On-line Case Report

Angiosarcoma in an area of lipodermatosclerosis

ANDREW JL JOWETT, SIMON D PARVIN

Department of Vascular Surgery, Royal Bournemouth Hospital, Bournemouth, UK

We present an unusual case in a post phlebotic lower limb that had previously developed an area of lipodermatosclerosis. An area within this broke down as a result of an angiosarcoma rather than the more probable: formation of a venous ulcer.

Keywords: Angiosarcoma – Lipodermatosclerosis

Angiosarcoma is a rare aggressive mesenchymal tumour. It is more commonly described in upper limbs affected by post mastectomy lymphoedema (Stewart-Treves syndrome).¹ Occurrence in oedematous lower limbs has been reported.²⁻⁴

Venous ulcers are known to have the potential to undergo malignant change in the form of squamous cell carcinoma. We present an unusual case in a post phlebotic lower limb that had previously developed an area of lipodermatosclerosis. An area within this broke down as a result of an angiosarcoma rather than the more probable: formation of a venous ulcer.

Case report

A 59-year-old woman presented with a painful lesion on the medial aspect of her left ankle. The lesion was situated within an area of lipodermatosclerosis at its lower end. The venous changes developed as a result of two deep venous thromboses; the first occurring 13 years previously following a hysterectomy and bilateral salping-oophorectomy (for endometriosis) and the second spontaneously 12 years later. The limb had become intermittently swollen in the year prior to presentation. An area within the lipodermatosclerosis had become nodular, begun to breakdown and weep (Fig. 1). There was no history or specific risk factors for malignancy or immune compromise.

On examination, a classical area of lipodermatosclerosis was seen on her lower leg with a raised nodular purple area within it measuring 12 cm x 10 cm, just above the medial malleolus, with several small areas of break down. The leg was mildly oedematous and pulses were normal.

A venogram showed incomplete filling of the calf veins, with little recanalisation of the common iliac vein and superficial femoral vein as well as recent thrombosis of the full length of the long saphenous vein. The blood was thus returning via the short saphenous vein and a single thigh tributary and crossing to the right side of the pelvis.

Figure 1 The area of lipodermatosclerosis and lesion as described.

Correspondence to: Simon D Parvin, Consultant Vascular Surgeon, Royal Bournemouth Hospital, Castle Lane East, Bournemouth, Dorset BH7 7DW, UK. E: simon.parvin@rbch.nhs.uk

An excision biopsy was performed. Histology revealed the skin surrounding the lesion to show changes compatible with vascular stasis with a proliferation of capillary sized vessels. There was a proliferation of malignant vascular channels in the subcutaneous fat and deep dermis with surrounding areas of necrosis and marked endothelial atypia. These findings together with multilayering, characteristically dissecting growth pattern and positive CD31 staining confirmed the diagnosis of cutaneous angiosarcoma.

In view of the histology result, a CT scan of the abdomen and pelvis together with MRI scan of the thigh were performed, failing to show any metastases. An X-ray of the lower leg revealed destruction of the tibial cortex, implying deep extension; as a result, above-knee amputation was performed. At 1-year follow-up, there was no evidence of recurrence in the groin, stump or distant sites.

Discussion

Angiosarcoma is a rare mesenchymal tumour accounting for about 1% of sarcomas. The usual sites of presentation are scalp, lymphoedematous extremities, breast and previously irradiated fields.² It is most common where first described, in limbs oedematous post mastectomy as cited by Stewart and Treves in 1948.¹

Angiosarcoma has been reported in limbs oedematous for other reasons (*e.g.* post deep vein thrombosis^{2,4} and immobility); however, we are not aware of its description within an area of lipodermatosclerosis *per se*. In non-post mastectomy limbs, it tends to occur after a prolonged period of oedema (*e.g.* 20 years).³

The macroscopic changes in the above case were typical: red-purple nodules that then ulcerated.⁴ These changes may be difficult to distinguish from an area of lipodermatosclerosis breaking down to form a venous ulcer.

Angiosarcoma developing in a limb oedematous for reasons other than post mastectomy is thought to be predisposed to by the propensity for proliferation of

capillaries. This is thought to occur as a result of the activation of fibroblasts leading to increased collagen production.³ Lipodermatosclerosis itself is also known to involve the formation of an enlarged capillary bed as a result of venous hypertension. It has been hypothesised that lymphoedema predisposes to this sort of tumour as it decreases local immunity allowing the proliferation of abnormal cells.^{3,4}

Treatment of angiosarcoma in a limb is debatable: isolated limb perfusion, amputation or both and radiotherapy or chemotherapy have been used, with no clear benefit of any one modality.^{2,5} Current reported survival is in the region of 20 months;² this may potentially be improved by early diagnosis permitting treatment before the development of micrometastases or by advances in chemotherapeutic agents.

Conclusions

We believe that the diagnosis of angiosarcoma should be considered if nodular areas appear in a region of lipodermatosclerosis in an oedematous limb and subsequently begin to break down. Further research is required to elucidate the appropriate treatment, however.

References

1. Stewart FW, Treves N. Classics in oncology: lymphangiosarcoma in postmastectomy lymphedema: a report of six cases in elephantiasis chirurgica. *CA Cancer J Clin* 1981; **31**: 284–99.
2. Maddox JC, Evans HL. Angiosarcoma of skin and soft tissue: a study of forty-four cases. *Cancer* 1981; **48**: 1907–21.
3. Grevelink JM, Rosenberg AE. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 18-1993 – A 57-year-old man with chronic lymphedema and enlarging purple cutaneous nodules of the leg. *N Engl J Med* 1993; **328**: 1337–43.
4. Schmitz-Rixen T, Horsch S, Arnold G, Peters PE. Angiosarcoma in primary lymphedema of the lower extremity – Stewart–Treves syndrome. *Lymphology* 1984; **17**: 50–3.
5. Grobmyer SR, Daly JM, Glotzbach RE, Grobmyer III AJ. Role of surgery in the management of postmastectomy extremity angiosarcoma (Stewart–Treves syndrome). *J Surg Oncol* 2000; **73**: 182–8.