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Primary cutaneous leiomyosarcoma of the face

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ABSTRACT

Superficial leiomyosarcoma, a rare malignant lesion, constitutes only 3%-5% of all soft tissue sarcomas. Formerly viewed as a single entity, cutaneous and subcutaneous leiomyosarcomas are now believed to be distinct diseases, with different biological behaviour. Cutaneous leiomyosarcomas derive from the erector pili muscles, smooth muscles that surround sweat glands or from vascular tissue. They occur predominantly on the lower extremities, with predilection for the hair-bearing extensor surface. They occur in the face in only 1%-5% of cases.

We report a rare case of cutaneous leiomyosarcoma on the face of an 82-year-old woman. The tumour was fully excised and exhibited a number of histological and immunohistochemical features that are characteristic of cutaneous leiomyosarcoma. The immunohistochemical study revealed positivity to vimentin, h-Caldesmon and smooth muscle actin. No staining was noted with antibodies against S-100, HMB45, wide spectrum cytokeratin and epithelial membrane antigen. Six-month follow-up revealed a well-healed scar without evidence of local recurrence and regional or distant metastases.

Leiomyosarcomas (LMS) are malignant non-epithelial tumours. They derive from smooth muscle cells of vessels, predominantly in visceral locations, such as the uterus, the gastrointestinal tract, the mesentery, the urogenital system or the retroperitoneal space. Superficial LMS, a rare malignant lesion, constitutes 4.0%–6.5% of all soft tissue sarcomas, an overall incidence of approximately 0.04% among all cancers[1,2]. Formerly viewed as a single entity (superficial LMS), cutaneous and subcutaneous LMS are now believed to be distinct diseases, with dramatically different potentials for local recurrence and metastases[3,4]. Although cutaneous LMS is associated with 30% to 50% local recurrence rate, a well-documented case of metastases has not been reported. Comparing with lesions from cutaneous structures, subcutaneous LMS is associated with a higher local recurrence rate (50% to 70%), metastases (30% to 40%) and death[1,5].

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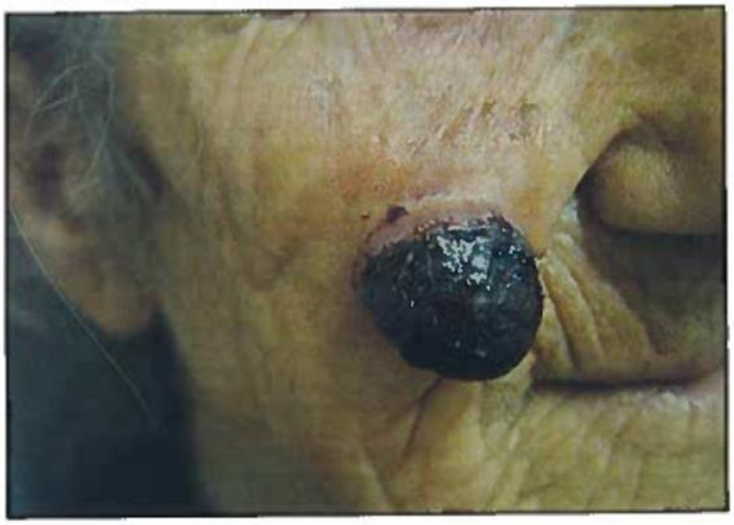

Fig. 1 - Clinical aspect of the tumour.

We describe the histological and immunohistochemical features of a cutaneous LMS in an elderly woman, treated with local excision.

CASE REPORT

An 82-year-old woman presented a rapidly enlarging nodule on her right cheek, which had grown over the last 2 months (Fig. 1). The patient did not recall any trauma in that region before the development of the tumour and denied exposure to ionizing radiation. The physical examination showed a solitary, elevated and well defined black nodule measuring 2.5cmx1.7cm with no adenopathies or additional cutaneous lesions.

The routine laboratory findings were normal. The thoracic, abdominal and pelvic CT scans were normal.

The lesion was excised with 1cm of surgical margin. The histological examination revealed cutaneous malignant tumour composed by a proliferation of atypical spindle-shaped cells arranged in sheets and interlacing fascicles with blunt-edge nuclei, clumping of chromatin and prominent nucleoli and numerous mitotic figures under an ulcerated epidermis (Fig. 2). The immunohistochemical study was positive with antibodies against vimentin, smooth muscle alpha-actin and h-Caldesmon (Fig. 3). No staining was noted with antibodies against S-100, HMB 45, Melan A, MiTF, epithelial membrane antigen (EMA) and a wide spectrum cytokeratin.

Six-month follow-up revealed a well-healed scar (Fig. 4) without evidence of local

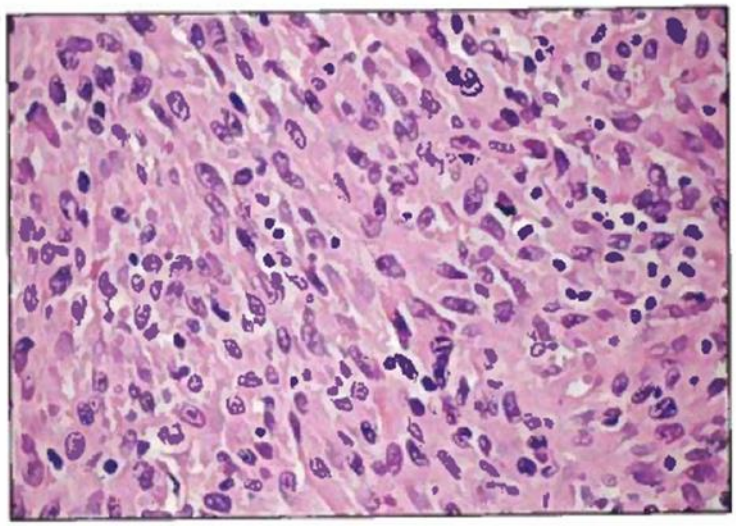

Fig. 2 - Cutaneous leiomyosarcoma composed of spindle-shaped cells with nuclear atypia and cytological pleomorphism (H&E,X400).

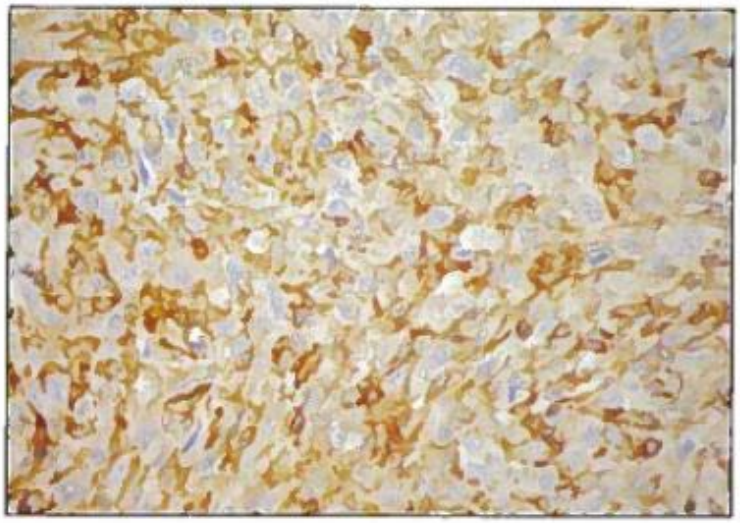

Fig. 3 - Focal staining of h-Caldesmon (X400).

recurrence and regional or distant metastases.

COMMENT

Although histologically similar, three different types of LMS are usually recognized, based on clinical and biological differences: (i) retroperitoneal and abdominal LMS; (ii) cutaneous and subcutaneous LMS and (iii) vascular LMS. Cutaneous LMS are derived from the erector pili muscles, smooth muscles that surround sweat glands in the nearby adipose tissue at or below the junction of the cutis and subcutis or from vascular tissue[2,5], whereas the subcutaneous variety derive from the smooth cell muscle of the blood vessels[4].

Primary cutaneous superficial LMS are very rare, accounting for only to 2% to 3% of all superficial soft-tissue sarcomas[2,6]. Clinically, superficial LMS typically occur primarily as solitary nodules[2].

Multicentricity in the skin with synchronicity almost always indicates metastasis from an underlying soft tissue or visceral malignancy[2,7]. Morphologic characteristics include irregular contours, pedunculation, umbilication and discoloration of the skin (e.g., pink, red, violet, tan, brown, black or grey). Ulceration is uncommon[2,5]. Pain is the most common symptom, occurring in 80% to 95% of patients[6]. Pruritus, burning and bleeding are also common[6].

Leiomyosarcoma is primarily a disease of middle age (50-70 years), although occasionally it occurs in young and old age[8,9]. There is no sex preference[5,9]. Cutaneous LMS occurs more frequently along the

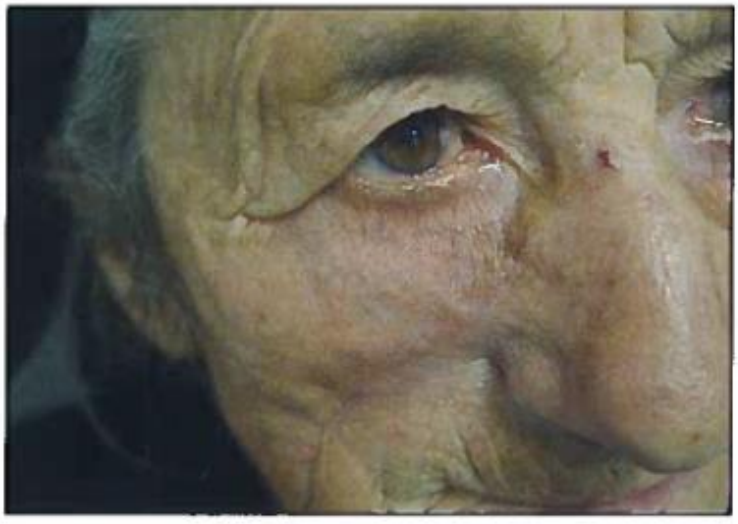

Fig. 4 - Well-healed scar, 6 months after surgery.

extensor surfaces of the extremities, consistent with areas of greatest hair distribution[1,2,6]. Approxi-mately 50%-70% occur on the lower extremities, most often proximally, 20%-30% occur on the upper extremities and 10%-15% on the trunk[6]. It occurs in the face in only 1%-5% of cases[6].

Superficial LMS can be divided into two distinct entities: cutaneous LMS and subcutaneous LMS. The histopathogenesis and overall clinical prognosis appear to be different between these two types. The subcutaneous LMS tend to be biologically more aggressive than their cutaneous counterparts. They exhibit a higher growth rate and the potential for recurrence and metastases is greater than for the cutaneous lesions. There is local recurrence in 50%-70% of patients, and metastases occur in 30%-40%, commonly affecting the lungs, liver and bones[2]. Ultimately, 30%-40% of theses patients die of their disease[1,2]. By contrast with the highly malignant behaviour of subcutaneous LMS, cutaneous LMS is a relatively benign disease. These lesions are usually contained within the dermis, although larger lesions may extend into the adjacent subcutis and epidermis[10].

We were unable to find a single well-documented report of metastatic cutaneous LMS and it is generally accepted that this lesion has minimal, if any, metastatic potential[2]. Local recurrence, however, is common and is seen in 30%-50% of patients[2]. No correlation of local recurrence rates with either tumour size or histopathological features was found; inadequate excision is, presumably, the predisposing risk factor[6]. Tumour related deaths have not been previously described [2,11].

Histological characteristics of LMS alone may occasionally lead to confusion with other cutaneous spindle-shaped tumours. They display a fairly homogeneous pattern of spindle cell proliferation arranged in interlacing cords[12]. The histopathological diagnosis requires an appearance of dense cellularity, nuclear atypia and pleomorphism, with at least one mitotic figure per 5 or 10 high power fields[2].

Cutaneous LMS may show different immunophenotypes, thus emphasizing the importance of using a large panel of antibodies: smooth muscle actin, desmin, vimentin, h-Caldesmon, cytokeratins, HMB45 and S-100 protein. Co-expression of vimentin, desmin and muscle-specific actin represents the characteristic phenotype of LMS[9]. Actin stained positively in 100% and desmin in 66% of cutaneous lesions[13,14]. h-Caldesmon (h-CD) is a protein combined with actin and tropomyosin that regulates cellular contraction, and has been thought to be expressed exclusively in vascular and visceral smooth muscle cells and LMS showed intense and extensive immunoreactivity for h-CD[15]. Absence of desmin expression has been observed in poorly differentiated neoplasms and may represent a feature of neoplasm that arises from vessel walls[14].

The surgical management of cutaneous LMS is contradictory in the literature. Because of its rarity, published reports have all been retrospective series and these have concentrated on diagnosis rather than management. The tumour is not sensitive to X-ray therapy and is not suitable for chemotherapy. Thus, wide local excision is the treatment of choice. Surgical management has varied from conservative excision, without

reference to margin dimension to 3 cm to 5 cm excision margin including subcutaneous fat and deep fascia[16].

The rate of recurrence has been claimed to decrease with wider excision but there are no documented figures for these claims[2,6,16]. Complete surgical excision is recommended, but it would seem that narrow margins should be sufficient[4]. However, all patients should be followed for a minimum of 5 years after excisional surgery[4]. If microscopic margins are negative for soft-tissue sarcomas, then local recurrence and survival are not affected by increasing the soft tissue excisional margin[17]. The reports of high local recurrence may represent high rates of incomplete surgical excision.

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