

Case Report: Cardiac Hemangioma

(#2001-7896 ... July 13, 2001)

Rachel L. Tillett, MRCS,¹ Peter M.C. Jiskoot,² Brian E. Glenville, MS, FRCS³

Department of Cardiothoracic Surgery,^{1,3} Department of Histopathology,² St. Mary's Hospital, London W2 1NY, UK

ABSTRACT

A report of an asymptomatic cardiac hemangioma in a middle-aged man is presented. This is followed by a literature review of mediastinal hemangiomas.

INTRODUCTION

Cardiac tumors are rare, with a post-mortem incidence of 0.002-0.33% [Chitwood, 1988]. Approximately three-quarters of these tumors are benign and one-quarter malignant [McAllister 1978]. Hemangiomas account for 5% of benign cardiac neoplasms in adults [McAllister 1978].

CASE REPORT

A 52-year-old South African male had a chest radiograph taken as part of a routine medical examination. The radiograph revealed a mediastinal mass in the area of the left heart border. The patient, who had no other symptoms, had been trying to lose weight and had successfully lost four kilograms.

The only relevant medical history was a laparoscopic Nissen's fundoplication for a symptomatic hiatal hernia, which was performed four months prior to presentation. The patient was an ex-smoker.

Physical examination of the patient revealed no abnormalities of the cardiovascular or respiratory system. Examination of the abdomen showed scarring from the previous laparoscopic surgery.

Further investigations revealed a normal full blood count, urea and electrolytes, with normal liver function and an erythrocyte sedimentation rate of 6 mm/hour. An electrocardiogram showed a sinus rhythm of 60 beats per minute, and an exercise tolerance test was also normal.

A computed tomography scan of the chest, with and without intravenous contrast, showed a 10cm homogeneous mass of soft tissue above the left ventricle [see Figures 1A and 1B, ⊙]. There was no evidence that the previous hiatal hernia had migrated through the diaphragm into the chest.

T1 and T2 weighted magnetic resonance images revealed a 10cm diameter mass lying within the middle mediastinum [see Figure 2, ⊙]. The mass appeared to be entirely contained

within the pericardium, lying immediately lateral to the main pulmonary trunk anterior to the left atrium, and in sagittal and coronal slices almost contiguous with the superior wall of the left ventricle. There was no apparent involvement or distortion of surrounding vessels, cardiac chambers or bronchi. The appearance of the mass was suggestive of an intrinsic tumor of the middle mediastinum, possibly arising from the heart.

Transesophageal echocardiography demonstrated a 6 x 7 cm encapsulated mass located within the pericardium. The mass appeared to be rising from the basal septum and extending laterally and posteriorly. The superior aspect appeared to lie immediately below the aortic valve. A small pericardial effusion was noted. The appearance was also consistent with that of a hydatid cyst.

A subsequent abdominal ultrasound scan revealed a normal liver, with no evidence of intra-abdominal cysts.

Hydatid serology, using enzyme-linked immunosorbent assay, was negative. However, because false negative results are not uncommon, hydatid infection could not be excluded and treatment with albendazole was therefore commenced.

To obtain a histological diagnosis, the mass was surgically explored. A large mass lying along the right ventricular outflow tract, measuring 10 x 8 x 6 cms, was found. Aspiration yielded 150†ml of dark blood, suggesting that the mass was very unlikely to be a hydatid cyst. However, hypertonic saline was injected into the mass to ensure that if any cysts or tumor cells were spilt, they would be less likely to cause secondary deposits. The operation was performed on cardiopulmonary bypass, using cold blood cardioplegic arrest. The lesion was incised and noted to have a trabeculated appearance. Two-thirds of the length of the left anterior descending artery was seen to be running through the mass. The lesion was therefore only partially excised, and the residual mass plicated. Complete removal would have involved making a new right ventricular outflow tract and half a left ventricle.

The resected material comprised four fragments with a total weight of 15 grams. It was covered by a smooth, gray membrane overlying light brown, spongy, multicystic tissue. Microscopic examination showed epicardial tissue at the surface consisting of a fibrous membrane lined by mesothelium [Figure 3, ⊙]. The deeper tissue consisted of many cavernous angiomatous spaces, with some blood in their lumina. The spaces were lined by flattened cells that immunohistochemically expressed Factor VIII-related antigen (von Willebrand factor) and CD31, but not epithelial membrane antigen or cytokeratin. This identified them as endothelial cells and not as mesothelial cells. Between the cavernous spaces there was connective tissue with randomly scattered bundles of altered-

Submitted July 11, 2001; accepted July 13, 2001.

Address correspondence and reprint requests to: Mr. Brian Glenville, MS, FRCS, Consultant Cardiothoracic Surgeon, 66 Harley St., London, W1G 7HD, UK. Email: bg@66harley.com

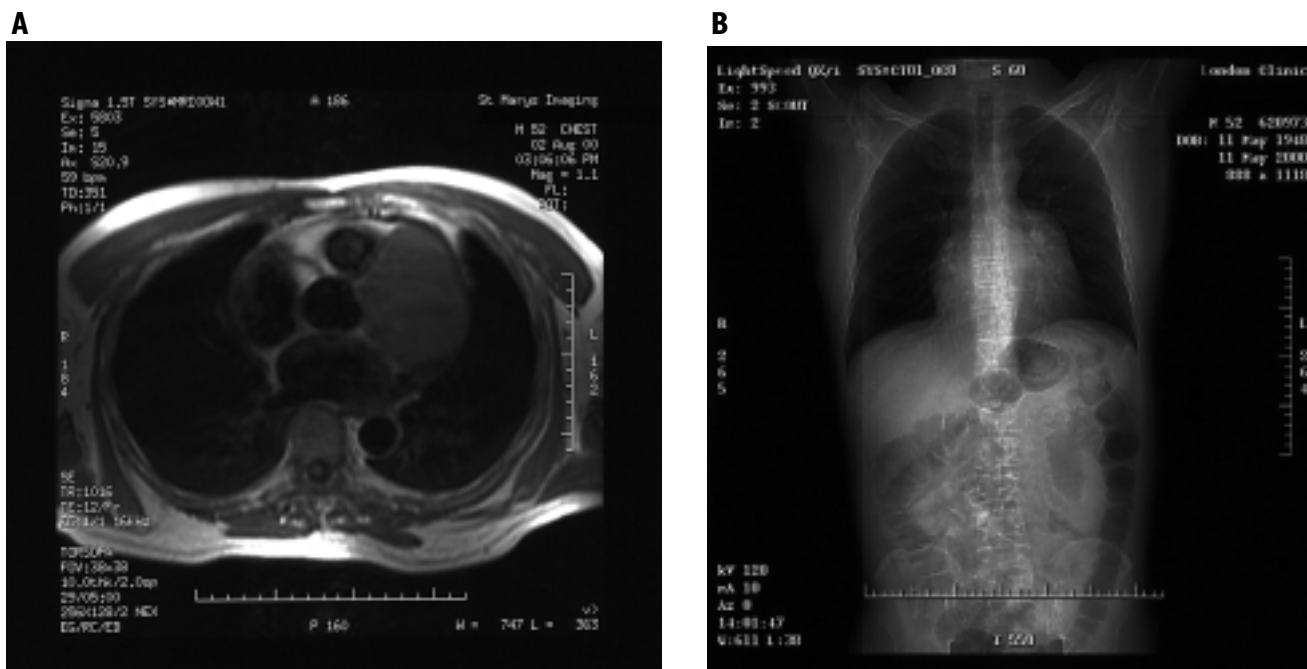


Figure 1. CT scan demonstrating the haemangioma

smooth muscle actin positive cells, lymphoid aggregates, arteries with thickened walls and narrowed lumina, adipose tissue, and nerve branches. However, no cardiac muscle was observed. There were also regressive changes, including areas of sclerosis and collections of hemosiderin-laden macrophages.

The preceding evidence indicates that the lesion was an epicardial hemangioma with features of both the cavernous and arteriovenous type.

The patient had an uneventful postoperative recovery and is very well four months after surgery.

DISCUSSION

Hemangiomas can be defined as benign, nonreactive processes in which there are an increased number of normal or abnormal blood vessels. The distinction between vascular malformations, vascular hamartomas, and hemangiomas is, however, vague [Enzinger 1995]. Hemangiomas of the heart are very rare: they account for 5% of benign cardiac tumors and represent 1.7% of all cardiac tumors [McAlister 1978].

These tumors occur in patients of all ages. Mediastinal hemangiomas have been recorded in a baby at one week post-term [Cohen 1987] and in a 76-year-old man [Ceccanti 1989]. However, they appear to be found most frequently in young adults and have an equal distribution between sexes [Davis 1978].

Hemangiomas of the mediastinum can present in a variety of ways. Many earlier cases were discovered in the post-mortem room. However, as radiological techniques and expertise have improved, more are being diagnosed and treated during life. Some tumors are asymptomatic and are discovered accidentally on routine chest radiographs [Toch 1965, Davis 1978, Grenadier 1989, Ishii 1990, Zalcmann 1990, Lineau 1995]. Others are symptomatic and their clinical features are often influenced by their location within the heart and mediastinum. Intramyocardial tumors may be silent.

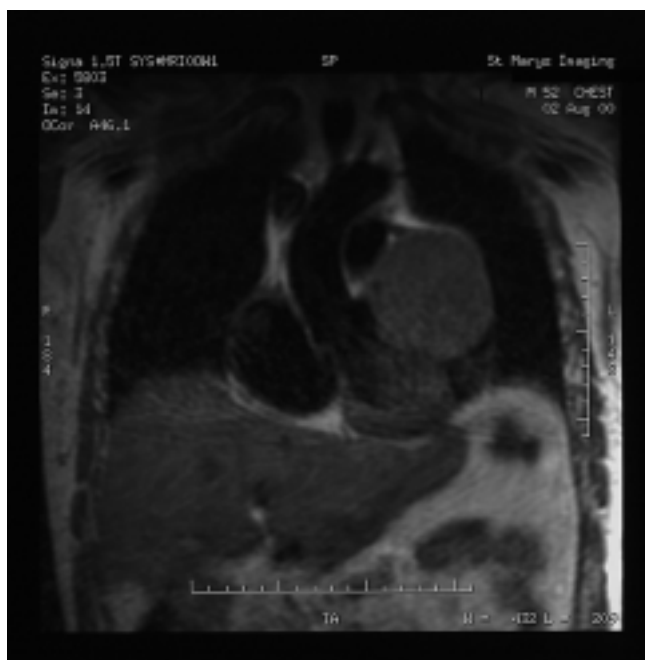


Figure 2. MRI scan demonstrating the haemangioma.

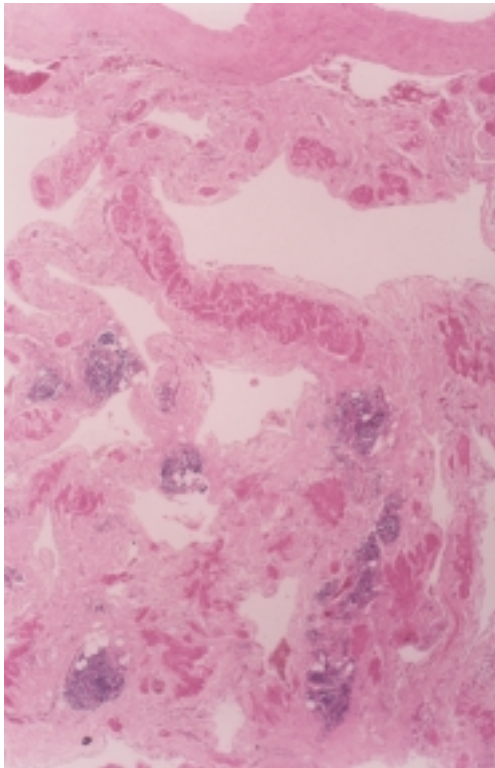


Figure 3. Epicardial hemangioma. At the top is shown the fibrous membrane lined by mesothelium. The tissue below shows cavernous hemangiomatous spaces with focally some blood in their lumina. The intervening connective tissue comprises bundles of smooth muscle cells and lymphoid aggregates. (Hematoxylin and Eosin used as tissue stain, original magnification: $\times 20$).

However, they can present with devastating arrhythmias or heart blockage if their mass obstructs the heart's conducting system [Burke 1990]. By projecting into a cardiac cavity, hemangiomas are capable of causing obstruction of blood flow and therefore mimicking mitral or pulmonary stenosis [Scully 1983]. If the tumor is lying within the pericardium, the patient may develop pericarditis, pericardial effusions [Ha 2000], or tamponade [Scully 1983, Burke 1990]. Patients may also develop systemic signs and symptoms such as fever [Takahashi 1993], weight loss, raised erythrocyte sedimentation rate, and elevated white cell count.

Hemangiomas can occur anywhere within the heart and mediastinum. Reports of hemangiomas affecting the right and left ventricles, right and left atria, and both the interventricular and interatrial septa exist [Abad 1990, Burke 1990]. However, they occur more commonly in ventricles than in atria. Tumors have been found in the superior, posterior, and anterior mediastinum. Those in the anterior mediastinum are frequently associated with the thymus [Ishii 1990, Zalcman 1990, Takahashi 1993]. One tumor seemed to arise from the sympathetic chain [Parker 1997].

Histologically, hemangiomas can be broadly classified into three types. Cavernous hemangiomas are composed of multi-

ple, dilated, thin-walled vessels. Capillary hemangiomas consist of very small vessels, while arteriovenous hemangiomas are made up of dysplastic, malformed arteries and veins. Cardiac hemangiomas containing papillary components have also been described. [Abad 1990]. The histological picture of the hemangioma in this case, with scattered bundles of smooth muscle and lymphoid aggregates, appears to be uncommon.

Diagnosing cardiac hemangiomas often involves several modes of radiological imaging. Chest radiographs are one method of detecting the tumor, and mediastinal masses may be seen readily by that means. All mediastinal masses warrant CT examination [Schurawitzki 1991]. Cohen et al. [Cohen 1987] state that CT did not aid in the preoperative diagnosis of hemangioma of any patient, but it successfully evaluated the extent of the tumor and invasion of adjacent structures. Takahashi et al. [Takahashi 1993] suggest that in thrombosed hemangiomas, areas of clot are not enhanced by contrast medium, and it would be difficult to differentiate a thrombosed hemangioma from cystic lesions. However, magnetic resonance imaging is better at characterizing clot formation due to the characteristic features of methemoglobin formation in clots.

Treatment of hemangiomas usually involves thoracotomy and resection of the lesion. For tumors involving important structures, incomplete resection may be inevitable [Guvener 1999]. Resection allows a histological diagnosis, reduction of tumor mass and improvement of clinical symptoms. In most cases reviewed, resection has been successfully performed.

In one case [Chalet 1993], after a hemangioma was excised an angiosarcoma developed at the same site. Despite this incident, prognosis is generally very good and in most patients the tumors do not recur.

REFERENCES

1. Abad C, Campo E, Estruch R, et al. Cardiac hemangioma with papillary endothelial hyperplasia: report of a resected case and review of the literature. *Ann Thorac Surg* 49:305-8, 1990.
2. Burke A, Johns JP, Virmani R. Hemangiomas of the heart: a clinicopathologic study of ten cases. *Am J Cardiovasc Path* 3(4):283-90, 1990.
3. Ceccanti J, Chauvin G, Guendon R et al. Hemangiome Tumoral Malformatif Geant Du Mediastin A Propos D'une Observation. *Ann Chir: Chir Thorac Cardio-Vasc* 43 (2):157-60, 1989.
4. Chalet Y, Mace L, Frac B, et al. Angiosarcoma 7 years after surgical excision of histiocytoid hemangioma in the left atrium [Letter]. *Lancet* 341:1217, 1993.
5. Chitwood WR. Cardiac neoplasms: current diagnosis, pathology and therapy. *J Card Surg* 3:119-54, 1988.
6. Cohen AJ, Sbaschnig RJ, Hochholzer L, et al. Mediastinal hemangiomas. *Ann Thorac Surg* 43:656-9, 1987.
7. Davis JM, Mark GJ, Greene R. Benign blood vascular tumors of the mediastinum. Report of four cases and review of the literature. *Diag Radiol* 126:581-7, 1978.
8. Enzinger FM, Weiss SW. Benign tumors and tumor-like lesions of blood vessels. In: Enzinger FM, Weiss SW, eds., *Soft tissue tumors*. St. Louis: CV Mosby, 579-626, 1995.

9. Grenadier E, Margulis T, Palant A, et al. Huge cavernous hemangioma of the heart: a completely evaluated case report and review of the literature. *Am Heart* 117 (2):479-81, 1989.
10. Guvener M, Dogan R, Demircin M, et al. Vascular hamartoma of the mediastinum. *Turk J Pediatr* 41:133-7, 1999.
11. Ishii K, Maeda K, Hashihara M, et al. MRI of mediastinal cavernous hemangioma. *Pediatr Radiol* 20:556-7, 1990.
12. Ha JW, Lee MH, Chang BC, et al. Cardiac hemangioma. *Clin Cardiol* 23:53-4, 2000.
13. Lineau C, Quinquenel ML, Maugendre S, et al. Hemangiome caveux intrathoracique. A propos d'un cas. *Rev Mal Resp* 12:625-7, 1995.
14. McAllister HA Jr, Fenoglio JJ Jr. Tumors of the cardiovascular system. In: *Atlas of tumor pathology*. Washington, DC, Armed Forces Institute of Pathology, 1978.
15. Parker JR, Knott-Craig C, Min KW, et al. Cellular hemangioma of the posterior mediastinum. Unusual presentation of a rare vascular neoplasm. *J Okla State Med Assoc* 90:7-9, 1997.
16. Schurawitzki H, Stiglbauer R, Klepetko W, et al. CT and MRI in benign mediastinal hemangioma. *Clin Radiol* 43:91-4, 1991.
17. Scully RE, Mark EJ, McNeely BU. Case records of the Massachusetts General Hospital. Presentation of case. *NEJM* 308(4):206-14, 1983.
18. Takahashi M, Murata K, Mori M, et al. Thrombosed cavernous hemangioma of the anterior mediastinum in a five year old boy: CT and MR appearances. *Radiation Med* 11(2):66-68, 1993.
19. Toch H, Hagstrom JWC, Steinberg I. Hemangioma of the mediastinum. Report of a case with compression of the spinal cord. *Am J Roent Genol* 94(3):580-3, 1965.
20. Zalcman G, Jancovici R, Paraf F, et al. Une tumeur rare du mediastin: l'hémangiome benin. *Rev Pneumol Clin* 46:31-34, 1990.

REVIEW AND COMMENTARY

1. Editorial Board Member KE221 writes:

There is too much histological detail for a lesion that the average heart surgeon may never see, or at most will see once in a career. The histology is well documented in the literature. It would be sufficient to say that the lesion had histology typical of a benign hemangioma.

Authors Response by Rachel L. Tillett, MRCS:

This tumor is an unusual type of hemangioma and therefore a discussion of its histological components is important and warranted. We did not find that this was an area well covered by the literature.