LETTERS TO THE EDITOR

IS MELKERSSON-ROSENTHAL SYNDROME A DISTINCT CLINICAL ENTITY
(A retrospective study of eight cases)

Eight patients, 3 females and 5 males, diagnosed clinically as Melkersson-Rosenthal syndrome (MRS), seen during the last three years, were studied. All eight patients showed the classical clinical triad of labial oedema, facial paralysis and lingua plica. Although there was history of waxing and waning of labial oedema and facial palsy in all the eight cases, these features were well established and had turned permanent at the time of examination. There was no positive family history in any of the patients. One patient, a 40-year-old male, had four hypopigmented and hypoaesthetic patches on his trunk, suggestive of borderline tuberculoid Hansen's disease. Apart from this, none of the patients had any clinical signs of any dermatological or systemic disease.

Biopsy from the swollen lip in all eight cases, showed non-caseating granulomatous foci, comprised of predominantly epithelioid cells, lymphocytes and a few histiocytes. Other investigations to rule out tuberculosis, parasitic infestation and septic foci were not contributory. Kveim test was not done due to non-availability of the antigen.

The patients were all treated with dapsone and reviewed once a month. Even after six months of treatment, there was no regression of any of the three signs. An interesting feature was that the patient with Hansen's disease showed resolution of the leprotic patches but his labial oedema and facial paralysis remained unchanged. Systemic corticosteroids, antibiotics, antiamoebics and antihelminthics were of no avail.

MRS has been described to be a reaction pattern of systemic diseases such as Crohn's disease, sarcoidosis and other granulomatous diseases.[1,2] There have been reports of MRS being associated with leukemia.[3] Some authors have suggested a hereditary factor.[4,5] But from the observations made in this study, wherein all the eight cases of MRS showed no suggestive aetiological factor and no response to any form of treatment, it may be opined that MRS is a distinct clinical entity of undetermined aetiology, although the syndrome could masquerade as any form of disease, including Hansen's disease.[6] A study on a larger number of cases may throw more light on this subject.

Jayakar Thomas and TC Muthuswami,
Department of Dermatology and Leprosy,
Government General Hospital,
Madras-600 003, India.

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TUBEROUS SCLEROSIS IN IDENTICAL TWINS

As I was going through the journal, I noticed the above-named paper by Govil and Govil (IJDVL, 1987; 53 : 127-128). The authors have described the cutaneous lesions of the twin brothers in detail and state that 'only Eapen et al have reported tuberous sclerosis among identical twin sister's. Eapen et al described 6 cases which includes a very brief description of the twins and conclude that 'occurrence of tuberous sclerosis in identical twin sisters which is being reported for the first time' (IJDVL, 1984; 50 : 60-63). I disagree with these authors for they have overlooked an important paper published much earlier. Vasista from Varanasi, India in her Letter to the Editor has clearly presented identical twin brothers with this condition and has described lucidly the criteria she used, such as the weight, height, blood group and finger prints etc, of the twins to diagnose zygosity. (Arch Dermatol, 1981; 117 : 456).

Whilst I agree with Pasricha and Seetharam that 'the problem with most western writers is that they almost completely ignore the Indian publications from their reviews even when these publications are in international journals (IJDVL, 1987; 53 : 52), it was unfortunate that a good piece of Indian contribution on this subject went unnoticed and therefore unquoted.

Kader Naina Mohamad,
Department of Dermatology,
Sultanah Aminah General Hospital,
80100, Johor Bahru, Malaysia.

ALOPECIA UNIVERSALIS, ANODONTIA, CALCINOSIS CUTIS, CATARACT, CARDIOMYOPATHY AND ARACHNOIDITIS

A 35-year-old male patient, delivered at full-term without any complications and having a parental history of consanguinity, lost his deciduous teeth spontaneously at the age of two years leading to partial anodontia which was later followed by complete anodontia. Hairs were lost all over the body at the age of 5 years leading to alopecia universalis. Multiple small, nodular calcifications appeared all over the skin at the age of 15 years. The patient developed gradually progressive weakness of the lower limbs with pain, first of the right limb followed one year later in the left limb, and strained and painful micturition at times associated with nocturnal bed-wetting, at the age of 30 years. His vision diminished gradually since the age of 32 years. The patient developed herpes zoster while in the hospital. There was no history of cough, haemoptysis, prolonged fever, weight loss or trauma to spine, however, the patient complained of frequent attacks of fever and pyodermas. None of his two sons aged 9 and 6 years, a daughter aged one year or other family members had a related illness.

The patient had pallor and pedal oedema, and the jugular venous pressure (JVP) was raised. Liver was enlarged 3 cm below the intercostal margin and soft. There was no ascites. The prostate was normal. The lungs were clear. The neurological examination showed normal speech, cranial nerves and higher functions. Cerebellar signs were absent. The motor and sensory examination of upper extremity was normal. In lower extremity, the sensory examination showed impaired (50%) vibration sensations below L_1 segment and (25%) below S_3 segment over the right leg. The tone of the muscles was decreased in both the lower limbs. The power of the muscles of the right leg was grade II/III and that of left lower limb was grade III/IV. Fasciculations

Fig. 1. Alopecia, anodontia and nodules over the scalp and face.

were absent and co-ordination of muscles was normal. The reflexes were normal in upper limb while knee jerks and ankle jerks were absent in both the lower limbs. Plantar was not elicitable bilaterally. Cremasteric reflex was absent over right side. Examination of the cardio-vascular system showed features of left ventricular hypertrophy and murmurs of MS, MR and AR. Ophthalmological examination showed bilateral immature cataract. There was no bony deformity. Genitalia were normal. The dermatological examination revealed a normal texture of the skin with normal sweating. Multiple nodular firm swellings, varying in size from 1 cm to 3 cm, were present all over the body including the scalp. The hairs and teeth were completely absent. The nails were normal.

Haematological investigations including serum calcium and phosphorous levels were normal. Urine and stools were also normal.

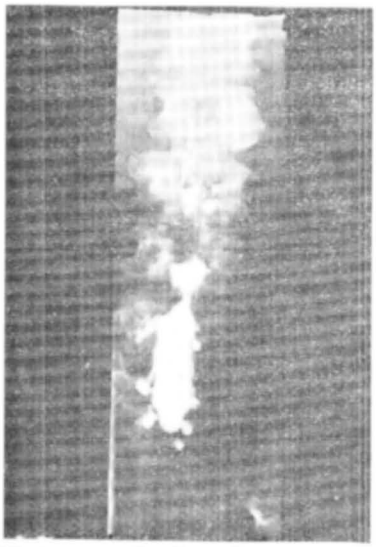

Fig. 2. Myelogram showing the "candle-guttering" appearance suggestive of arachnoiditis in the lumbar region.

VDRL test and Mantoux test were negative. ECG showed features of left ventricular hypertrophy. EEG, IVP and roentgenograms of skull, long bones and chest were normal. CSF examination was normal apart from a raised protein content. Myelography showed features of arachnoiditis in the lumbo-sacral region. Study of chromosomal pattern could not detect any abnormality. Histopathological examination of the cutaneous nodule showed features of calcinosis cutis and that of uninvolved skin showed normal features including the sweat glands.

Multisystem involvement in this patient in the form of alopecia universalis, anodontia, calcinosis cutis, cardiomyopathy, bilateral cataract, and features of lumbar arachnoiditis with a parental history of consanguinity is interesting. It is not known how far these manifestations are inter-related.

P K Nigam, Gurmohan Singh and Lata Sharma

Department of Dermatology, Venereology and Leprosy, Institute of Medical Sciences, Banaras Hindu University Varanasi-221 005, India.

SKIN METASTASIS IN CARCINOMA PENIS

The recent article on the rarity of cutaneous metastasis from carcinoma of the penis[1] has prompted me to describe a similar experience. A 58-year-old man came with swellings and discharge over the lower abdomen and thigh of 3 months duration (Fig. 1). Past records showed that 6 months earlier, he had undergone partial amputation of the penis for an ulcerative lesion on the glans which was diagnosed histopathologically as squamous cell carcinoma. Three months later, he noticed a painless swelling, increasing in size and extent, over the right inguinal region followed by similar involvement of the lower abdomen and upper thigh on the left side. On the right, the swelling broke down forming 2-3 sinuses. An oval mass 6 cm × 3 cm, non-tender, firm and arising from the lymph nodes was felt in the right inguinal region. The sinuses were centrally located, discharging sero-purulent material and the surrounding area was covered with crusts overlying friable skin tissue. Two separate nodules, each measuring 3 cm × 2 cm, firm and non-tender were present in the left lower abdomen above the inguinal region and the middle of the upper thigh respectively. The nodules were subcutaneous and the left inguinal lymph nodes were slightly enlarged, discrete and non-tender. The overlying skin was freely mobile and appeared normal. The amputated end of the penile shaft appeared healthy. Scrotum and scrotal contents were normal. Routine blood and urinalysis, and chest X-ray were normal. Mantoux test read 10 mm. Culture of discharge from the sinus grew *Streptococcus* and biopsy from the adjoining region showed chronic nonspecific dermatitis with no acid-fast bacilli on Ziehl-Neelsen stain. Culture of the skin tissue for *M. tuberculosis* was negative. Biopsy from the intact nodule over the left thigh revealed squamous cell carcinoma.

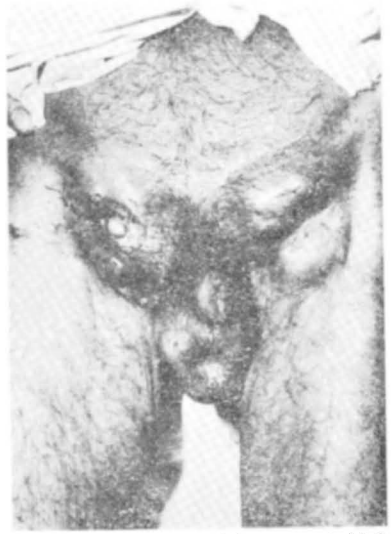

Fig. 1. Swellings over lower abdomen and thigh.

The possibility of cutaneous metastasis was suspected from the history and confirmed by biopsy. Attempts were made to rule out coexisting scrofuloderma in the right inguinal region. It is presumed that these were also due to metastasis, and secondary infection had altered the histopathologic picture or a deeper biopsy from a different site may have been confirmative.

V Ramesh
Department of Dermatology
and Leprology,
Safdarjung Hospital,
New Delhi-110029, India.

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