermatologist in K.P.J (Shah Alam) & Pantai Hospital Klang.

Dr Sivasundram suffered a stroke in 2012 and became bed bound and passed away peacefully after a period of 10 months.

He leaves behind his loving and devoted wife Pathmavathy d/o Selvadurai, a retired secondary school teacher, all his siblings and a host of relatives and close friends.

Dato’ Dr SK Ratti, Dr. Gangaram Hemandas and Dr. Saraswathy
Welcome to the 23rd WORLD CONGRESS OF DERMATOLOGY

IMPORTANT DATES

Online Abstract Submission Opens
January 15, 2014

Early Bird Registration Fee Deadline
January 31, 2014

Website: http://derm2015.org
RCD 2014

Regional Conference of Dermatology (Asian-Australasian)
in incorporating the
6th Annual Meeting of the
Asian Academy of Dermatology and Venereology

Theme: Enhancing Best Practice in Clinical and Cosmetic Dermatology

IMPORTANT DATES

Early Registration
31st October 2013

Abstract Submission
31st October 2013

Congress Administration
G-1, Medical Academies of Malaysia, 210, Jalan Tun Razak, 50400 Kuala Lumpur, Malaysia
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Email: secretariat@asianderm.org
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