Cutaneous leiomyoma in two sisters

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Cutaneous leiomyoma (CL) is an uncommon cutaneous tumour arising from the smooth muscle fibres in arrectores pilorum muscle, mammary glands, scrotum, labia majora or the wall of blood vessels[1]. CL arising from arrectores pilorum muscle are characterized by multiple dusky brown tender nodules often occurrence on the arms[2]. The occurrence of CL in family members has been reported in the medical literature but is rare[1,3]. We report here two sisters with this condition.

Case report

A 27 year old housewife (VJ) from Nakulugamuwa presented to the skin clinic at General Hospital Matara with multiple skin coloured nodules and papules on the right arm of 7 years duration.

The lesions were slowly growing in size and numbers during this period. Most of the nodules were painful when touched or compressed. Occasional pain was experienced even without any stimulation. She has had one painful nodule removed surgically about one year ago.

She had no other significant medical problem. There was no parental consanguinity or similar symptoms in her parents. However on direct questioning she admitted that her sister has similar but smaller nodules on her right arm.

On examination she was a slightly obese well built women in good general health. She had multiple dusky brown nodules and papules varying from a few millimetres to 1.2 centimetres in diameter on the right arm mostly on the lateral aspect (Figure 1). The larger lesions were painful to touch and compression. There was one scar of a previous surgical excision of a nodule.

She had no other nodules in other areas of the body and the systems examination was normal. An excision biopsy of the most painful nodule was done. The specimen was fixed in 10% formol saline and was histologically analysed.

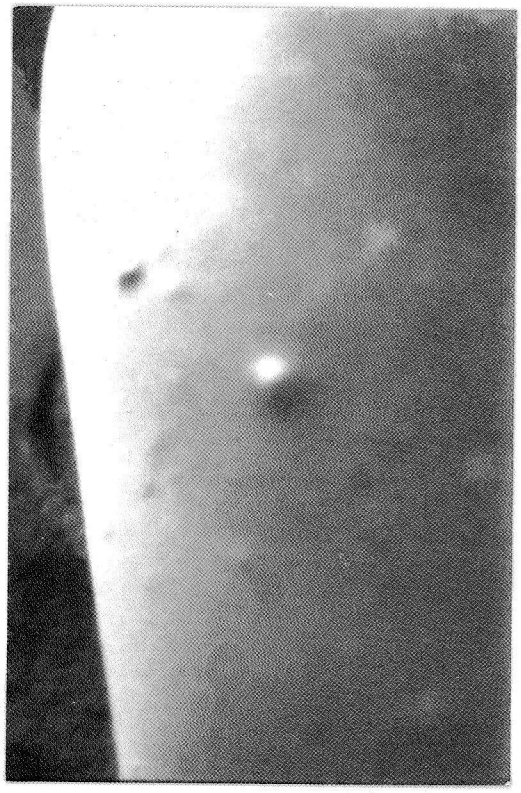

Figure 1. Dusky brown nodules on the right arm

The haematological investigations showed; Hb 15g%, PCV 45%, WBC count 10600/mm^3 with 39% neutrophils, 60% lymphocytes and 1% eosinophils, a normal blood picture and an ESR of 8mm. The liver transaminases were normal.

Haematoxylin and eosin (H & E) stained sections showed an ill circumscribed dermal proliferation of interlacing bundles of spindle cells with scanty collagen (Figure 2). The spindle cells had elongated blunt ended ("cigar shapped") nuclei (Figure 3) and eosiniphilic cytoplasm. They

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stained red with Masson trichrome and stained yellow with Van Geison stain confirming the smooth muscle origin. There was no storiform appearence, vascular structures or myxoid areas with serpigenous nuclei to suggest a dermatofibroma, vascular leiomyoma or neurofibroma respectively.

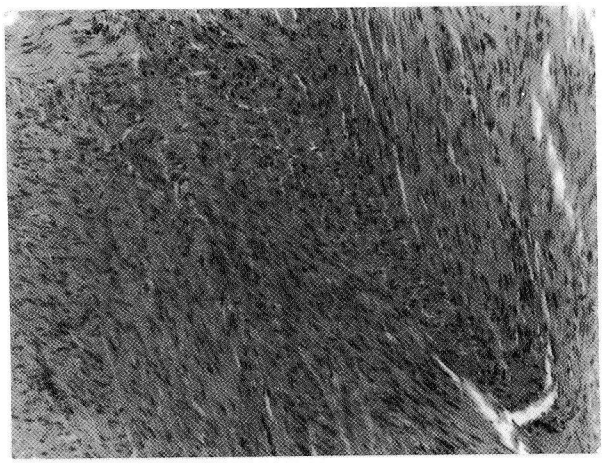

Figure 2. Interlacing smooth muscle fibres in the dermis (H & E, 10 × 10)

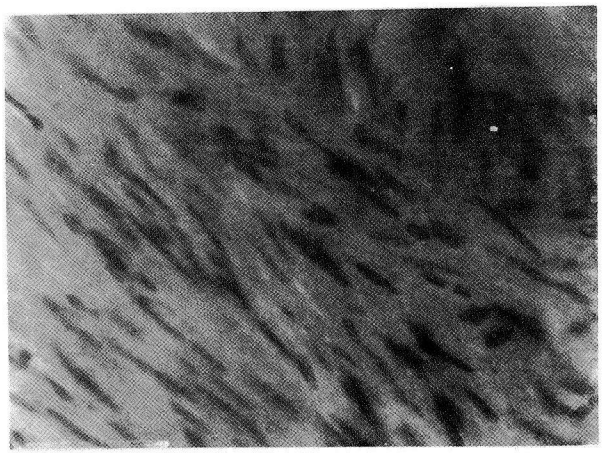

Figure 3. Elongated blunt ended nuclei ("cigar shapped") of spindle cells (H & E, 10 × 40)

Examination of the sister of the patient (29 years) revealed similar but smaller dusky brown papules and nodules on the right arm. Some of these were painful to touch and compression.

Discussion

Cutaneous leiomyoma arising from pilomotor muscle is commoner than dartotic myomata and angioleiomyomata. In our patient the clinical history, examination findings and histologic and histochemical appearences were characteristic of leiomyoma cutis (CL).

Clinically, dusky brown, painful or tender nodules and papules enlarging slowly in the region of upper limbs or trunk makes one suspect cutaneous leiomyoma. Neurofibromata, dermatofibromata and multiple glomus tumours were thought to be unlikely even though they can give rise to painful tender nodules on the body.

Our patient did not show any evidence of overt erythrocytosis although the Hb (15g%) was slightly above the normal Sri Lankan average for a woman. She also did not have any clinical evidence of uterine fibroids. Erythrocytosis and uterine fibroids have been reported to be associated in some cases of CL[2,3].

The exact mode of transmission of this condition is not known although autosomal dominant inheritance with incomplete penetrance is likely[1]. Verma et al (India, 1973) reported leiomyoma cutis in two brothers and Butler (USA, 1984) reported its occurrence in mother and daughter[1,3]. Most of the familial leiomyomata reported have been multiple. Mamillary and dartotic leiomyomata are usually solitary and are not familial[2].

The course of CL is usually benign but progressive. No definite treatment is available for multiple lesions of CL. Excision of lesions often cause appearance of new lesions in the same area[2]. The severity of pain may make the patient demand treatment and extensive lesions may require plastic surgery[2]. In our patient also two most painful nodules were removed for relief of symptoms. The sister of the patient elected not to undergo biopsy or surgical treatment.

References

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