Primary cutaneous leiomyosarcoma of the flank: Case report and review

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Abstract

Cutaneous leiomyosarcoma is a rare soft tissue sarcoma. We present a case of a 50-year-old man, with a rare location in the flank. Clinical presentation showed an ulcerative lesion. Imaging features were evocative of malignancy. Surgical excision was performed, involving oblique muscles. The diagnosis of low-grade leiomyosarcoma with free margins was made. No adjuvant therapy was indicated. Clinical and radiological follow-up of the patient during two years did not reveal recurrences. Cutaneous leiomyosarcoma of the flank is extremely rare and takes part in a group of infrequent and unknown tumors. Clinical presentation is unspecific and histological evidence is mandatory for diagnosis. Tumor extension in depth constitutes the major determinant factor for therapeutic management and prognosis. Wide surgical resection is crucial to minimize recurrence risk especially when poor prognostic factors take place.

Keywords: Smooth muscle; Leiomyosarcoma; Cutaneous; Flank; Surgical excision

1. Introduction

Leiomyosarcomas (LMS) are a very rare smooth muscle malignant tumors. They represent 3–7% of superficial soft tissue sarcomas (Gustafson et al., 1992). Superficial leiomyosarcomas (SLMS) are divided into two groups. Cutaneous LMS arise in the dermis with possible extension into the underlying tissues. Subcutaneous LMS originate from the subcutis and have more aggressive behavior (Guillen and Cockerell, 2001). SLMS may arise anywhere on the body with predilection for the lower extremities.

Due to their infrequency and unspecific clinical presentation, the diagnosis of LMS is usually delayed (APol et al., 2012). Tumor localization and its expansion to the adjacent tissues may limit surgical excision and determine mainly the prognosis. We present a rare case of cutaneous leiomyosarcoma that occurred in the flank.

2. Case report

A 50-year-old man, smoker, without medical history, presented with an ulcerative lesion of the right flank discovered 4 months ago. On clinical examination, there was a 5 × 4 cm swollen and indurate lesion with ulcerated center. Ultrasonography showed a dermal mass measuring 5 × 4 × 3 cm, with hypoechoic and heterogeneous echostructure, and poorly vascularized on Doppler. On computed tomography, the lesion was hyperdense, ill-defined and coming in contact with the right oblique muscles. Tumor displayed enhancement and there were no signs of...
intra-abdominal extension (Fig. 1). Biopsy revealed dermic mesenchymatous proliferation consisting of fascicles of spindle cells with atypical and hyperchromatic nucleus within a fibrous stroma suggesting the diagnosis of sarcoma. Surgical excision, involving the oblique muscles, was made. Surgical wound was closed by placement of rotation flap. The remainder of patient’s course was uneventful. Gross pathological examination revealed a white tumor of $6 \times 5 \times 3$ cm, ulcerated on surface, with hard consistency and ill-defined limits (Fig. 2). Histological examination showed a well differentiated sarcomatous proliferation made of spindle-shaped cells provided with cigar-shaped nuclei (Fig. 3). There was nuclear atypia and the mitotic activity was moderate (9 mitoses per 10 high-power fields). There were large foci of necrosis (20%). Surgical margins were free. Immunohistochemically, spindle cells were positive for actin and negative for desmin, caldesmone and PS-100 (Fig. 3). The diagnosis of leiomyosarcoma grade 1 according to the “Fédération Française des Centres de Lutte Contre le Cancer” – FNCLCC grading system was made. No adjuvant therapy was indicated. The patient was followed-up biannually based on clinical exam, soft tissue and abdominal ultrasonography and chest radiograph. Monitoring during two years did not reveal local recurrence or metastasis.
3. Discussion

SLMS occur usually in elderly individuals with male predilection (Guillen and Cockerell, 2001). They are located most of the time on the lower extremities (50–70%). Upper extremities localizations are less frequent (20–30%) and the trunk ones are rare (10–15%) (Washer and Lee, 1992). Reviewing the literature, LMS of the trunk occur mainly in the back and the buttocks. Flank locations are especially rare and only one case of anterior abdominal wall tumor has been reported. Moreover, unusual sites have been reported involving the face, penis, breast, orbit and external auditory canal (Fields and Helwig, 1981). Etiology of cutaneous LMS is unknown. Mainly predisposing factors are presented by traumatic scars, radiotherapy and leiomyomas (Tellechea et al., 1993). Our patient had no history of physical trauma or precancerous lesions. SLMS arise from the arrector pili muscles of the hair and sweat glands in the cutaneous subtype and from the smooth muscle of vessels in the subcutaneous one (Skoulakis et al., 2010). Histological distinction between these two groups, particularly essential because of its therapeutic and prognosis implications, is difficult to make because of possible extension of each tumor to both skin layers (Massi et al., 2010). Cutaneous LMS don’t have a specific clinical presentation. The usual appearance reported is a solitary nodule, frequently with red color and ranging from 5 to 60 mm (Kuflik et al., 2003). Its surface can be smooth, ulcerated as in our case, rough, verrucous and sometimes indurated and hemorrhagic. Histologically, tumor is composed of a highly cellular proliferation organized in fascicles of spindle-shaped cells with irregular arrangement of cell bundles. Cells have a stretched and round ended nucleus giving a “cigar” appearance (Fig. 3). The degree of differentiation may vary within a single tumor. Mitotic figures are seen all over the lesion (Massi et al., 2010; Limaïem et al., 2007). Criteria for malignancy include the presence of mitoses of at least one per 10 high power fields, high cellularity, significant nuclear atypia and tumor giant cells (Limaïem et al., 2007). Histological variants of cutaneous LMS can lead to diagnosis difficulties as in the case for epithelioid, granular cell, desmoplastic, inflammatory and myxoid LMS (Limaïem et al., 2007). Diagnosis is also frequently difficult to establish based on biopsy because of fragments small size and multiple sections on resection specimen is mandatory (Jensen et al., 1996).

Immunohistochemical studies help in the diagnosis of poorly differentiated forms. Classical immune-phenotyping consists of positive staining for vimentin, desmin and smooth muscle actin (Massi et al., 2010; Limaïem et al., 2007). Staining in our case was positive for actin but negative for desmin. Actin looks to be more sensitive than desmin for smooth muscle tumors, even though the antibody is not always specific (Massi et al., 2010; Limaïem et al., 2007). In a large review concerning soft tissue tumors, transgelin, a new myogenic marker, was found to be the best marker in LMS (Robin et al., 2012). Treatment of cutaneous LMS consists of wide resection with 3–5 cm lateral margin and a depth that includes subcutaneous tissue and fascia in order to avoid recurrences (Deneve et al., 2013). Mohs micrographic technique may be useful to ensure complete tumor removal while saving surrounding tissues (Lane and Kent, 2005). Adjuvant therapies have not been standardized. LMS are known by their resistance to both radio- and chemotherapy (Limaïem et al., 2007). Their use is reserved in case of LMS with large extensions limiting complete excision and cases of recurrence or metastatic diseases (Deneve et al., 2013). Follow-up is based on clinical examination and CT scan. Local recurrence and distant metastasis can occur years after the initial excision (Limaïem et al., 2007). Cutaneous LMS have a better prognosis compared with the subcutaneous subtype with less local recurrence rates (30–50% versus up to 70%) and uncommon metastasis (Pashaei et al., 2005). Principal poor prognostic factors include tumor size ≥ 5 cm, deep location with fascia involvement and high malignancy grade (APol et al., 2012).

4. Conclusions

Cutaneous leiomyosarcoma of the flank is extremely rare and takes part in a group of infrequent and unknown tumors. Clinical presentation is unspecific and histological evidence is mandatory for diagnosis. Tumor extension in depth constitutes the major determinant factor for therapeutic management and prognosis. Wide surgical resection is crucial to minimize recurrence risk especially when poor prognostic factors take place.

Conflict of interest

We have no conflict of interest to declare.

References


