CASE REPORT

Angiosarcoma of common iliac vein

Kamuran Ibis,¹ Ufuk Usta,² Rusen Cosar,³ Cem Ibis⁴¹Department of Radiation Oncology, Trakya University Medical Faculty, Istanbul, Turkey²Department of Pathology, Trakya University Medical Faculty, Edirne, Turkey³Department of Radiation Oncology, Trakya University Medical Faculty, Edirne, Turkey⁴Department of Surgery, Istanbul University Istanbul Medical Faculty, Istanbul, Turkey

Correspondence to

Dr Kamuran Ibis, kamuranibis@gmail.com

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SUMMARY

Angiosarcoma is a rare malignant tumour of endothelial cells. Primary angiosarcoma of venous origin is extremely rare, and has a very poor prognosis. A 63-year-old woman with retroperitoneal mass underwent en bloc resection on a part of iliac vein followed by adjuvant radiotherapy. No recurrence was detected during 3 years of follow-up.

BACKGROUND

Primary malignant tumours of venous origin are extremely rare. Primary angiosarcoma of an iliac vein is also a very rare diagnosis. We present a case of angiosarcoma of the common iliac vein that underwent a surgical resection followed by radiotherapy. Adequate local control of the tumour could be achieved through surgical resection followed by adjuvant radiotherapy without chemotherapy.

CASE PRESENTATION

A 63-year-old woman without a history of previous exposure to polyvinyl chloride or any other toxic agent or to radiation was admitted to a hospital due to lower abdominal pain that had lasted for more than 6 months. She had no history of tobacco or alcohol use. She had been using antihypertensive drugs for 20 years. No pathology was detected in the physical examination.

INVESTIGATIONS

An abdominal MRI revealed a contrast-enhanced nodular mass in the inferior aspect of the bifurcation of the aorta. The negative whole body I-123 metaiodobenzylguanidine (MIBG) scan excluded the possible diagnoses of pheochromocytoma, paraganglioma and neuroblastoma. CT and F-18 fluorodeoxyglucose positron emission tomography/CT (FDG PET/CT) demonstrated the nodular mass

with focal hypermetabolic activity with a maximum standard uptake value (SUVmax) of 8.7 only on the lesion side (figure 1A, B).

TREATMENT

In surgery, a 3×3 cm well-demarcated mass in the anterior aspect of the bifurcation of the aorta was detected. The lesion could easily be separated from the left common iliac artery but the surgical team observed an obvious continuity of the lesion into the left common iliac vein. With the help of the vascular surgery team the mass was removed en bloc with the invaded area of the left common iliac vein. The left common iliac vein was repaired with a synthetic patch graft. The patient was discharged after an uneventful postoperative period on the fifth day after surgery.

Microscopically, the tumour was mostly composed of mesenchymal cells with epithelioid morphology forming a solid pattern (figure 2A). In better differentiated areas there were slit-like or well-formed anastomosing vascular spaces that were lined by highly atypical endothelial cells and that often had hub-nail morphology (figure 2B, C). The mesenchymal tumour was infiltrating smooth muscle fibres (figure 2D). The tumour cells had nuclei with prominent pleomorphism, and high mitotic activity with frequent atypical mitotic figures (figure 2E). Immunohistochemically (IHC), the tumour showed diffuse and strong cytoplasmic positivity for antibodies vimentin, Factor VIII, CD31 (figure 2F) and CD34 (figure 2G), and was negative for pankeratin, α smooth muscle actin (figure 2H), desmin, CD68, CD117, Dog1 and S100 protein. Ki-67 proliferation index was over 50% in most areas of the tumour.

With detailed histopathological examination of the surgical specimen, along with preoperative radiological and intraoperative findings, the tumour

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Figure 1 (A) Axial CT demonstrates a soft tissue mass (arrow) between common iliac arteries and veins. (B) Axial 18-FDG PET/CT (F-18 fluorodeoxyglucose positron emission tomography/CT) demonstrates FDG uptake in this area.

Figure 2 (A) The tumour area showing solid pattern of tumour cells with epithelioid morphology (H&E×100). (B and C) A better differentiated area of the tumour forming vascular spaces lined by atypical endothelium with hobnail morphology (H&E ×100 and ×200, respectively). (D) The mesenchymal tumour infiltrating smooth muscle fibres (H&E×50). (E) The tumour cells with prominent nuclear atypia and frequent mitotic figures (H&E×400). (F and G) Strong cytoplasmic positivity for CD31 and CD34 (IHC×100). (H) The negative reaction of the tumour for α smooth muscle actin and positivity of infiltrated muscle fibres by the tumour (IHC×50) (IHC, immunohistochemical stain).

was interpreted as an angiosarcoma originating from the vasa vasorum in the adventitial layer of the left common iliac vein. Unfortunately, no frozen section examination was considered directly after surgical resection of the mass. Microscopically, the surgical margin was found to be positive. Neither re-excision of the common iliac vein and grafting nor adjuvant chemotherapy was offered to the patient. External beam radiation therapy was planned, and the patient received 54 Gy in 30 Fr through three-dimensional conformal radiotherapy.

OUTCOME AND FOLLOW-UP

No distant metastasis or local recurrence was detected in the 3-year follow-up period (including physical examination and routine abdominal and thoracic CT scans).

DISCUSSION

Angiosarcomas are a relatively rare histological subtype of sarcomas that represent less than 1% of all sarcomas. In contrast to their rarity, angiosarcomas appear in a large variety of clinical forms in different parts of the body.¹

The most common clinical form of angiosarcoma is a cutaneous tumour, characteristically in the scalp or face of an old-aged patient. Nevertheless it can occur in any part of the body, including deep soft tissues, breast, visceral organs and bone.² Most cases are sporadic, however, they can be seen in a previously irradiated field,³ and in a chronically lymphedematous extremity (Stewart-Treves syndrome).⁴ Angiosarcomas can be multifocal,⁵ and metastasise to lymph nodes and distant soft tissues.⁶

With the help of IHC staining, the diagnosis of poorly differentiated angiosarcoma, which often presents as an undifferentiated malignant neoplasm, can be confirmed. The most useful marker is CD31, which, unfortunately, is not ideal due to its low-level staining characteristics in other tumours, and also positive staining for macrophages.⁷ Angiosarcomas can usually be intensely stained with CD31, often highlighting an obvious cytoplasmic membrane. Although most angiosarcomas can be detected with CD34 and factor VIII, poorly differentiated tumours can only be stained infrequently with these markers.⁶

Angiosarcomas are very aggressive tumours, and most patients die due to disseminated disease. According to its highly infiltrative nature, the tumour extends rapidly and is often multifocal,

resulting in recurrences.² Early diagnosis can be life-saving. With the help of PET-CT imaging we were able to promptly decide to operate. PET-CT imaging in diagnosing the angiosarcoma has been reported before.⁸ Venous angiosarcomas, though rare, have previously been reported.^{8–13} The literature review in PubMed through the key words “vein”, “venous”, “vessel” and “angiosarcoma” revealed only 20 cases reporting an angiosarcoma of primary venous origin. Primary tumours of major blood vessels can be divided into different groups depending on the original site.¹⁴

In the preoperative and also perioperative setting it was extremely difficult to distinguish the origin of the tumoral mass in our case. This shows the complexity of diagnosis and treatment of such tumours and the need for a multidisciplinary approach.¹⁵ The treatment of localised disease is surgery followed by radiotherapy, which is reported to be highly successful with local control. It has been reported that paclitaxel-based and doxorubicin-based regimens can be valuable in patients with advanced disease.^{15 16} All data regarding uncommon great vessel angiosarcomas were obtained from case reports and case series including a limited number of patients. While post-operative radiotherapy is used to improve local control, post-operative chemotherapy is controversial. However, Fatima *et al*¹⁷ recently reported better survival rates with triple therapy

Learning points

- ▶ Angiosarcomas rarely originate from veins.
- ▶ Positron emission tomography/CT is valuable in detecting primary lesions and metastases.
- ▶ Small case series including cardiac, aortic and venous angiosarcomas suggest surgical resection followed by adjuvant radiochemotherapy. However, radiotherapy may help to control the disease even with positive surgical margin without chemotherapy, particularly in cases where the angiosarcoma cannot be resected with negative surgical margin.
- ▶ Close follow-up is sine qua non for detecting recurrences.

including surgery, radiotherapy and chemotherapy than with single treatment modality. Resection and grafting of the common iliac vein could be an option to achieve a negative surgical margin; however, this option was not offered to the patient by the surgical team. Chemotherapy was not offered to the patient by the medical oncology team either. Systemic involvement was excluded with the help of PET-CT scanning. This encouraged us to achieve local control of the venous angiosarcoma with radiotherapy, resulting in a positive surgical margin in this particular patient. There has been no recurrence during 3 years of follow-up.

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Competing interests None.

Patient consent Obtained.

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