

## MULTIPLE CUTANEOUS LEIOMYOMATA

**Dr. Hamda Al Ansari**

*Department of Dermatology and Venereology*

**Dr. Bahram Azadeh**

*Laboratory Med. and Pathology Department*

### Introduction:

Leiomyoma is a benign muscle tumor arising from erector pili, smooth muscle of scrotum, labia majora or nipples and from media of blood vessels. The most frequent is leiomyoma cutis arising from erector pili (Pilar leiomyoma). They appear as firm painful, tender dusky brown nodules which are usually less than 15mm in diameter. The tumor usually starts as a single nodule which increases in size and number and may coalesce to form plaques. They may be unilateral or affect more than one site. Less commonly the leiomyoma arises from smooth muscle of scrotum and nipple. Angiomyomas arise from media of blood vessels and is usually solitary subcutaneous nodule most frequently seen on lower limbs.

### Case Report:

A male patient 30-years old from Srilanka presented with painful swellings of his gluteal region. The course was progressive and the nodules increased in size and number over a period of one month. The patient sought medical advice and was diagnosed as multiple gluteal

abscesses and surgical drainage was attempted without improvement. Patient was referred to dermatology in April 2001, and on examination he had on his left gluteal region multiple greyish nodules which were tender and painful and two showed site of surgical incision - the nodules were firm and the regional lymph nodes were not enlarged. (Fig. 1, 2).

The biopsy showed epidermal hyperplasia (Fig. 3). Fascicles and bundles of fusiform cells with blunt-ended "cigar-shaped" nuclei. This proliferation had infiltrated all dermal layers with entrapment of sweat glands and hair follicles (Fig. 4, 5). Features of fibrous histiocytoma like foamy macrophages, multinucleated giant cells and iron pigments were not present. There were no abnormal mitotic figures. Foci of necrosis (Fig.6) with infiltrates of macrophages and neutrophils were noticed presumably due to earlier attempts at incision and drainage by a surgeon who had thought the lesions were abscesses. On immunostaining tumor cells were positive for vimentin, smooth muscle (Fig., 7, 8, 9) actin and HMF35, but were negative for desmin, CD 34, S100 protein, HMB 45, neurofilament and CD 68, the latter highlighted the inflammatory macrophages. These light microscopic and immunohistochemical findings were consistent with multiple cutaneous leiomyomata, although myofibroblastomas were considered in the differential diagnosis. Complete excision was advised for treatment and exact classification of the tumor.



*Fig. 1 & 2: Multiple Leiomyomata of gluteal region.*

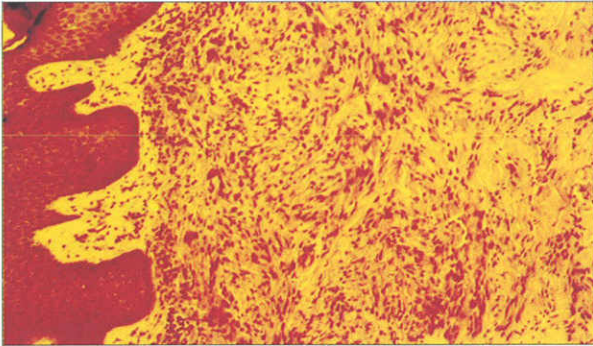


Fig. 3: Epidermal hyperplasia with bundles of fusiform cells infiltrating all dermal layers superficially simulating fibrous histiocytoma (H & E).

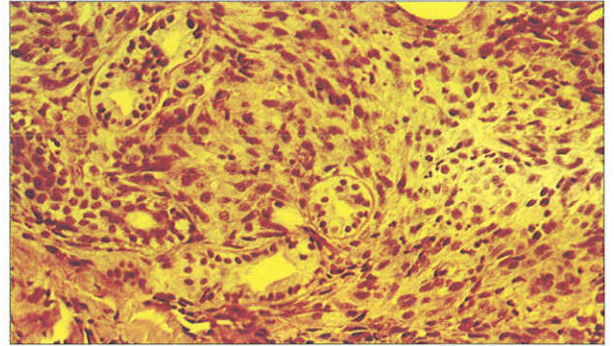


Fig. 4: Entrapment of sweat glands by bundles of fusiform tumor cells (H & E).

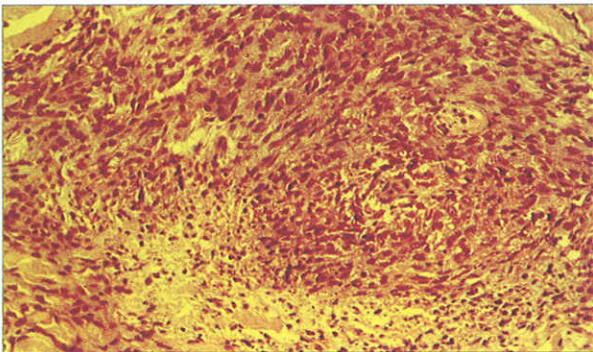


Fig. 5: Bundles of fusiform cells have encircled sweat glands and a hair follicle. Note prominent nucleoli in blunt-ended nuclei of the tumor cells suggesting smooth muscle differentiation (H & E).

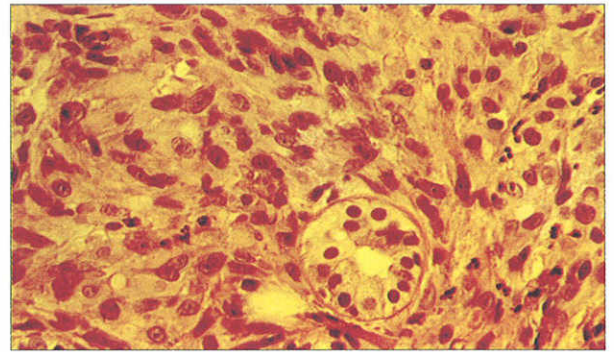


Fig. 6: Fascicles of tumor cells in deep dermis associated with necrosis and inflammatory cellular infiltrates (H & E).

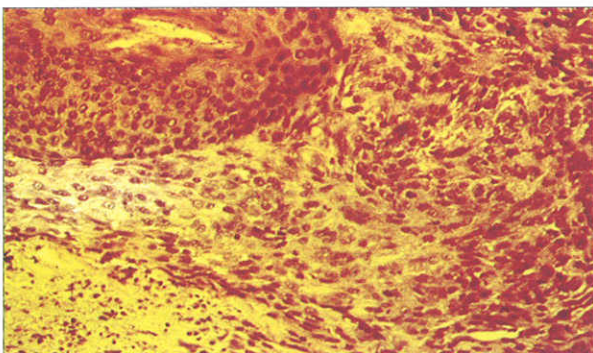


Fig. 7: Tumor cells in the deep dermis around a hair follicle. Note necrosis and inflammatory cellular infiltrates (H & E).

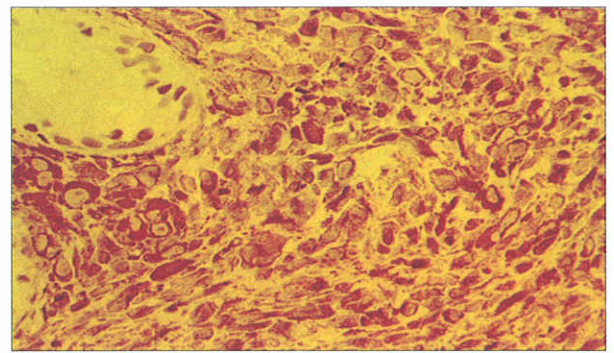


Fig. 8: Tumor cells show positive immunostaining for vimentin. Compare with the negatively stained epithelial cells of the sweat gland (ABC).

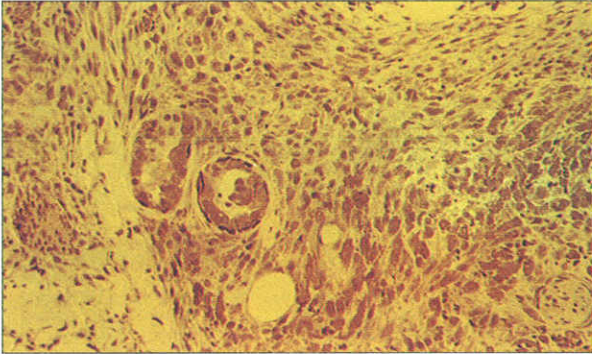


Fig. 9: Tumor cells show positive immunostaining for smooth-muscle actin. Compare with strongly positive arrecta muscle, and myoepithelial cells of the sweat gland and a negatively stained nerve (ABC).

#### Discussion:

Light microscopic appearances including elongated blunt-ended nuclei and positive immuno-histochemical staining for vimentin, smooth muscle actin and HHF 35 seen in the present case are consistent with (multiple) cutaneous leiomyomata; negative staining for desmin by itself does not exclude a diagnosis of leiomyoma. However, myofibroblastic tumors should be considered in the differential diagnosis. Tumor cells in myofibroblastomas

consist of stellate to spindle cells with tapered or sharp-ended nuclei. Myofibroblasts are typically positive for vimentin and alpha-smooth-muscle actin. Desmin is not a useful discriminant between leiomyomas and myofibroblastomas<sup>(1)</sup>. In a series of malignant myofibroblastic tumors (myofibrosarcoma) immunohistochemistry showed smooth muscle actin in 13 of 15 cases, muscle-specific actin in 7 of 9 and desmin in 6 of 14<sup>(2)</sup>. Electron microscopy will be helpful for differential diagnosis between smooth muscle and myofibroblastic tumors. Ultra structurally, myofibroblasts have a surface characterized by prominent fibronectin fibrils and fibronexus junctions, which are distinct from “basement membrane”. This can permit a distinction to be made between smooth muscle and myofibroblastic tumors. Myofibroblasts have abundant rough endoplasmic reticulum; modestly developed peripheral myofilaments with stress fibers and fibronexus junctions<sup>(1)</sup>. The disease is rare and is difficult to diagnose histopathologically and often requires surgical excision to alleviate pain although complete surgical excision may not be attained<sup>(3)</sup>.

The present case showed unilateral multiple nodules of left gluteal region and patient’s main complaint was pain and tenderness. Treatment is by surgical excision. Medical treatment namely Calcium channel blockers may alleviate pain.

#### References:

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- 2- Montgomery E, Goldblum JR, Fisher C. Myofibrosarcoma: a clinicopathologic study. *Am J Surg Pathol*; 2001. 25: 219-28.
- 3- Latoni JD; Neuburg M; Matloub HS: Pilar Leiomyoma: a case report and review of the literature. *Ann-Plast-Surg*. 2000;