A rare case of primary epithelioid angiosarcoma of the ureter

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ABSTRACT

Angiosarcoma is an extremely rare vascular malignancy with a 1-year survival rate of 50%, regardless of tumour origin. Distant metastases are common and occur in the lungs, bone, lymph nodes and soft tissues. The majority of patients with angiosarcoma present with localised disease, although 25–45% have distant metastases at presentation. There are few reported cases of angiosarcomas of the bladder and we report the second case in the literature of primary angiosarcoma of the ureter. We suggest that, in atypical manifestations of suspected urothelial malignancy, the multidisciplinary team considers the possibility of rarer, aggressive tumours early. This may influence clinical decision making towards offering radical treatments earlier, before it is too late to do so owing to aggressive oncological behaviour.

KEYWORDS

Angiosarcoma – Haematuria/etiology – Ureter/pathology – Male – Ureteral neoplasms

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Introduction

Angiosarcoma is an extremely rare vascular malignancy with a 1-year survival rate of 50% regardless of tumour origin.¹ We report the second case of epithelioid angiosarcoma of the ureter in the literature.

Case history

A 67-year-old male with mild cognitive impairment presented with clot retention. Cystoscopy revealed blood oozing from the left ureteric orifice and attempted ureteroscopy was abandoned due to poor views. A computed tomography (CT) urogram showed an obstructed left kidney with a dilated pelvicalyceal system and ureter to the level of the common iliac bifurcation. There was increased density of the ureter at this level, raising suspicion of a urothelial lesion. A poorly demarcated, partly calcified soft-tissue mass involving the distal right common iliac artery was also reported and was thought to represent possibly an aneurysm, an inflammatory mass or a nodal mass. Management options including radical nephroureterectomy were discussed at the multidisciplinary team meeting. However, the patient initially opted for a conservative approach with clinical and symptomatic review.

Unfortunately, the patient re-presented 3 weeks later with persistent haematuria and symptomatic anaemia. His case was re-discussed and he was listed for a left nephroureterectomy during that admission. Histopathology revealed a high-

grade vascular tumour, which was later confirmed as an epithelioid angiosarcoma.

Three weeks following nephroureterectomy, the patient underwent CT of the chest, abdomen and pelvis. This demonstrated progression of the poorly defined partially calcified soft-tissue mass in the region of the left common iliac artery, with possible periureteric extension of the primary tumour. The patient was deemed unfit for palliative chemotherapy.

Discussion

Angiosarcomas are rare, aggressive soft-tissue sarcomas of endothelial origin.^{2,5} Although they can arise anywhere, angiosarcomas have a higher predilection for the head and neck region or the extremities.^{2,4} They can also arise following irradiation and, hence, secondary breast angiosarcomas are an important subgroup.² Although angiosarcoma was first reported in the skin and thyroid gland, a significant number of patients are diagnosed with a primary abdominal lesion or in other visceral sites such as the lung or adrenals.⁵ Distant metastases are common and occur in the lung, bone, lymph nodes and soft tissue. The majority of patients present with localised disease, although 25–45% have metastases at presentation.⁵ The prognosis remains poor, with an overall 1-year survival rate of 50% and 5-year survival rate of just 35%.^{1–5}

Intrinsic non-epithelial tumours of the ureter are rare.⁶ Histologically, distinguishing angiosarcoma from epithelioid

haemangioendothelioma is challenging. The presence of vascular differentiation with irregular anastomotic vascular channels, high nuclear to cytoplasmic ratio and pleomorphism is considered diagnostic.⁵ In addition, the presence of solid growth favours the diagnosis of epithelioid angiosarcoma.⁵ There are fewer than ten reported cases of angiosarcoma of the bladder and we report only the second case in the literature of primary angiosarcoma of the ureter.⁶ In the previous reported case, the patient underwent a radical nephroureterectomy. The histological features were of a low-grade angiosarcoma. The patient made a good recovery, owing to the tumour's small size, mobility and the fact that it was completely excised.⁶ We suggest that, in atypical manifestations of suspected urothelial malignancy, the multidisciplinary team considers the possibility of rarer tumours early. This may influence clinical decision making towards offering radical treatments earlier, before it is too late to do so owing to aggressive oncological behaviour.

In this instance, achieving a ureteric biopsy might have persuaded the patient to opt for an earlier nephroureterectomy.

This would be the gold standard of management but an advanced stage at presentation and a high chance of metastases would appear to make surgical resection less successful in such cases. The emphasis therefore would be on identifying metastatic disease that might make surgery unwise unless it provides good palliation, as it did in this case.

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