CASE REPORT

Multi-Disciplinary Approach to Haemangiopericytoma Uteri

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Introduction

Haemangiopericytoma accounts for less than 2% of soft-tissue sarcomas, but this uncommon mesenchymal neoplasm may be found throughout the body. They arise from the pericytes¹ and comprise of uniform elongated cells surrounding capillaries and post-capillary venules. Histological diagnosis of haemangiopericytoma is often difficult to distinguish from other soft-tissue neoplasms which may have areas of rich haemangiopericytoma-like vascularity.

We report here a case of haemangiopericytoma in the pelvis, treated by pre-operative embolization and surgery, with preservation of all of the abdominal and pelvic viscera.

Clinical Presentation

A 34-year-old pregnant woman with a previous diagnosis of myalgic encephalopathy (ME) by her general practitioner, began to experience progressively worsening episodic pelvic pain from the 8th week of gestation onwards. Ultrasonography demonstrated a mass behind the uterus, thought originally by her gynaecologists to be a fibroid.

She continued the pregnancy and at 38 weeks’ gestation, underwent elective Caesarian section, and delivered a healthy male infant. At the same time, a large highly vascular mass was found behind and attached to the uterus. Biopsy of the mass yielded histology suggestive of haemangiopericytoma.

On the 10th day postpartum, she had a MRI scan which confirmed the position of the large tumour. By this stage, she complained of constipation, with feelings of incomplete urination. She also had self-limiting episodes of bleeding per rectum, and there was symptomatic anaemia. The pelvic discomfort had persisted throughout.

Clinical Management

At 3 months post partum, she was referred to our centre for surgical intervention. Clinical examination revealed a large tender mass measuring 15 by 10 cm, extending from the pelvis into the abdomen. Colonoscopy showed the mass indenting, but not invading, the sigmoid colon, with evidence of mucosal venous congestion. Angiography showed that the tumour received its blood supply from branches of the internal iliac arteries on both sides, and also from the inferior mesenteric artery. The feeding branches were selectively catheterized and embolized with Contour particles (500–1000 U) followed by coils in the large branches of the internal iliac arteries. The final images showed very little filling of the tumour.

At laparotomy, a well-circumscribed tumour (16 × 11 × 6 cm), weighing 656 g, and adherent anteriorly to the uterus, was removed. The uterus and all other abdominal viscera were preserved intact. The para-aortic and internal iliac lymph nodes were biopsied. Because of the vascularity of the lesion, a

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cell-saver was employed and patient only had four units of banked blood transfused throughout the perioperative period. The woman remained well in the early postoperative period but serial scanning will be required to rule out recurrent disease.

**Histology**

The tumour was a smooth-surfaced mass (16 × 11 × 6 cm) with a lobulated appearance. Bisection of the tumour revealed variegated tan and mucoid cut surfaces. Haemorrhage and necrosis was not a feature. Microscopically, the tumour was variably cellular and consisted of cells with small basophilic, angulated nuclei and scant cytoplasm arranged around anastomosing capillary channels. Some areas showed a myxoid appearance, whilst others were densely cellular with mild nuclear polymorphism. There were scant mitoses (<3/10 high power field). The lymph nodes biopsied were all negative for tumour, however, one large vein in the capsule showed tumour invasion. A panel of immunohistochemical markers were used in order to assist with diagnosis. Results were as follows: staining was strongly positive for vimentin in all of the tumour cells, but staining for CD34 (Q bend 10), smooth muscle actin, Desmin, S100, and cytokeratin (cam5.2) were negative. The slides were reviewed in conjunction with an expert in soft-tissue pathology. The appearances were thought to be consistent with the diagnosis of haemangiopericytoma, however, the negative SMA was unusual and a differential diagnosis of solitary fibrous tumour was proposed as an alternative consideration. The low mitotic count and absence of necrosis imply low grade tumour but biological behaviour is unpredictable and follow-up with exclusion of metastases is advised.

**Discussion**

Haemangiopericytoma, though rarely encountered in clinical practice, is a well-described vascular neoplasm. The tumour arises from contractile spindle cells called pericytes of Zimmerman, which surround capillaries and post-capillary venules. Since its first description in 1942,1 numerous accounts of this neoplasm have been reported. Most of these articles are individual case reports, and are concerned more with the clinical and therapeutic aspects, rather than the problems of histological diagnosis.

Haemangiopericytoma may present at any age. There are reported cases from birth to 92 years with a peak incidence in the fifth and sixth decades.2 There is no significant difference in incidence amongst male or female. Although these tumours are ubiquitous and occur wherever capillaries are found, they tend to occur most commonly in the lower extremities and in the retroperitoneum.3 The uterus is a relatively uncommon site with only about 100 cases reported worldwide.

The clinical presentation is usually neither striking nor characteristic; with a majority of patients presenting with a slowly enlarging painless mass. Pain or tenderness is rarely a feature and may be due to pressure on surrounding viscera and nerves. Some of the symptoms clearly relate to the site. Tumours located in the pelvic fossa may cause urinary retention, constipation (as in our case), dysuria, hydronephrosis, and even haematuria or bleeding per rectum. Tumours situated in other sites can cause epistaxis, cough, dyspnoea, upper gastrointestinal bleeding, or abdominal distension. Various paraneoplastic effects have been reported and these include hypoglycaemia, virilization, or gynecomastia.4 Since the tumour is highly vascular with low resistance to blood flow, there is often telangiectasia of the overlying skin with elevated skin temperature. It may also act as an arteriovenous shunt, and precipitate cardiac failure.

B-mode gray-scale ultrasound examination is cheap, non-invasive and simple; but relatively non-specific. Lesions may be hyper- or hypo-echoic, may contain cysts,5 or appear entirely solid. Recently, with the advance of Doppler ultrasound technology, this has become the first-line investigation in haemangiopericytoma, to distinguish it from other solid tumours, or from arteriovenous malformations.6

Angiographically, because of the highly vascular nature of the tumours, they appear as dense, well-circumscribed lesions due to the accumulation of contrast medium in the delicate capillary-like vessels (‘the diffuse capillary blush’) of the tumour vascular bed.7 Draining veins can also be detected on angiography or on thin-cut CT scans, which also show the mass with central areas of low attenuation consistent with necrosis.

The tumours are seen well on MR images, and have the expected high signal of soft-tissue tumours on T2-weighted images. This is perhaps the reason why MRI is becoming the imaging test of choice for haemangiopericytoma. If the tumour is causing mass effect on the other abdominal viscera, additional images might be necessary, such as an excretory urography, or barium gastrointestinal investigations. A chest radiograph
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is mandatory to exclude metastases. Endoscopy should be performed when appropriate. Biopsy should be approached with caution because of the risk of major haemorrhage.

These tumours are very vascular and risk of haemorrhage per and postoperatively is high, therefore, embolization of some of the feeding vessels to decrease vascularity at the time of angiography should be considered.8 This is often an adjunct to successful resection of large haemangiopericytoma.

Surgery is the only curative treatment; and in the case of uterine haemangiopericytoma, might include total hysterectomy and bilateral salpingo-oophorectomy. Grossly, the lesions tend to be well circumscribed with a thin vascular pseudocapsule, and have a smooth, sometimes slightly nodular or lobulated surface.3 Some are firmly attached to the underlying tumour bed, whilst others can be shelled out easily. The tumours are often covered by a plexiform meshwork of large vessels, and the whole surface is susceptible to a general ooze. Sometimes with uterine tumours, it is necessary to ligate the internal iliac arteries to halt the bleeding.9 Massive transfusion is often necessary, and the use of cell-saver is appropriate.

The tumours range from 1 to 21 cm in diameter, most being 4–8 cm.1 Haemangiopericytoma display a solid homogeneous appearance, with dilated vascular spaces. The colour varies from white, pink and tan, to brown or purple. In large tumours, areas of cystic degeneration and haemorrhage may be present. Calcification is unusual, and necrosis is common in the more malignant tumours.

Microscopically, the tumour is composed of tightly packed spindle-shaped cells about thin walled, endothelium lined vascular channels.3 Ultrastructural studies have confirmed the pericyte origin of these tumours. The cytoplasm is ill defined in most of the cases, and the nuclei vary from round to oval. Solid cellular areas may be seen with intervening areas of mixed cellularity and myxoid appearance, representing cell degeneration. Silver impregnation techniques can be used to confirm that these cells are outside the basement membrane of the endothelium, and hence are pericytes rather than endothelial cells.10 A distinction between benign and malignant haemangiopericytoma cannot be made in all cases, but the presence of high mitotic activities (>4/10 high power field), focal necrosis, haemorrhage, or cellular polymorphism are suspicious of rapidly growing tumours that are likely to recur. However, tumours that appear histologically benign can still harbour the potential to behave aggressively, therefore, the clinical behaviour of haemangiopericytoma cannot be predicted with certainty.

Haemangiopericytoma are rare tumours, and may create differential diagnostic problems, especially when they have an atypical appearance. They may resemble other more common sarcomas such as fibrous histiocytomas, mesenchymal chondrosarcomas, liposarcomas, and synovial sarcomas. But these difficulties can usually be resolved by the use of a panel of immunohistochemical markers as described.11 One study has shown that immunohistochemical staining with a monoclonal antibody to proliferating cell nuclear antigen (PCNA), a 36 kDa nuclear protein which is associated with the cell cycle, correlates well with the histological grade of haemangiopericytoma.12 Analysis of DNA ploidy and proliferative index by flow-cytometry can also be used in predicting the behaviour, stage, and response to treatment in haemangiopericytoma.13 This technique has been widely used in several other epithelial neoplasms, including carcinoma of the colon, breast and ovary.

In cases where the tumour is unresectable or partially resected, chemotherapy and radiotherapy, which usually involves high dose of irradiation, can be used.3 Jha N et al. reported nine patients with haemangiopericytoma who received radiotherapy, as an adjunct to surgery. These nine patients have survived from 3.5 to 20 years free from disease, and the results suggested that postoperative radiation therapy should be considered as an integral part of the primary treatment of haemangiopericytoma.14 Chemotherapy using a combination of cyclophosphamide, nitrogen mustard, actinomycin-D and vincristine,15 has been advocated in locally recurrent or metastatic tumours.

Since most reports in the literature concern small series of patients or individual cases, it is difficult to predict the prognosis of patients with haemangiopericytoma. The 10-year actuarial survival rate seems to be in the region of 70%.3 The incidence of metastases ranges from 11.7 to 56.5%. The lung and the skeleton are the most common metastatic sites, and lymph-node spread is uncommon. The interval from the time of first diagnosis to the occurrence of metastasis is about four and a half years, with a range from 1 to 14 years.3 Solitary metastatic pulmonary lesions can be resected with good long-term survival.15 There has also been a report of primary pulmonary haemangiopericytoma with metastases in the abdominal wall and also in the mesenterium, presenting 6 months and 12 months after the original resection of the pulmonary primary lesion.19

Conclusion

Haemangiopericytoma is a tumour with a relatively good prognosis but because of the tendency to late,
local or distant recurrence, excisional surgery at the time of diagnosis must be followed by long-term follow-up. Metastatic disease needs aggressive surgery, radiotherapy and chemotherapy, alone or in combination. Long-term survival is thus possible.

References