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RESEARCH ARTICLE

CUTANEOUS LEIOMYOSARCOMA: A RARE PRESENTATION

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ABSTRACT

Cutaneous leiomyosarcoma is a rare malignant soft tissue sarcoma that accounts for about 2-3% of all superficial soft tissue sarcomas. It mostly present in middle aged to elderly males with a predilection for lower limbs. Due to non specific morphologic features with a host of differential diagnosis, immunohistochemistry plays important role in arriving at the final diagnosis. We present the case of 77 year old woman with cutaneous leiomyosarcoma located on the upper back.

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INTRODUCTION

Soft tissue sarcomas are rare, among which leiomyosarcoma is a malignant neoplasm of smooth muscle cells which usually affects the uterus, gastrointestinal tract, and retroperitoneum (Pop *et al*., 2011). The etiology of leiomyosarcoma is unknown. Some relationship has been established with radiation, chemical exposure, chromosomal defects and trauma (Monsef Esfahani *et al*., 2004). Superficial leiomyosarcomas can be subdivided into two types according to its primary site of origin: deep subcutaneous and superficial cutaneous types (Lin and Tsai, 1999). Deep subcutaneous leiomyosarcoma is comparatively more common than superficial cutaneous subtype. Primary cutaneous leiomyosarcoma (PCL) of the skin is a rare soft tissue tumor that accounts for about 2-3% of all superficial soft tissue sarcomas (Lin and Tsai, 1999). It may affect any body site but most commonly involve the lower limbs.

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We hereby report a case of PCL in a 77 year old female who presented with a nodular mass on the right side of the back.

The Case

A 77 year old female patient with no significant past history presented with a painless pedunculated mass on the back, which had been progressively increasing in size since past 6 months. Systemic examination and investigations, including complete blood count, fasting blood sugar levels, liver function and renal function tests were within normal limits. A clinical diagnosis of malignant mesenchymal lesion was considered. Resection with a 5 cm resection margin was performed and specimen was analysed for histological examination. Gross examination revealed a partly skin covered grey brown, soft tissue mass measuring 7 cm x 6.5 cm x 4 cm. Overlying skin showed brownish discolouration. The cut surface of the mass was greyish white, solid, fleshy with few hemorrhagic areas. (Figure1) Microscopic examination revealed a circumscribed neoplasm originating from the dermis and extending into the underlying subcutis.

Figure 1. (A) Gross photograph showing an irregular, pedunculated skin covered swelling, measuring 7.5 cm x 6.5 x 4 cm, with overlying skin showing blackish discolouration. (B) Cut surface shows a greyish white, solid, fleshy irregular tumor mass with areas of haemorrhage

The growth pattern of the neoplasm was predominantly fascicular, with the tumor bundles intersecting each other at wide angles. Individual tumor cells had acidophilic fibrillary cytoplasm and pleomorphic nuclei with coarse and irregularly dispersed chromatin. Mitotic count of 6 mitotic figures per 10 high power fields along with atypical mitosis and multinucleate tumor giant cells were seen. (Figure 2) Focal necrosis was noted at the edges. The overlying epidermis was thinned out. The tumor was grade II according to French federation of cancer centre sarcoma group grading. A provisional diagnosis of spindle cell malignant neoplasm of the skin was kept. Numerous diagnostic possibilities including fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, monophasic synovial sarcoma and spindle cell variant of squamous cell carcinoma were considered and immunohistochemistry was used for confirmation. On immunohistochemistry, tumor cells showed strong immunopositivity for Smooth muscle actin (SMA) and Vimentin (Biogenex USA). (Figure 3) Epithelial Membrane Antigen (EMA), CD34 and S-100 markers were negative. Based on the microscopic and immunohistochemistry findings, a final diagnosis of primary cutaneous leiomyosarcoma was rendered. The patient is currently on periodic follow-up since 18 months and no recurrence has been identified so far.

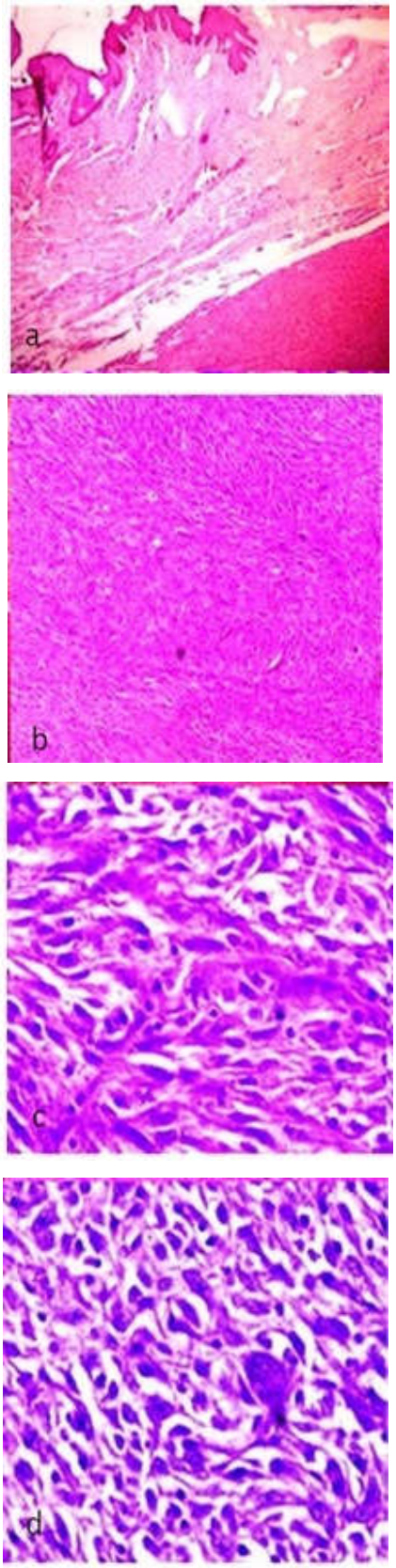

Figure 2. (A) Light microscopy revealed a dermal neoplasm with extension into the subcutaneous tissue (H & E, x40). (B) Neoplasm showed interlacing fascicles of elongated spindle cells (H & E, x100). (C) Tumor cells are spindle shaped with moderate to abundant pink, fibrillary cytoplasm and pleomorphic hyperchromatic nuclei frequently exhibiting atypical mitosis (black arrow) (H & E, x400). (D) Tumor cells with markedly pleomorphic nuclei and multinucleate tumor giant cell (H & E, x400)

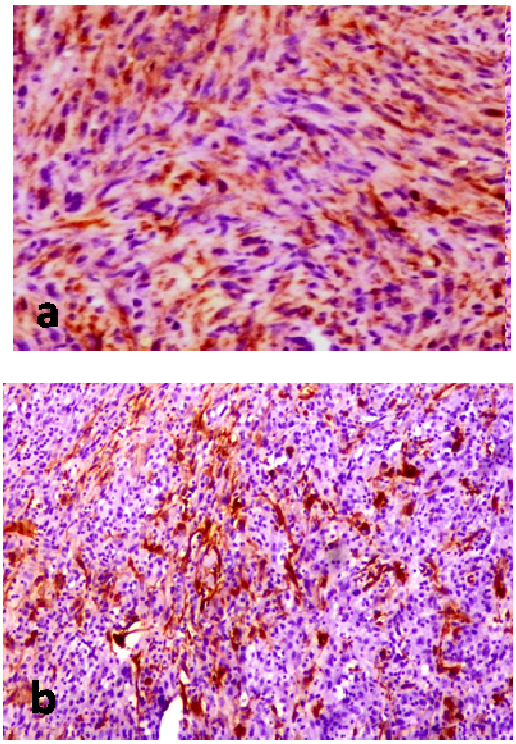

Figure 3. Immunohistochemical staining: Neoplastic cells showing positive immunostaining for (a) Vimentin (Vimentin, x400), (b) Smooth muscle actin (SMA, x400)

DISCUSSION

Sarcoma of the soft tissue is a rare condition, among which leiomyosarcoma is a rare malignant neoplasm of smooth muscle cell. PCL is an exceedingly uncommon malignant superficial soft tissue sarcoma (Auroy *et al*., 1999). PCL are uncommon soft tissue tumors which and are usually located on the extremities with a predilection for hair-bearing surfaces (Lin and Tsai, 1999; Weedon *et al*., 2006). Fifty to 75% of lesions occur on the legs, 20-30% on the arms and only 10-15% of lesions occur on the trunk (Lin and Tsai, 1999). The most common predisposing factors reported are trauma and radiation. Clinical appearance of PCL is non-specific with a wide range of differential diagnoses including squamous cell carcinoma, amelanotic melanoma, and basal cell carcinoma (6). Dermal leiomyosarcoma is usually seen as a solitary nodule ranging from 0.4 to 6 cm. Most case series described in the literature report a predilection for the middle aged to elderly male (Lin and Tsai, 1999; Auroy *et al*., 1999).

Fields and Helwig, presented a large case series of PCL in which 95% of the patients presented with a solitary nodule with a median size of 1.8 cm at presentation. Twenty-four percent of their patients presented with a painful mass (Fields *et al*., 1981). The histological features of leiomyosarcomas include highly cellular infiltrative tumor with predominantly fascicular pattern of growth. Tumor cells are highly atypical predominantly spindle shaped with blunt ended pleomorphic nuclei. Cells have eosinophilic cytoplasm with few showing perinuclear vauolations. Cutaneous lesions present a grey zone between them and overlying epidermis (Fauth *et al*., 2010).

Mitosis, equivalent to one or more per 10 high-power fields, high cellularity and bizarre myomatous cells are the generally accepted criteria for malignancy (Fields *et al*., 1981). Two growth patterns have been described by Kaddu *et al*, a nodular pattern characterized by high cellularity, prominent nuclear atypia, conspicuous mitosis and a diffuse pattern that is less cellular, well differentiated and inconspicuous mitosis (Kaddu *et al*., 1997). Classical immunophenotype of PCL comprises of positive vimentin, desmin and SMA staining. Focal positivity can occasionally be seen for Pan-muscle actin, HHF-35, CK, EMA, CD34 or S-100 (Auroy *et al*., 1999; Weiss and Goldblum, 2001). Electron microscopy can reveal intracytoplasmic myofilaments in cases which are difficult to diagnose. The most effective treatment of cutaneous leiomyosarcoma is wide excision with 3-5 cm lateral margin and a depth that includes subcutaneous tissue and fascia (Swanson *et al*., 1988). Several poor prognostic factorsof PCL have been identified by Jensen *et al*. (1996). These includes tumor size ≥5 cm, acral location, deep localization with fascia involvement and high histological grade. Cutaneous leiomyosarcomas have fewer incidences of local recurrence (30-50%) and negligible potential for distant metastases (0-10%). In contrast, subcutaneous leiomyosarcomas may develop local recurrence in about 40-60% and distant metastasis in 20-60% of patients (Fields *et al*., 1981). Hence long term follow-up should be emphasized on to keep in check any local recurrence or metastasis.

Conclusion

PCL is an uncommon entity that must be kept in the list of differential diagnosis of a malignant spindle cell neoplasm of the skin. An extensive histopathological examination, along with a proper immunohistochemical panel is mandatory for definitive diagnosis. Because of high recurrence rate, a long term follow-up of patient is recommended.

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