Short reports

Giant retroperitoneal hemangiopericytoma

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Summary

Hemangiopericytomas are rare vascular tumours that are derived from pericytes. Retroperitoneal hemangiopericytomas are usually bulky but clinically silent when diagnosed because of their slow rate of growth. A 49-year-old man, who presented with only vague symptoms of abdominal fullness for several months, was found on computed tomography to have a huge well-defined mass with areas of low attenuation and well-enhanced septa. The tumour was successfully resected and was confirmed to be a malignant retroperitoneal hemangiopericytoma. It measured 30 cm in the greatest dimension. We are prompted to present this case as it is believed to be the largest tumour reported to date.

Keywords: hemangiopericytoma, retroperitoneal neoplasm, computed tomography

Hemangiopericytoma is a rare vascular tumour representing approximately 1% of all vascular neoplasms.¹ It is derived from pericytes and was first described by Stout and Murray in 1942.² Pericytes are cells with long processes that surround capillaries and serve to change the calibre of the capillary lumen.³ The exact function of pericytes is still unknown, but it has been postulated that these cells have contractile power.⁴ Hemangiopericytomas tend to be large and well encapsulated.⁵ They have been observed in many parts of the body and in a number of viscera but most commonly in the soft tissue of the lower extremities.^{3,6} Approximately 25% of hemangiopericytomas arise from the retroperitoneum and pelvic cavity.⁷ Although the median size of the excised tumours reported is 6.5 cm, the largest malignant hemangiopericytoma recorded was 23 cm in its greatest dimension.⁸ We are prompted to report a giant retroperitoneal hemangiopericytoma of 30 cm in its greatest dimension. To our knowledge, this is the largest tumour to date in the english literature.

Case report

A 49-year-old male presented with progressive abdominal fullness in the past six months which had become worse in the last two weeks. There were no complaints of abdominal pain or other gastrointestinal symptoms. He had had an anterior wall myocardial infarction six years earlier and was under regular cardiac care. On

physical examination, the abdomen appeared globular in shape. It was non-tender, with dullness on percussion. Blood examination was negative. Small bowel series revealed peripherally displaced bowel loops with a central space-occupying lesion (figure 1). Computed tomography (CT) scan of abdomen with intravenous contrast demonstrated a huge lobulated retroperitoneal soft tissue mass with cystic low attenuation zones and intervening enhanced septations (figure 2). On exploratory laparotomy, a huge mass originating from the pancreatic region with adhesion to serosa of the stomach and transverse colon was found. The patient underwent complete excision of the tumour mass with distal pancreatectomy and splenectomy. He was later discharged in stable condition. Gross pathology revealed a large, well-encapsulated tumour mass measuring 30 × 20 × 15 cm and weighing 3750 g (figure 3). The tumour appeared grey in colour, and was soft with an irregular surface. On dissection, there was extensive necrosis and haemorrhage. Microscopically, the tumour showed tightly packed cells around thin-walled vascular channels ranging from capillary-sized vessels to large sinusoidal spaces. The tumour cells had small round-to-oval nuclei and a moderate amount of clear cytoplasm. Spindle cells were also noted in places. The intervening vascular channels usually formed a continuous ramifying vascular network or a 'staghorn' configuration. There were focal haemorrhages, necrosis and a few mitotic figures (1–2 per 10

Figure 1 Small bowel series showing a space-occupying lesion with displacement of the bowel loops.

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Figure 2 Contrast-enhanced CT scan demonstrates a large retroperitoneal tumour with multiple large and small areas of low attenuation corresponding to tumour necrosis and haemorrhage.

Figure 3 Excision demonstrated a large retroperitoneal hemangiopericytoma.

high power fields). The histopathologic picture of the tumour was compatible with malignant hemangiopericytoma (figures 4 & 5). Furthermore, immunohistochemical stains for actin, desmin, S-100, *Ulex europaeus* agglutinin 1 were negative, while stains for vimentin were positive.

Discussion

Hemangiopericytoma is a rare vascular neoplasm, which is believed to arise from the cells surrounding capillaries and postcapillary vessels.² Because the growth rate of the tumour is generally slow, it is often large by the time it is diagnosed, as in our case. As a result, it is usually presented with no symptoms other than occasional pain or abdominal fullness which seems to be related to perineural invasion.⁷⁻⁹ According to the literature, it can be found anywhere in the body but most commonly appears at the lower extremities (34.9%); 24.5% arises from the retroperitoneum and pelvic cavity.⁷ It has an equal sex distribution and occurs at between 4 and 72 years of age.⁴ The median age at diagnosis is 45 years.⁷ Some congenital cases have been reported.^{10,11} The present patient is 49 years old, close to the quoted age and has non-specific complaints. The plain radiographic feature of most hemangiopericytomas, is a well-demarcated soft tissue mass. Calcification is noted only

Figure 4 Photomicrograph of the tumour showing vascular structures lined by a layer of endothelium and surrounded by tumour cells with oval nuclei and a moderate amount of clear cytoplasm. A mitotic figure is presentation (arrow) (original magnification, $\times 100$).

Figure 5 Low power view demonstrated the hemangiopericytoma with massive necrosis (original magnification, $\times 40$).

occasionally in the tumour (less than 1% of cases).^{3,8,12-14} This non-specific finding is of no help in diagnosis and does not allow its differentiation from other soft tissue tumours.¹⁵ Angiography may be of help in defining the anatomy and planning surgical strategy but is not diagnostic.⁸ CT scanning is superior to ultrasound in diagnosis of the retroperitoneal tumour because of delicate cross-sectional anatomy and abundant fat content.^{16,17} Hemangiopericytoma is prominently enhanced in an intravenous contrast CT scan due to its dense vascularity, particularly in the periphery of the lesion, which is helpful for the diagnosis.¹⁸ The mass is usually well circumscribed, lobulated, large in size and has multiple areas of low attenuation due to necrosis, haemorrhage or cystic degeneration.^{5,18} All the above CT findings are presented in our case.

CT presentation of a large, lobulated, soft tissue mass with cystic low attenuation zones and enhancement of solid areas or septations with or without speckled calcifications is sug-

Clinical features
<ul style="list-style-type: none">asymptomatic, but vague abdominal fullnessoccasional pain due to perineural invasion, usually a late manifestation

gestive of but not specific for malignant hemangiopericytoma.¹⁹ A similar appearance on CT scan of the retroperitoneum may be seen in liposarcoma, although liposarcoma is hypovascular and therefore not well enhanced on intravenous contrast CT scan and in addition, it occasionally has CT number indicative of fat. Malignant fibrous histiocytomas may also appear as lobulated, inhomogenous masses with ill-defined margins and inhomogenous contents on intravenous contrast CT scan. Generally, of the CT images of liposarcoma, malignant fibrous histiocytoma and hemangiopericytoma, hemangiopericytoma shows the greatest degree of contrast enhancement.

Angiography can demonstrate the nature of the tumour and its blood supply, however, CT scan plays an extremely important role in demonstrating its size and relationship to adjacent viscera. A definite diagnosis of hemangiopericytoma is made only upon pathological confirmation but the above radiological findings are helpful for the differential diagnosis and treatment planning.

Pathologic diagnosis of this tumour is based on the presence of large numbers of branched, sinusoidal vascular channels surrounded by and enclosed within nests and masses of spindle-shaped cells which can be occasionally ovoid or even round. Silver impregnations can be used to confirm that these cells are outside the basement membrane of the endothelium and hence are pericytes rather than endothelial cells. Distinguishing the tumours as benign or malignant is based on the numbers of mitotic figures, increased cellularity, presence of cellular anaplasia and foci of haemorrhage and necrosis.^{7,20} McMaster *et al* classified the hemangiopericytoma histologically as benign, borderline malignant, and malignant on the basis of microscopic findings such as vascular patterns, shape of pericytes, anaplasia of pericytes, number of mitotic figures and reticulum.⁴ Focal haemorrhage, necrosis and mitotic figure were found in our case which hence was diagnosed as a malignant hemangiopericytoma. Immunohistochemical methods using monoclonal antibodies have been used recently to distinguish hemangiopericytoma and other mesenchymoma, but also they cannot make a definite diagnosis of this rare tumour.²¹

The treatment of choice for hemangiopericytoma is wide surgical excision.^{3,8,9} Because of its extreme vascularity large retroperitoneal hemangiopericytomas have sometimes been considered unresectable due to a high risk of inordinate blood loss. In previous reports, pre-operative vascular embolisation has been found to be an effective adjunct to the

Treatment options

- wide surgical excision
- pre-operative vascular embolisation to minimise blood loss
- postoperative radiation therapy to improve local control rates
- radiation therapy also for unresectable tumours and recurrences
- chemotherapy not proven useful

Summary points

- rare vascular tumour
- clinically silent
- largest tumour to date 30 cm

risky procedure.^{22,23} Adjuvant postoperative radiation therapy can improve local control rates in patients treated initially with surgery,²⁴ but complete remission of hemangiopericytoma from radiation alone is rare.²⁵ For unresectable hemangiopericytomas and recurrent problems, radiation therapy has been used as an alternative treatment.²⁶ Chemotherapy has not been proven useful for the management of resectable hemangiopericytoma^{7,23}; only partial or short-term remission of metastatic tumours has sporadically been reported after chemotherapy.²⁷

Hemangiopericytomas have a uniquely variable range of malignant potential. Ten-year survival rate for benign lesions is approximately 80%, but only 29% for malignant lesions.⁸ Local recurrence predates metastases in more than 50% of reported cases.²⁵ Due to the location of disease, high recurrent rate and possible metastasis, long term periodic CT scans are necessary, even after complete excision of the tumour.¹⁷

In conclusion, hemangiopericytoma should be considered in the differential diagnosis of a well-defined lobular mass arising in the peritoneum or retroperitoneum, which has multiple large and small areas of low attenuation, intervening enhanced septa, with or without speckled calcifications in CT images. CT is helpful in diagnosis and demonstrating the size, location and relationship to adjacent viscera, which is important for planning treatment.

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1 Ayella RJ. Hemangiopericytoma: a case report with arteriographic findings. *Radiology* 1970; **97**: 611–2.

2 Stout AP, Murray MR. Hemangiopericytoma: vascular tumor featuring Zimmermann's pericytes. *Ann Surg* 1942; **116**: 26–33.

3 Binder SC, Wolfe HJ, Deterling RA. Intra-abdominal hemangiopericytoma. *Arch Surg* 1973; **107**: 536–43.

4 McMaster MJ, Soule EH, Ivins JC. Hemangiopericytoma: a clinicopathologic study and long-term follow-up of 60 patients. *Cancer* 1975; **36**: 2232–44.

5 Goldman SM, Davidson AJ. Retroperitoneal and pelvic hemangiopericytoma, clinical, radiologic and pathologic correlation. *Radiology* 1988; **168**: 13–17.

6 Angervall L, Kindblom LG, Nielson JM, Stever B, Svendsen P. Hemangiopericytoma: a clinicopathologic, angiographic and microangiographic study. *Cancer* 1978; **42**: 2412–27.

7 Enzinger FM, Smith BH. Hemangiopericytoma: an analysis of 106 cases. *Hum Pathol* 1976; **7**: 61–82.

8 Morris P, Stahl R, Liriano E, Dardik H. Giant retroperitoneal pelvic hemangiopericytoma. *J Cardiovasc Surg* 1991; **32**: 778–82.

9 Backwinkel KD, Diddams JA. Hemangiopericytoma: report of a case and review of the literature. *Cancer* 1970; **25**: 896–901.

10 Kanffman SL, Stout AP. Hemangiopericytoma in children. *Cancer* 1960; 13: 695–710.

11 Tulenko JF. Congenital hemangiopericytoma: case report. *Plast Reconstr Surg* 1968; 41: 276–7.

12 Kent KH. Hemangiopericytoma. *AJR* 1957; 77: 347–56.

13 Mujahed Z, Vasilas A, Evans JA. Hemangiopericytoma: a report of four cases with a review of the literature. *AJR* 1959; 82: 658–66.

14 Sutton D, Pratt AE. Angiography of hemangiopericytoma. *Clin Radiol* 1967; 18: 324–9.

15 Yaghmai I. Angiographic manifestations of soft tissue and osseous hemangiopericytomas. *Radiology* 1978; 126: 653–9.

16 Stephens DH, Sheedy PF, II, Hattery RR, Williamson B, Jr. Diagnosis and evaluation of retroperitoneal tumor by computed tomography. *AJR* 1977; 129: 395–402.

17 Pinson CW, Remine SG, Fletcher WS, Braasch JW. Long term results with primary retroperitoneal tumors. *Arch Surg* 1989; 124: 1168–73.

18 Lorigan JG, David CL, Evans HL, Wallace S. The clinical and radiologic manifestation of hemangiopericytoma. *AJR* 1989; 153: 345–9.

19 Alpern MB, Thorsen MK, Kellman GM, Pojunas K, Lawson TL. CT appearance of hemangiopericytoma. *J Comput Assist Tomogr* 1986; 10: 264–7.

20 Imachi M, Tsukamoto N, Tsukimori K, et al. Malignant hemangiopericytoma of the omentum presenting as an ovarian tumor. *Gynecol Oncol* 1990; 39: 208–13.

21 Jimenez-Avala M, Diez-Nau MD, Larrad A, et al. Hemangiopericytoma in a male breast. Report of a case with cytologic, histologic and immunochemical studies. *Acta Cytol* 1991; 35: 234–8.

22 Smullens SN, Scatti DJ, Osterholm JL. Preoperative embolization of retroperitoneal hemangiopericytomas as an aid in their removal. *Cancer* 1982; 50: 1870–5.

23 Smith RB, Machleder HI, Rand RW, Bentson J, Tarbas P. Preoperative vascular embolization as an adjunct to successful resection of large retroperitoneal hemangiopericytoma. *J Urol* 1976; 115: 206–8.

24 Staples JJ, Robinson RA, Wen BC, Hussey DH. Hemangiopericytoma – the role of radiotherapy. *Int J Radiat Oncol Biol Phys* 1990; 19: 445–51.

25 Craven JP, Quigley TM, Bolen JW, Raker EJ. Current management and clinical outcome of hemangiopericytomas. *Am J Surg* 1992; 163: 490–3.

26 Munoz AK, Berek JS, Fu YS, Heintz PA. Pelvic hemangiopericytomas: a report of five cases and literature review. *Gynecol Oncol* 1990; 36: 380–2.

27 Wong PP, Yagoda A. Chemotherapy of malignant hemangiopericytoma. *Cancer* 1978; 41: 1256–60.

Carcinoid syndrome due to a malignant somatostatinoma

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Summary
Somatostatinoma is one of the rarest tumours of the endocrine pancreas. Cardinal manifestations of a somatostatinoma include gallstones, mild diabetes mellitus, steatorrhoea, diarrhoea and dyspepsia. Like any other pancreatic islet cell carcinoma, a somatostatinoma may also produce several different hormones such as adrenocorticotrophic hormone, calcitonin, vasoactive intestinal polypeptide, pancreatic polypeptide, gastrin, insulin, and glucagon. In many cases, the clinical picture is dominated by the effect of these other hormones. We present a patient with somatostatinoma in which an immunocytochemical study of the specimens from pancreas and liver showed a weak positive reaction for gastrin besides a strong positive reaction for somatostatin. Interestingly, this patient also showed the signs of carcinoid syndrome which was successfully treated with octreotide.

Keywords: somatostatinoma, carcinoid syndrome, octreotide

Gut endocrine tumours have a low incidence of about 1 in 200 000 population and 60% of these are carcinoids.¹ Somatostatinomas appear to be one of the rarest gut endocrine tumours, and the yearly incidence is estimated to be as low as 1 in 40 million people.² In the majority of the patients, metastatic spread is evident at the time of presentation or shortly thereafter.

Metastatic spread is usually to the liver, with involvement of lymph nodes and contiguous spread also being common. It has been suggested that the expression of the classic triad of symptoms may be more common when liver metastases are present.³ Total tumour resection is the first line of therapy in patients with pancreatic somatostatinoma, while chemotherapy is also frequently used either as the primary mode of therapy in disseminated disease or as adjunctive therapy after surgery.¹

Carcinoid syndrome is a clinical entity which is usually caused by the humoral secretions of carcinoid tumours that originate in the midgut. Lesions other than carcinoid tumours sometimes secrete serotonin and present with symptoms of the carcinoid syndrome (see box 1). In its most complete form, the carcinoid syndrome involves several different organ systems such as the vasomotor, cardiopulmonary and gastrointestinal systems. The cardinal manifestations of this syndrome consist of hepatomegaly, cutaneous flushing, facial telangiectasia, hypotension, diarrhoea, endocardial

Carcinoid syndrome: causes
<ul style="list-style-type: none">• carcinoid tumours• medullary carcinomas of the thyroid• oat-cell carcinomas of the lung• pancreatic islet cell cancers• neuroblastomas• other chromaffin tumours

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