Primary cutaneous leiomyosarcoma: A rare malignant neoplasm

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ABSTRACT

Primary cutaneous leiomyosarcoma (PCL) is an exceedingly uncommon malignant superficial soft tissue sarcoma with a predilection for middle-aged to elderly male. Morphologic differential diagnosis includes a host of other malignant spindle cell neoplasms, thereby necessitating the use of a panel of immunohistochemical markers to arrive at a definitive diagnosis. We report a case of PCL arising in the right leg of a 70-year-old male.

Key words: Caldesmon, cutaneous, leiomyosarcoma

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INTRODUCTION

Superficial leiomyosarcomas are rare malignant tumors that account for only 5–10% of all soft tissue sarcomas.[1] They can be subdivided into two types according to its primary site of origin: deep subcutaneous and superficial cutaneous types.[2]

Subcutaneous leiomyosarcoma is by far the most common subtype. Primary cutaneous leiomyosarcoma (PCL) is a rare subtype with few isolated case reports and occasional case series described in the literature. PCL accounts for only 2-3% of all superficial soft tissue sarcomas.[2]

We report the clinical, histopathological and immunohistochemical findings in a 70-year-old male patient with PCL arising in the right leg.

CASE REPORT

A 70-year-old male patient presented with a painful nodule in the right leg, which had been progressively increasing in size for 4 months. On palpation, an irregular, firm, tender exophytic swelling, measuring 5 cm x 5 cm, with overlying skin showing ulceration was noted on the lateral aspect of the lower one third of the right leg [Figure 1A]. Inguinal lymph nodes were not palpable. His past history was significant of a trauma and subsequent non-healing ulcer formation at the same site in the right leg.

Systemic examination and investigations, including complete blood count, fasting blood sugar level, liver function test and renal function test, were within normal limits. Hepatitis B virus surface antigen and human immunodeficiency virus enzyme-linked immunosorbent assay were non-reactive.

A clinical diagnosis of squamous cell carcinoma was considered and fine needle aspiration cytology of the swelling was requested.

Fine needle aspiration cytology of the same was reported as a spindle cell neoplasm with suspicion of malignancy. The swelling was excised with a 5 cm resection margin and submitted for histopathological examination [Figure 1B].

Gross inspection revealed a grey brown, irregular, nodular soft tissue mass, measuring 5 cm x 5 cm x 2.5 cm and partly covered with skin. The cut surface of the mass was greyish white, solid, homogenous and fleshy [Figure 1C].

Light microscopy revealed a poorly delineated dermal neoplasm extending into the underlying subcutis. The neoplasm was composed of bundles of elongated spindle cells arranged in interlacing fascicles, with intensely pink, fibrillary cytoplasm and pleomorphic nuclei with coarse irregularly dispersed chromatin. Mitosis, including atypical, amounting to 22 mitotic figures per 10 high power fields and multinucleate tumor giant

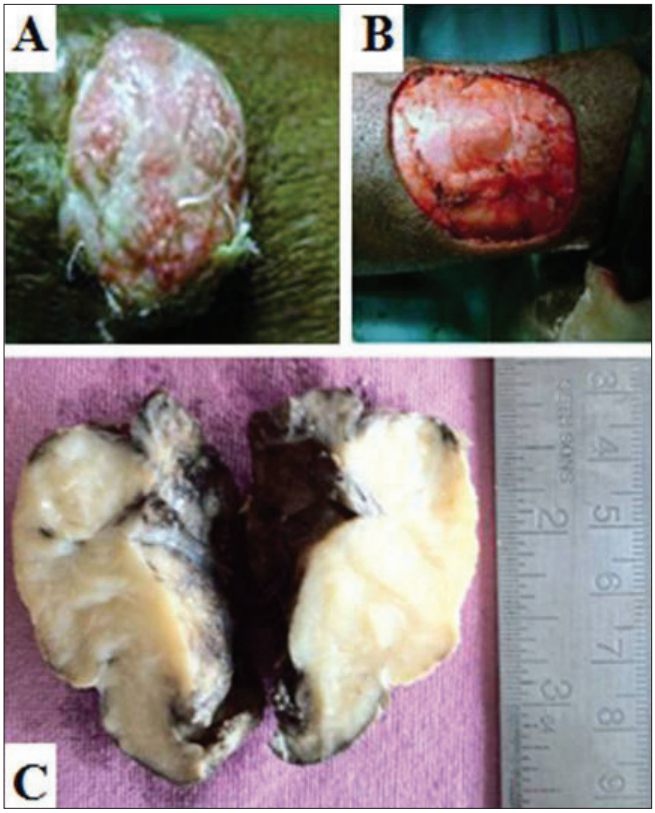

Figure 1: (A) Clinical photograph showing an irregular, exophytic swelling, measuring 5 cm x 5 cm, with overlying skin showing ulceration on the lateral aspect of the lower one-third of the right leg. (B) Clinical photograph of the post-tumor excision site. (C) Gross photograph showing greyish white, solid, homogenous and fleshy cut surface of the irregular tumor mass measuring 5 cm x 5 cm x 2.5 cm

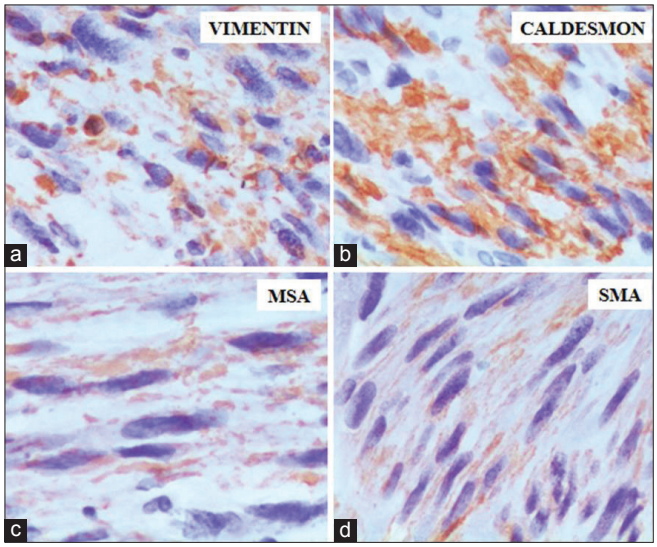

Figure 3: Immunohistochemical staining: Neoplastic cells showing positive immunostaining for (a) Vimentin (Vimentin, x400), (b) Caldesmon (Caldesmon, x400), (c) muscle-specific actin (MSA, x400) and (d) smooth muscle actin (SMA, x400)

cells, was evident. Focal necrosis was noted at the edges. The overlying epidermis was thinned out with areas of ulceration [Figure 2].

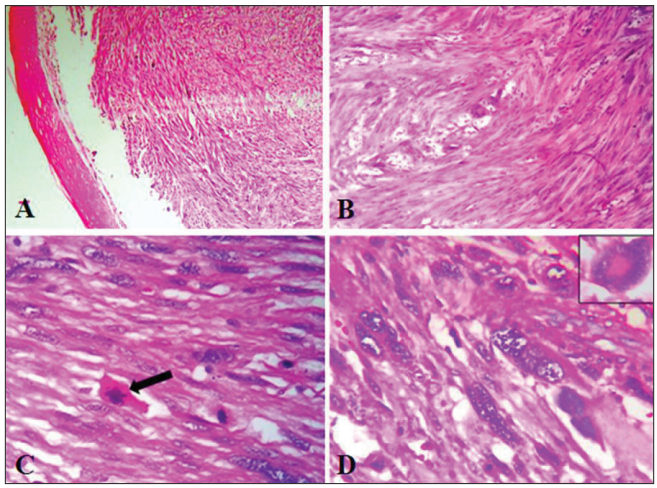

Figure 2: (A) Light microscopy revealed a dermal neoplasm with extension into the subcutaneous tissue (H & E, x40). (B) Neoplasm composed of interlacing fascicles of elongated spindle cells (H & E, x100). (C) Tumor cells are spindle shaped with intensely pink, fibrillary cytoplasm and pleomorphic hyperchromatic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

A provisional diagnosis of spindle cell malignant neoplasm of the skin, which typically includes a host of diagnostic possibility, namely fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, monophasic synovial sarcoma, malignant fibrous histiocytoma and spindle cell variant of squamous cell carcinoma, was considered and immunohistochemistry was advised for confirmation.

On immunohistochemistry, tumor cells showed strong immunopositivity for vimentin, muscle-specific actin (MSA), smooth muscle actin (SMA), caldesmon and desmin and negative for pancytokeratin, Epithelial Membrane Antigen (EMA), myogenin, CD34 and S-100 protein [Figure 3].

Based on the immunophenotypic features, a final diagnosis of PCL was rendered. The patient is currently on periodic follow-up since 1 year and no recurrence or metastasis has been identified so far.

DISCUSSION

PCL is an exceedingly uncommon malignant superficial soft tissue sarcoma.[3] It is postulated to arise from the arrector pili muscles. The most common predisposing factors reported are trauma and radiation. Malignant transformation of a leiomyoma has also been reported.[4] A history of possible trauma was reported by our patient as well.

Although PCL most commonly arises in the extremities, particularly hair bearing surfaces, it may occur at any anatomic

site on the body.[3,4] Most case series described in the literature report a predilection for the middle aged to elderly male.[4,5]

In a large case series reported by Fields and Helwig,[4] 95% of the patients with PCL presented with a solitary nodule with a median size of 1.8 cm at presentation. Pain was reported by 24% of their patients at presentation, whereas pain could be elicited on pressure in an additional 27% of the patients.

Histologically, PCL is characterized by a poorly circumscribed proliferation of interwoven fascicles of spindle shaped atypical myomatous cells that merge with a collagenous stroma.[4,6] Mitosis, equivalent to one or more per 10 high-power fields, high cellularity and bizarre myomatous cells are the generally accepted criteria for malignancy.[4] Kaddu *et al*.[6] described two growth patterns: 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. Morphologic differential diagnosis includes a galaxy of other malignant spindle cell neoplasms, namely desmoplastic malignant melanoma, spindle cell synovial sarcoma, spindle cell angiosarcoma, fibrosarcoma, malignant fibrous histiocytoma and malignant peripheral nerve sheath tumor. Immunohistochemistry therefore is instrumental in arriving at a definitive diagnosis.[1]

Cutaneous leiomyosarcoma often exhibit immunophenotypic polymorphism, thereby mandating the use of a large panel of antibodies. Desmin, SMA and caldesmon staining are positive in a vast majority of cases. However, none of these markers is absolutely specific for smooth muscle, and positivity for two of these markers is more supportive of leiomyosarcoma. Pan-muscle actin HHF-35 immunostaining is sometimes focally present. Focal keratin, EMA, CD34 or S-100 positive areas may occasionally be encountered.[1,3]

Jensen *et al*.[7] identified several poor prognostic factors, namely 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.[4]

A wide local excision with a 3–5 cm margin is usually an effective treatment for PCL.[2,4] The role of adjuvant therapy is controversial.[2]

In conclusion, PCL is a rare entity that must be borne in mind when encountered with a malignant spindle cell neoplasm of the skin and usually requires a panel of immunohistochemical markers to distinguish it from other cutaneous malignancies with spindle cell morphology.

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