CASE REPORT

Delayed diagnosis of dermal leiomyosarcoma mimicking keloid scar

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SUMMARY

A 43-year-old man developed an abnormal scar 6 months following excision of a leiomyoma from his left shoulder. The scar was elevated, irregular in shape, pink-red in colour, hard in consistency and it was extending beyond the margins of the original wound. A diagnosis of a keloid scar was considered and the patient was managed as such. He underwent a planned procedure for intralesional excision of the keloid scar. The histopathological examination showed a diagnosis of leiomyosarcoma. This case report presents a delayed diagnosis of dermal leiomyosarcoma mimicking a keloid scar. The patient subsequently underwent wider excision of the tumour with curative intention.

BACKGROUND

Cutaneous leiomyosarcoma is a rare type of tumours representing 1%–3% of all sarcomas.¹ The aetiology of leiomyosarcoma is unknown. It may arise from leiomyomas, trauma and radiation.^{2,3} It tends to be the most common in middle-aged men.^{1,3} Early diagnosis is important for successful management and curative intention. The risk of metastasis

and recurrence is 3.3% and 24.4%, respectively.⁴ Delayed diagnosis may impact the outcome. It can be due to a variety of reasons including lesions with misleading features, neglected lumps and inaccurate diagnosis.^{5,6} We present a case of delayed diagnosis of dermal leiomyosarcoma mimicking a keloid scar in a patient with a history of leiomyoma excised from the same site. Having a high index of suspicion and routine histopathological examination of specimens is the key for successful management.

CASE PRESENTATION

A 43-year-old man was referred to the plastic surgery unit for the management of abnormal scar on the right shoulder, at the site where a leiomyoma was excised by curettage 2 years prior to presentation. The lesion was slowly growing in size and became more painful with time. It became infected few times and antibiotic therapy was commenced. The medical history included left eye glaucoma for which he was prescribed eye drops. He had no other regular medications. He had no known allergies and was an ex-smoker. On examination, the lump was raised, firm, pink-red in colour and extending outside the borders of original surgery (figures 1–4). He was reviewed by two plastic surgery consultants who advised intralesional excision of the lesion followed by steroid injections.

INVESTIGATIONS

The initial curettage specimen consisted of four pieces of tissue, the largest measuring 23×10×9 mm. Histological examination showed that expansion of the dermis by a smooth muscle proliferation comprised bland spindle cells arranged in short fascicles, set among dermal collagen fibres. The proliferation lacked cytologic atypia or mitotic activity and there was no infiltration of the subcutaneous tissues. Immunohistochemical analysis showed diffuse positive staining with desmin and smooth muscle actin, and negative staining for S100, CD31 and CD34. Overall, the appearances were felt to be those of a dermal leiomyoma. The completeness of excision could not be assessed due to the fragmented nature of the specimen.

The intralesional excision of what was felt to be a keloid scar consisted of a specimen measuring 55×46×17 mm with overlying skin. On sectioning, the tissue consisted almost entirely of tumour which was firm, homogeneous and tan-white. Histological examination showed a grade 1 leiomyosarcoma expanding the dermis consisting of moderate

Figure 1 A tangential view showing an irregular lobulated pink lesion on the left shoulder.

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Figure 2 A view from above showing the lesion on the left shoulder.

atypical spindle cells arranged in fascicles. There was focal infiltration of subcutaneous tissues. Mitoses were easily identified (8 per 10 high-power fields) but there was no necrosis. There was widespread involvement of the deep and peripheral margins. Vascular invasion was not identified. Immunohistochemical analysis showed diffuse positive staining with desmin, smooth muscle actin and h-caldesmon. There was negative staining for S100, CD31 and CD34. This immunoprofile is consistent with a tumour of smooth muscle lineage (Figure 5).

Figure 3 A view from above showing cutaneous leiomyosarcoma on the left shoulder.

Figure 4 A view taken from the front of the left shoulder showing an elevated irregular lesion.

The subsequent wider excision specimen comprised a skin ellipse measuring 61×22 mm with a 16 mm subcutaneous tissue. There was a single 2 mm diameter nodule of residual tumour in the dermis with a surrounding scar and giant cell reaction. The residual tumour nodule was fully excised and clear of the nearest margin by 6 mm.

OUTCOME AND FOLLOW-UP

The patient subsequently underwent wider excision with curative intention. The tumour was excised with 6 mm peripheral margins and 10 mm deep margins. Subsequent discussion at the sarcoma multidisciplinary team advised clinical follow-up to monitor for recurrence of leiomyosarcoma and a chest radiograph. The chest radiograph did not identify significant lung lesions.

DISCUSSION

Leiomyosarcoma is a malignant tumour of pure smooth muscle lineage. The most common soft tissue sites are retroperitoneum,

Figure 5 (A) Dermal leiomyoma comprised cytologically bland spindle cells arranged in short fascicles, set among dermal collagen fibres. The proliferation lacks atypia or mitotic activity. (B) Dermal leiomyosarcoma consisting of moderately atypical spindle cells arranged in fascicles with easily identified mitoses. (C) Diffuse positivity with desmin. (D) Diffuse positivity with h-caldesmon.

blood vessels (especially the inferior vena cava and the veins of the lower extremities) and non-retroperitoneal locations, in particular the soft tissue of the lower extremities. Leiomyosarcoma also arise in the uterus and the gastrointestinal tract and do so with much greater frequency than soft tissue.⁷ Leiomyosarcoma of skin and subcutaneous tissue is a special entity comprising 1%–3% of all sarcomas.¹

The aetiology is unknown. Leiomyosarcomas usually have complex karyotypes, along with genomic instability. Alterations in the retinoblastoma-cyclin D1 and the TP53 pathways are frequently observed; they arise with greater frequency in patients with Li-Fraumeni syndrome. Trauma and radiation exposure have also been proposed as predisposing factors. Malignant transformation of a leiomyoma is felt to be a very rare occurrence.²

The clinical presentation of superficial soft tissue leiomyosarcoma is of a painful cutaneous or subcutaneous mass. It can occur at any anatomical location, however, it is more common in hair-bearing areas of the body.^{5,8}

Other clinical differential diagnoses include granulomatous lesions (eg, cutaneous tuberculosis and sarcoidosis), benign lesions (eg, fibroma and neurofibroma) and other malignant cutaneous lesions, dermatofibrosarcoma protuberans and Kaposi sarcoma.⁶

Leiomyosarcoma should be managed in a multidisciplinary team setting.⁹ Dermal leiomyosarcomas have an excellent prognosis; although they can locally recur, if the tumour is completely confined to the dermis, there is almost no risk of distant metastasis. As such, some authors feel that a diagnosis of 'sarcoma' in these cases is non-appropriate and the alternative designation of 'atypical intradermal smooth muscle neoplasm' has been proposed.¹⁰ Surgical treatment is with wide local excision.

A study by Aneiros-Fernandez *et al* reported 313 patients diagnosed with cutaneous leiomyosarcoma. In this study, the risk of recurrence, metastasis and mortality is 24.4%, 4.22% and 3.33%, respectively, after a median of 3.4 years of follow-up. In subcutaneous lesions, the risks of local recurrences, metastasis and mortality are much higher: 36.63%, 43.23% and 37.82%, respectively, after a median follow-up period of 4.4 years.⁴

Delayed diagnosis of leiomyosarcoma was reported in the literature due to a variety of reasons, including unusual site and clinical features of tumour and neglected lesions by elderly patients.⁵ However, few cases report delayed diagnosis of sarcomas mimicking a keloidal lesion.

A similar case, from Brazil, was published in 2015, reporting leiomyosarcoma in a lesion diagnosed as a keloid scar on the lateral aspect of the lower limb. The lesion was treated as a keloid scar until referral was made to a plastic surgeon for reassessment. However, the author failed to state clearly whether there was history of trauma and scar reported prior to the onset of lesion.¹¹

A second case reported leiomyosarcoma in a buttock lesion diagnosed as a keloid scar following excision of a histiocytoma.¹²

A third case reported high-grade soft-tissue sarcoma, fibrosarcomatous dermatofibrosarcoma protuberans (FS-DFSP), in

Learning points

- ▶ High index of suspicions is warranted for suspicious skin lesions resembling keloid scars following excision of leiomyomas.
- ▶ Previous surgery and scar do not exclude other differential diagnoses of a keloid scar.
- ▶ Histopathological study should always be requested for specimens obtained from 'keloid scars'.

a patient with a lesion on the anterior chest wall treated as a keloid scar. Unlike our patient, there was no history of trauma or scar before the onset of the lesion.¹³

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