CASE REPORT

Primary Sarcoma of an Iliac Artery Aneurysm

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Introduction

Vascular tumours are heterogenous neoplasms ranging from the common benign hemangiomas of small vessels, to the very rare and malignant spindle cell sarcomas of large vessels. Since the first report of a primary tumour of a large artery in 1873 there have been only 45 reported cases in the world literature.

The majority (75%), of large vessel tumours arise in the thoracic and abdominal aorta. Of the peripheral arteries, the iliac and femoral are the most frequently affected. We report a case of an undifferentiated sarcoma ensuing in a degenerative aneurysm of the iliac artery. We also review the literature as it pertains to tumours arising in the aorta and its major branches.

Case History

A 56-year-old male, heavy smoker, presented with right flank and lower back pain of 8 months' duration. The pain radiated to the right scrotum and thigh. He also complained of intermittent claudication in the left lower extremity. An IVP and a retrograde urogram, done at another hospital, revealed an obstructed right kidney, and a dilated ureter with partial obstruction where it crossed the iliac vessels. Abnormal findings on physical examination included a 5 cm tender pulsating mass in the right iliac fossa and absent peripheral pulses in the left lower extremity.

A CT scan of the abdomen reported bilateral iliac artery aneurysms with obstruction of the right ureter at the level of the right iliac aneurysm. The left common iliac artery aneurysm was thrombosed. An aortogram confirmed the CT scan findings (Fig. 1). With his symptoms persisting, we urgently operated on the patient.

There was ectasia of the infrarenal abdominal aorta with localised aneurysmal dilatation of the common iliac arteries. The left iliac artery was thrombosed and contained laminated thrombus. Grossly, the aneurysm wall and thrombus were normal in colour and consistency. The right iliac artery aneurysm consisted of an inflammatory mass; the wall was thick, shiny and white. The aneurysmal dilatation extended into the internal iliac artery; the external iliac was normal. The ureter and iliac vein were entrapped.

Fig. 1. Transfemoral arterial DSA of the aorto-iliac arteries showing a large aneurysm of the right common iliac artery with contrast medium filling a small compartment of the aneurysm. There is thrombosis of the left common iliac artery.
in the inflammatory mass; otherwise the retroperitoneum was normal. Inside the lumen the thrombus looked unusual; it was light gray, translucent, gelatinous, very friable, with no evidence of penetration into the aneurysm wall. There were no perivascular lesions growing into the aneurysm wall.

The aorto-iliac segment was reconstructed with a bifurcated woven Dacron graft, using Prolene sutures for the anastomoses. A biopsy was taken but the aneurysmal wall was not resected. The patient tolerated the procedure without complications.

Our operative diagnosis was an inflammatory aneurysm with retroperitoneal fibrosis. However, the tissue diagnosis on the right iliac artery aneurysm contents was that of sheets of undifferentiated tumour cells. In the patient's record we could not find any abnormal preoperative laboratory parameters suggesting that he had a malignancy or metastasis. His body weight, chest X-ray, blood count, serum proteins, liver function tests, sedimentation rate, and temperature were normal.

Unfortunately, the patient refused further investigation or surgery and elected to be treated abroad where his family resided. Six months later, we learned that the patient was terminally ill. He had received only a course of radiotherapy. Unfortunately, we do not have any additional information about his management or condition.

**Histology**

The contents of the right iliac artery aneurysm consisted mainly of fragments of a partially necrotic tumour, mostly composed of sheets of highly pleomorphic malignant cells with numerous bizarre giant cells (Fig. 2). In areas, however, tumour cells were spindle shaped and exhibited a fascicular arrangement. There were also fragments of atheromatous plaque and pieces of fresh as well as organising thrombus. Some of the thrombus material was adherent to tumour, and few foci of tumour cells were noted within atheromatous fragments. Biopsy of the aneurysm wall revealed portions of arterial wall with tumour infiltrating the adventitia associated with fibrosis and chronic inflammation. The contents of the left iliac artery aneurysm consisted of laminated thrombus.

The light microscopic appearance of this tumour was consistent with a poorly differentiated sarcoma. Immunohistochemical stains showed the tumour to be weakly positive for vimentin (a mesenchymal marker), and negative for factor VIII-related antigen and *Ulex europeus* I lectin (both endothelial markers); stains for cytokeratin (an epithelial marker) and S-100 protein (a melanocytic marker) were also negative. The electron microscopic appearance of the tumour was also consistent with a sarcoma; however, there were no additional ultrastructural differentiating features.

**Discussion**

Vascular tumours arise from one of two groups of cells that are normally present in blood vessels; the endothelial cells lining the lumen, and non-endothelial cells located in the vessel wall. The latter group includes pericytes, smooth muscle cells, and glomus cells. The majority of vascular tumours arise from small vessels and most of these are benign. Tumours of large arteries, on the other hand, are rare and invariably malignant.

Small vessel tumours, on the other hand, have characteristic appearances that are easily recognised with light microscopy and routinely stained sections. Identifying the different types of large vessel tumours, however, requires the additional use of immunohistochemical stains or electron microscopy. Despite these techniques, the histologic categorisation of large vessel tumours remains difficult. Consequently, Wright et al. proposed dividing primary tumours of large arteries into intimal and mural, based on their location in the artery wall, and on their clinical behaviour.

The intimal varieties are predominantly intraluminal tumours. The most commonly reported symptoms are related to arterial occlusion with intermittent claudication more frequently reported than embolic metastases. Mural sarcomas, on the other hand,
Iliac Aneurysm Sarcoma

References


Fig. 3. Contrast enhanced CT scan obtained through the level of the common iliac arteries showing the aneurysm of the right common iliac artery with well defined and intact wall and three intraluminal components of different attenuation values. A thrombus (big black arrow head), an enhancing tumor mass (big open arrow head), and the true lumen (small arrow head). The left common iliac artery aneurysm is filled with homogeneous thrombus.

arise from the media or adventitia and produce locally aggressive masses that rarely metastasise. The associated clinical picture is invariably non-specific, with systemic symptoms of malignancy predominating. Symptoms of vascular insufficiency can be present if the arterial lumen is invaded as well.

In our patient, the symptoms were related to the obstructed right ureter and the local pressure effects of the aneurysm. In retrospect, however, we realised that the right iliac aneurysm appearance, on CT images, was atypical. The CT scan shows a well circumscribed right common iliac artery aneurysm, with three intraluminal structures of different CT attenuation values. There is a low density mass representing a thrombus, an enhancing mass the tumour, and the round dense lumen of the distal common iliac artery (Fig. 3). These CT findings contrast with the homogenous thrombus occluding the contralateral iliac aneurysm. Only three published reports of primary tumours of the aorta describe CT findings, but there is no reference to any appreciable attenuation difference between the intraluminal tumour and thrombus.

Angiographic definition of an intraluminal malignancy from thrombus or atheroma can be difficult, especially if the tumour is sessile or covered with thrombus. It should be kept in mind that, when an arterial obstruction is discrete or a filling defect is polypoid, in the absence of atherosclerosis or embolism, a neoplastic lesion should be considered. This fact becomes more significant if there are systematic signs or symptoms of malignancy.

Finally, the gross appearance of the aneurysm contents, intraoperatively, should have alerted us to the unusual nature of the thrombus. An intraoperative frozen section biopsy of this bizarre looking thrombus would have confirmed the diagnosis of a malignancy, and certainly altered our surgical approach. We failed to recognise this rare entity preoperatively and intraoperatively, and subsequently, our surgical approach was inappropriate.

The management of primary arterial tumours remains unclear, with a reported average survival of 1.5 years. Even in the absence of distant metastasis local recurrence is an important cause of death. The poor survival, therefore, seems to be related to late diagnosis and inadequate surgical resection. When the index of suspicion is high, additional diagnostic modalities, and in some instances a biopsy of the lesion can be helpful. The finding, on CT scanning or MR imaging, of a contrast enhancing filling defect with attenuation values different from thrombus or blood, should raise suspicion as to the nature of the lesion. In selected cases, a diagnostic biopsy using endovascular techniques or CT guidance, can be helpful in the management plan. Certainly, the diagnostic yield of these invasive techniques must be weighed against their risks, after considering the general condition of the patient.

Improvement in the overall survival of patients requires complete resection of the involved artery and any invaded contiguous structures. The role of adjuvant therapy is not known. Since there are no series addressing the management of primary large artery tumours, it is reasonable to approach them similar to retroperitoneal soft tissue sarcomas; i.e. radical surgery seems to be the treatment of choice.


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