LEIOMYOMA CUTIS

(A clinico-pathological review with a case report)

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Summary

The classification and clinico-pathological review of leiomyoma cutis is presented.

A case of multiple hair muscle leiomyoma in a male patient is described.

Stout believes that the incidence of multiple lesions occurs twice as often in men than in women.

No familial or hereditary factors were seen in the case reported, though Kloepfer reported a familial case.

The painful nature of the lesions brought the patient to seek medical attention, this symptom we believe is of importance in diagnosing multiple leiomyoma.

The skin biopsy confirmed our clinical diagnosis of multiple hair muscle leiomyoma.

Classification:

Besnier (1880) classified Leiomyoma of the skin as solitary or multiple. The former occurred in the dartoid zones i.e. the perianogenital, axillary regions and the nipple. Multiple lesions might occur anywhere on the body.

Babe's (1884) on anatomical and pathological data grouped leiomyoma as (i) Leiomyoma derived from the smooth muscle of blood vessels. (ii) Those derived from the musculi arrectores pilorum, sometimes in *naevi.* (iii) Those derived from the smooth muscle layer of the 'dartoiques zones'. Included in this group are the more diffuse leisons which occur on the genitalia, legs in combination with lymphan-giectasia and Bazins pachydermia myxomatodes. (iv) Those derived from the smooth muscle of deeper tissues or from embryonic displace ments – bronchogenic leiomyoma belongs here.

Other types have subsequently been added to this classification, such as those originating from the smooth muscle of sweat glands, those from the superior tarsal muscle and a number of mixed types.

Recently, Fitzpatrick, et al[2] classified leiomyoma in three types (i) multiple cutaneous leiomyoma (ii) solitary angioleiomyoma and (iii) solitary genital leiomyoma (myomes dartoiques).

Aetio-pathology

Leiomyoma of the skin may occur at any age. Some familial hereditary cases have been observed. Kloepfer et al (1958) reported multiple leiomyomas

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in a family and related the disease to the presence of an autosomal dominant gene. Stout (1937) felt that multiple lesions occur twice as often in men as in women, but solitary tumours occur equally in the two sexes. They vary in size from pin-point to that of a man's head and in hue from skin colour to reddish brown and grow slowly, small tumours may spontaneously disappear. They may be difficult to diagnose particularly if the typical pain is absent. This pain is described as stabbing, pinching, burning or griping and is usually limited to the tumour. Sometimes it may radiate and may be paroxysmal. It may arise spontaneously or be provoked by emotional stress, fatigue, cold, menstruation or pregnancy, or by pressure, especially when manipulated. It is believed that contraction of the tumour gives rise to pain through ischaemia, or by pressure or traction on nerves.

Grzybowski (1933) demonstrated neurofibrils in leiomyomata by silver impregnation and believed them responsible for the symptoms. Halter and Hornemann (1952) hold the same opinion. Peyri (1943) Ekestrom (1950) suggested a relationship between vascular leiomyomata and painful glomus tumours. An angioleiomymatous glomus tumour was described by Vilanova and Grace – Barbera (1944). Neither Ekestrom, Saunders and Fitzpatrick (1956) succeeded in demonstrating neurofibrils in vascular leiomyomata. Jansen and Dreesson (1958)[8] did not observe an abnormal amount of nerve tissue in material prepared by silver impregnation technique. They gained the impression that leiomyoma were poorly provided with nerves: Ormea (1951) noted hypertrophy and degeneration of nerve tissue, swollen neurofibrils in serpiginous arrangement, with dilatations and changes in the terminal network. These accompanied changes in smooth muscle i.e. homogenitation and degeneration of nuclei. Nodl (1953) found similar microscopic changes, and believed the primary importance had to be attributed to the hypertrophy of the terminal network, he found changes in nerve tissue where no changes in muscle tissue could be observed. He concluded that multiple leiomyomata was to be looked on as a neurocutaneous syndrome.

Leiomyoma derived from hair muscle is the commonest form encountered (Sutton 1912). They sometimes occur in naevi – Jesionek and Werner (1907) described such a case as 'naevus verrucosus pigmentosus' to which adjective 'leiomyomatosus' might justifiably be added.

The diagnosis of hair muscle leiomyoma can be made if it can be demonstrated that muscle fibres of the arrectores pilorum fuse with the myoma. Hair muscle leiomyoma are seated in the dermis, they are not encapsulated. The muscle fibres run in bundles in every direction appear to invade the surrounding connective tissue (pseudo infiltrative growth) but signs of malignancy are absent. Occasionally foci of degeneration are seen as in uterine leiomyomata, however malignant degeneration is extremely rare.

Sobotka (1908) pointed out that smooth muscle fibres unconnected with hair follicles may be presented at the junction of the cutis and subcutis around the umbilicus, forehead, cheeks, ears, back and extensor surfaces of the thighs. It is therefore difficult to judge whether a given leiomyoma originated from 'embryonic nests', as a rule, these leiomyoma are located deeper than the hair muscle tumours, but sometimes it may be impossible to differentiate them.

Leiomyoma derived from deeper tissue and from embryonic nests may arise from the puborectal aponeurosis,

the recto-vaginal septum or the prostate gland. They are surrounded by firm fibrous capsules and may attain a considerable size.

Multiple leiomyomas may be present in association with other soft tissue tumours such as lipomas, fibromas dermoid tumours, epidermoid inclusion cysts. These lesions may co-exist with bony exostoses and intestinal polyposis (Gardener's syndrome)–Weary et al[6].

Pathologically, leiomyomas are spheri cal and show a whorled cut surface, those that originate in the dermis are usually not encapsulated, though subcutaneous leiomyomas are usually encapsulated. Vascular leiomyomas are encapsulated (Magner and Hill, 1961). In the leiomyoma are seen spindle cells arranged in interlacing fascicles. The nuclei are elongated and cytoplasm contains longtitudinal fibrillae. In vascular leiomyomas the smooth muscle cells are arranged in concentric fashion around vascular spaces, the vessels in the tumour do not contain elastic laminae (Stout 1937).

Solitary leiomyoma must be differentiated from glomus tumour, eccrine spiradenoma, tricho-epithelioma cylindroma and neurofibroma. Both are non encapsulated, however the spindle cells in leiomyomas are shorter and have abundant fibrillar cytoplasm, and the interlacing fascicles are thicker than the delicate loosely arranged fascicles of neurofibroma.

Rarely, a subcutaneous smooth muscle tumour shows nuclear atypicalism and mitoses. A few lesions of this type are proved to be primary leiomyosarcomas (Stout & Hill 1958). Others may prove to be metastasis from an occult, deeply situated leiomyosarcoma. Primary leiomyosarcoma that arises in the subcutaneous tissue metastasize to the lungs and rarely lymphnodes (Phelan et al 1962. Rising and Booth[5]). However there is no good evidence that the rare superficial leiomyosarcomas arise from pre-existing leiomyomas (Fisher & Hillwig)[1].

Histochemically, leiomyoma differs from fibroma, neuroma and neurilemmoma by the high concentration of amylophosphorylase in the tumour cells (Niemi[4]). Although the Bodian stain reveals no increase in nerve fibres in leiomyoma (Montgomery and Winkalmann 1959), histochemical, staining using the acetylcholine esterase technique demonstrates considerable proliferation of neural elements (Niemi)[4]. Electron microscopy shows that the tumour is composed of rather immature smooth muscle cells containing a small to moderate amount of glycogen. (Fitzpatrick, et al[2]).

Case Report

A 45 years old male patient attended the skin O.P.D. of the Nair Hospital, Bombay with complaints of numerous nodular swellings over the chest, back, upper extremities and thighs since last 10 years. The lesions were associated with severe pain since the last 5 years, and since the lesions were now painful the patient came up for dermatological attention.

There was no history of similar lesions in his family. He was average built and nourished. Routine investigations and systemic examination showed no abnormality. Local examination showed numerous nodular lesions over the sternal region and sides of the chest (Fig 1). Besides similar lesions were seen over the midline of the back and sides, supra-scapular region, extending over the deltoid regions-bilaterally (Fig. 2), and over the upper arms. Sparse lesions were also seen over the thighs. The lesions were of diverse sizes and shapes, some were discrete and others tended to conglumerate, they were flesh coloured, reddish brown movable and not attached to the deeper t ructures.

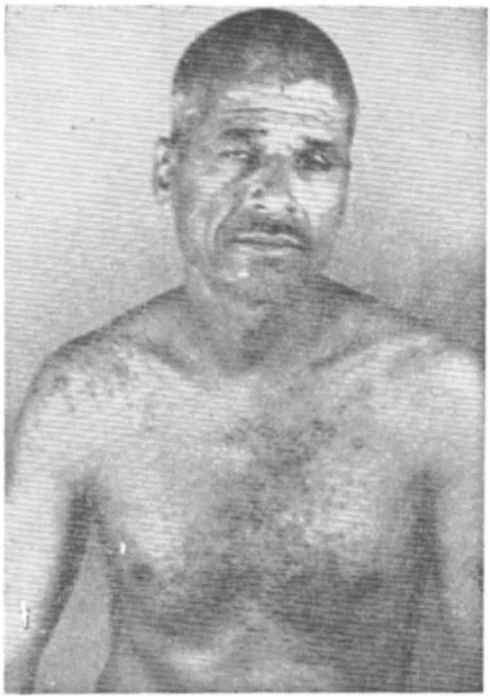

Fig. 1
Multiple hair muscle lesions over sternal region and sides of chest.

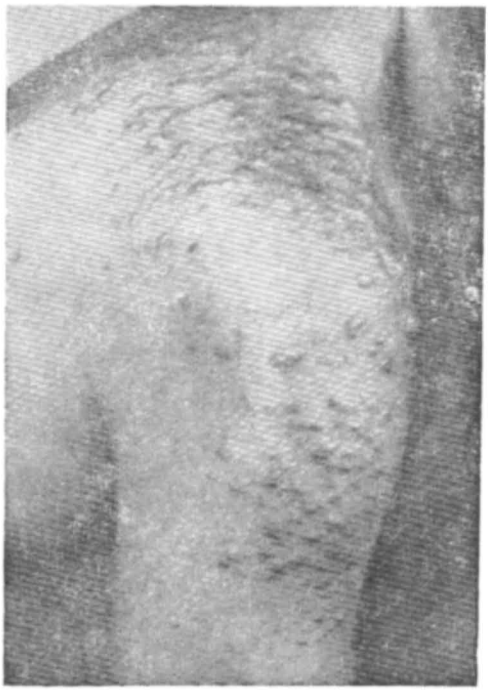

Fig. 2
Lesions over the supra scapular and deltoid region.

These lesions were extremely painful, the pain was stabbing in nature and at times of a dull ache, on pressure the pain was at times severe, and particularly the lesions pressurised during sleeping posture, would produce severe pain, at times disturbing his sleep. It was primarily the severe pain of the lesions, and the difficulty to sleep in any posture, which pressurised the lesions producing pain, that forced the patient to seek dermatological attention. Pressure manipulation of the individual lesions showed and confirmed the painful nature of the lesions.

Skin Biopsy

H. E. stained sections showed interlacing bundles of smooth muscle fibres staining pink traversing the corium in various directions, collagen bundles were seen intermingled with the smooth muscle bundles. The muscle fibres were

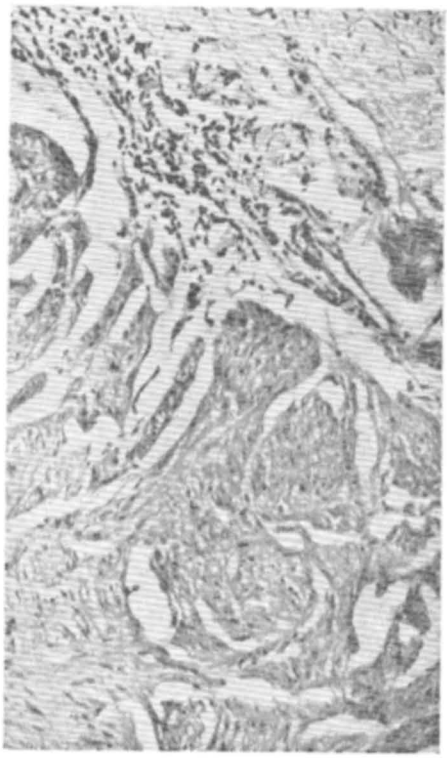

Fig. 3
H. E. Stain.

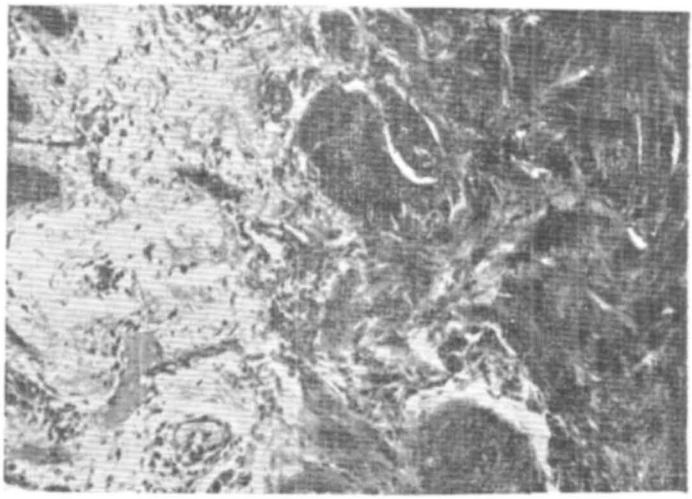

Fig. 4
Masson's stain.

slightly wavy and showed long blunt-edged nuclei, (Fig. 3). Masson's stained sections (Fig. 4) showed the muscle tissue as dark bright red areas and collagen blue.

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