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Case Report

CASE REPORT

Multi-system bleeding risk with a cutaneous angiosarcoma at an arteriovenous fistula site

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Abstract

We present a case of angiosarcoma at an arteriovenous fistula site in a non-immunocompromised patient presenting as a soft tissue swelling with associated findings suggestive of lung malignancy, metastases and bleeding diathesis. This patient died of an otherwise unexplained subdural haematoma. Given the ability of this tumour to metastasise early and the poor prognosis of angiosarcoma without adequate resection, this needs to be considered early in any differential diagnosis of soft tissue masses near an arteriovenous fistula.

INTRODUCTION

Angiosarcoma is an extremely rare soft-tissue sarcoma arising in endothelial tissue. It comprises <1% of sarcomas and there have been case reports of angiosarcomas in almost every organ [1]. The most common presentation of angiosarcoma is a cutaneous disease. These are broadly grouped as scalp and face presentations, those related to chronic lymphoedema (Stewart–Treves syndrome) and iatrogenic radiation-related disease. Angiosarcoma has been linked with exposure to toxins such as vinyl chloride. Angiosarcoma metastasises early and thus has an extremely poor prognosis [1].

We present the case of a patient, with chronic kidney disease (CKD) and arteriovenous fistula (AVF), who developed skins lesions overlying the fistula.

CASE REPORT

A 71-year-old patient with a medical history of ischaemic heart disease, insulin-dependent diabetes mellitus, hypertension and CKD stage 4 was referred to respiratory clinic with four weeks of increasing shortness of breath, cough, weight loss and subsequent haemoptysis.

A left brachio-cephalic AVF had been created 2 years and 10 months (34 months) previously but had never been used for haemodialysis. Three months before referral to respiratory clinic, he had been seen with a swollen and warm fistula site and diagnosed with partial fistula thrombosis, confirmed by duplex ultrasound. Pain and tenderness developed over the following month, consistent with mild thrombophlebitis.

In respiratory clinic, the patient had a 60 pack-years smoking history, potential exposure to asbestos and a significant swelling/mass at the AVF site. He was admitted from clinic for further investigation of bilateral pleural effusions with significant anaemia (haemoglobin 64 g/L on a baseline 110 g/L), and the arm swelling. CT thorax demonstrated bilateral pulmonary and pleural nodular and mass-like opacities with associated bilateral pleural and pericardial effusions. A chest drain was

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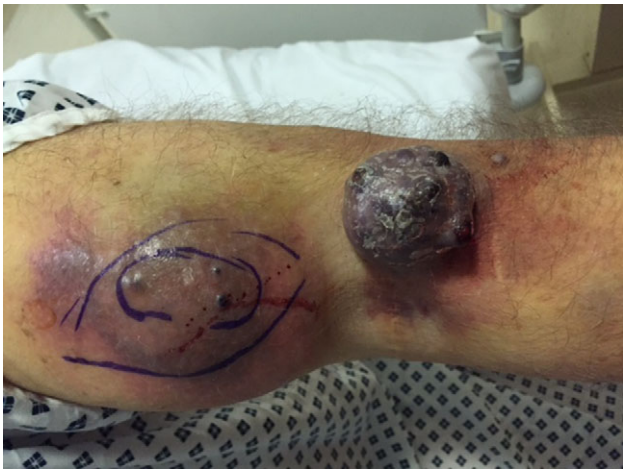

Figure 1: Macroscopic appearance: anterior left upper limb with distal fungating lesion and proximal swelling.

inserted and the strongly blood-stained pleural fluid was sent for cytology that showed non-specific inflammatory cells and no malignant cells. On admission the patient was noted to have a fever with erythema at the AVF site, diagnosed with cellulitis and treated with intravenous flucloxacillin. He received 2 units of packed red blood cells.

Admission blood tests had demonstrated no apparent clotting abnormality (prothrombin time 11.4 s, international normalized ratio 1.1, activated partial thromboplastin time ratio 1.2, platelets 481 $\times$ 10^9/L). These remained stable repeatedly throughout admission with nadir platelets of only 269 $\times$ 10^9/L. He was taking aspirin for secondary prevention of ischaemic heart disease, but no anticoagulants or other drugs predisposing to bleeding.

Review of the swelling by renal surgeons suggested the possibility of a distal fistula aneurysm with proximal haematoma (Fig. 1). Given the unusual nature of this lesion a decision was taken to proceed to surgery. Aspirin was withheld pending surgical intervention.

The patient was taken to theatre on Day 5 of admission and the significant necrotic soft tissue mass with associated haematoma was excised. The preserved, non-aneurysmal, thrombosed fistula was ligated and loose closure was used with a plan to re-explore at Day 8. The lesion was sent for histology due to its abnormal macroscopic appearance. The wound continued to bleed and haemostasis under a brachial block was performed on two occasions along with further packed red blood cell transfusions to a total of 6 units. Aspirin and prophylactic low molecular weight heparin were stopped indefinitely on Day 7. The patient then became hypotensive with a fever, and chest x-ray showed new basal collapse with worsening pleural effusions. The patient was treated with piperacillin/tazobactam for hospital acquired pneumonia.

Further extensive tissue debridement was performed at Day 9 with plastic surgeons in joint care with complete closure of the wound on Day 12. Two further units of red blood cells were transfused. The patient remained stable until Day 16. He continued to have blood-stained fluid from his chest drain. He suddenly deteriorated with tachypnoea, reduced Glasgow Coma Score (GCS) and left-sided weakness. A CT head was performed showing right fronto-temporo-parietal subdural haematoma with mass effect, intraventricular haemorrhage, evolving hydrocephalus and right uncal herniation.

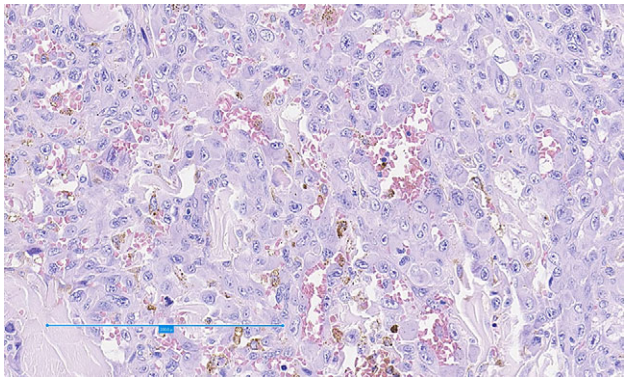

Figure 2: Microscopic appearance: slide of the excised lesion (at ×40 magnification, H&E stain) showing undifferentiated angiosarcoma. Scale represents 200 μm.

In view of his co-morbid state, a palliative care plan was instituted. He died 4 days later. Subsequent histology confirmed undifferentiated angiosarcoma from the arm tissue excised (Fig. 2).

DISCUSSION

A variety of clinical presentations of angiosarcomas have been described including painful enlarging soft tissue masses, long bone fractures and high-output cardiac failure related to arteriovenous shunting. Of relevance, 4 of 22 patients in case reports presented with a bleeding diathesis [2]. In patients with end-stage renal failure a high proportion of case reports have a renal transplant who are likely to be at higher risk due to the immune suppression required [2]. However, there appears to be a predilection for angiosarcoma in AV fistulae that is separate from iatrogenic immunosuppression [3–5]. The cause for this is unknown, although a number of potential theories have been propounded [5].

Treatment relies on complete excision, including limb amputation if required, and local adjuvant radiotherapy [1, 2]. Chemotherapy has some role in metastatic disease but not with curative intent [2].

This case demonstrates the importance of multi-disciplinary team approach to complex renal patients. Angiosarcoma would normally be a relatively unlikely explanation for an AVF-associated mass. ‘Red flags’ for malignancy or an unexplained bleeding diathesis in this scenario should prompt the clinician to give consideration to angiosarcoma as a differential diagnosis. Early diagnosis is key to successful management and a high index of suspicion by all clinicians provides the best opportunity to treat this condition before early metastasis or irreversible haemorrhage.

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest with this publication. A.B. and P.J.Y. are guarantors for this article.

CONSENT

Patient details were anonymised in keeping with ICMJE recommendations and the declaration of Helsinki. The patient’s next of kin consented to publication of this case report.

REFERENCES

1. Koch M, Nielsen GP, Yoon SS. Malignant tumours of blood vessels: angiosarcomas, haemangioblastomas, and haemangiopericytomas. *J Surg Oncol* 2008;**97**:321–9.
2. Oskrochi Y, Razi K, Stebbing J, Crane J. Angiosarcoma and dialysis-related arteriovenous fistulae: a comprehensive review. *Eur J Vasc Endovasc Surg* 2016;**51**:127–33.
3. Chanyaputhipong J, Hock DL, Sebastian MG. Disseminated angiosarcoma of the dialysis fistula in 2 patients without kidney transplants. *Am J Kidney Dis* 2011;**57**:917–20.
4. Farag R, Schulak JA, Abdul-Karim FW, Wasman JK. Angiosarcoma arising in an arteriovenous fistula site in a renal transplant patient: a case report and literature review. *Clin Nephrol* 2005;**63**:408–12
5. Webster P, Wujanto L, Fisher C, Walker M, Ramakrishnan R, Naresh K, *et al*. Malignancies confined to disused arteriovenous fistulae in renal transplant patients: an important differential diagnosis. *Am J Nephrol* 2011;**34**:42–8.