ABSTRACT

Background: The mediastinum is a highly uncommon site for hemangiopericytoma, a rare tumour arising from the pericytes of Zimmmernann in the outer wall of capillaries. Histopathology and immunohistochemistry are two primary diagnostic tools which differentiate these from synovial sarcoma and solitary fibrous histiocytoma. Surgery is the mainstay of treatment for this tumour with variable malignant potential; adjuvant therapy does not help much. Case Summary: We report a case of primary mediastinal hemangiopericytoma in a 15-year-old lady which recurred after complete surgical excision and a second surgical operation resulted in recurrence again. The first histopathological report after removal of the tumour, revealed a primary cystic hemangiopericytoma, histopathology of the subsequently resected tumour established malignant hemangiopericytoma. Conclusion: Hemangiopericytomas are potentially malignant tumours with high recurrence rate and are very difficult to cure.

KEYWORDS: Hemangiopericytoma, Mediastinal mass, Cystic mediastinal tumour

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Introduction

The pericytes of Zimmermann are modified smooth muscle cells with long processes, which wrap around capillaries to change its calibres, give rise to hemangiopericytoma [1]. It is a rare tumour of vascular origin, uncommonly found in musculoskeletal system and skin, rarely found in the mediastinum [2]. The reported cases of primary mediastinal hemangiopericytoma in the literature are quite scanty, and presentation as the mediastinal cyst is rare [3]. Hemangiopericytomas are potentially malignant tumours with high recurrence rate and are very difficult to cure and are now included in the group of the solitary fibrous tumour, a single spectrum of mesenchymal tumours, of which hemangiopericytoma is a cellular phenotypic variant. The criteria for definitive malignancy are difficult to establish, though increased tumour size, mitotic count, cellularity, the presence of haemorrhage/necrosis, nuclear polymorphism and presence of sharply demarcated anaplastic/poorly differentiated foci give a high degree of suspicion of malignancy[4]. We present a case of primary cystic mediastinal hemangiopericytoma and discuss the clinical symptoms, the diagnosis, the correlation of radiographic features and difficulties in management for its high recurrence rate. The first histopathological report after removal of the tumour, revealed a primary cystic hemangiopericytoma, histopathology of the subsequently resected tumour established malignant hemangiopericytoma.
A 15-year-old female complaining of shortness of breath, cough, and low-grade fever for six months was admitted to our hospital. Chest radiography on admission revealed a large mass occupying the mediastinum and left thoracic cavity (Fig-1). Computed tomography (CT) with contrast enhancement showed a giant mass of approximately 10X15X15 cm in size. It compressed the heart, descending thoracic aorta with the left lung atelectasis and occupying almost the whole left thoracic cavity (Fig-2, 3, 4). The lesion was well demarcated and cystic on radiological appearance. Biomarkers for cancer including AFP, CEA, CA 19.9, CA 125 and Beta-HCG were normal. We decided to extirpate the lesion by extended left posterolateral thoracotomy. The patient underwent thoracotomy on 3 April 2015; the giant cystic mass was located in the left hemithorax and adherent to the left side of the pericardium. There were extensive adhesions between lesions and the pericardium, as well as with the compressed the left lung. Dissection was performed with great care. For better operative view we punctured the lesions and aspirated its fluid contents (Fig-5). This resulted in a reduction in the size of the lesion, which was very helpful for dissection around the lesions and complete resection was successful without any difficulty despite its size and adhesions. The left lung was fully expanded after removal of lesions. Resected lesion was sent for pathological examination. Pathologic findings revealed abundant capillaries and small vessels with a single layer of endothelium, around which there were tightly packed tumour cells, and the diagnosis was a primary cystic hemangiopericytoma (Fig-6). The postoperative period was uneventful, and the patient was discharged home on 10th postoperative days.

Follow-up at one month, three months revealed no abnormality, her chest X-rays were normal. The patient did not report at six months as she lived in a remote area. Her symptoms recurred at 9th month; Chest X-ray revealed tumour recurrence (Fig-7,8). CT scan of the chest with contrast enhancement was done on 26 May 2016, which showed a recurrent tumour of 8.63x 9.28x 8.5cm dimension, lobulated, cystic with the thick irregular wall, pushing the left hemidiaphragm below and compressing the left lung upward. She was again operated on 13 June 2016.
This time it was a complicated operation, required six units of whole blood transfusion, four units of freshly frozen plasma transfusion, the tumour could not be removed completely, and few densely adherent pieces were left behind. Histopathological examination revealed malignant hemangiopericytoma.

Immunohistochemical analysis showed that the tumour was positive for CD34, anti-smooth muscle actin, type IV collagen and vimentin and negative for S-100 protein and cytokeratins. These findings confirmed the diagnosis of hemangiopericytoma. We could not remove the chest tube for 14 days; the patient was discharged home on the 20th postoperative day with advice to take chemotherapy and radiotherapy. The patient did not take that. In a phone interview on 25 January 2016, the patient said that she was bed-ridden and hoped only best supportive care.

Discussion

Following its first description by Schmidt in 1937 and naming by Stout and Murray in 1942, Hemangiopericytoma has been reported on various sites, most commonly in the thigh, head and neck regions, but mediastinal hemangiopericytoma in general, and its cystic variety, in particular, remains a rarity [1]. Hemangiopericytomas are now included in the broad group of the solitary fibrous tumour, which are predominantly patternless tumours having pleomorphic spindle cells admixed with collagen, whereas its cellular variant hemangiopericytomas are composed of ovoid, monomorphic cells with thin walled anastomosing vessels. However, if a different part of the same tumours is examined histologically, both patterns can be seen in the same tumour, as both being CD34 reactive, hemangiopericytomas are now included within the spectrum of solitary fibrous tumours though representing a cellular variant[4]. Our patient had a history of shortness of breath, cough and low-grade fever. However, benign tumours may be asymptomatic, detected incidentally on chest radiograph but may present with hemothorax due to spontaneous rupture [5]. This locally aggressive and potentially malignant tumour, frequently recur after excision, metastasize and have no predilection for age and sex, and constitute only 6% of the primary mediastinal tumours [6,7].

On computed tomography (CT) with contrast enhancement, the findings in our patient were of a mass which was well defined, 10x15x10 cm, occupying two-third of the left hemithorax pushing the mediastinum to the right and compressing the left lung upward. The smooth interface with the adjacent lung and a broad base on the mediastinum indicated the origin of the tumour in the mediastinum rather than the lung. The central low-density attenuation and peripheral irregular lining in some images and contrast enhancement showed a vascularized and potentially malignant tumour. There was no calcification. FNAC was not attempted because of the fear of haemorrhage. The radiographic features were consistent with primary mediastinal hemangiopericytoma though the curvilinear encircling structure suggesting a large feeder vessel was not found [3]. Some investigators have advised preoperative embolization to reduce vascularity [8], others have advised preoperative aspiration for immediate relief of symptoms [8] but careful intraoperative aspiration and sometimes suction evacuation help in reducing the size of the tumor with minimum chance of spillage but facilitates dissection around the tumor, reduces blood loss while separating vascular adhesion around it. Arrhythmia particularly bradycardia is common during separation from mediastinum necessitating careful monitoring and requiring atropine and inotrope agent administration.

Microscopically, hemangiopericytomas consist of tightly packed tumour cells around thin-walled, single layered endothelium-lined vascular channels ranging from capillary-
Fig. 7,8. CT scan of the chest with contrast enhancement revealed a recurrent tumor of 8.63x 9.28x 8.5cm dimension, lobulated, cystic with thick irregular wall, pushing the left hemidiaphragm below and compressing the left lung upward.

sized vessels to large gaping sinusoidal spaces. The rounded pericytes are paler and have vacuolated cytoplasm whereas the endothelial cells are deeply stained and our findings are completely consistent with that [1]. Though MRI scan, angiography, contrast-enhanced CT scan gives a clue to the vascular origin of the tumour, the definitive diagnosis can be established only after a histological diagnosis of excised tumour[10]. Wide excision is the mainstay of treatment, preoperative embolization may reduce vascularity of the tumour, and adjuvant chemotherapy and radiotherapy are usually not rewarding [6]. Local recurrence or eventual distant metastasis are quite common even after complete excision of the tumour, approximately 80% of which turn out as a malignant tumour. The overall recurrence rate of hemangiopericytomas originating in any organ system is approximately 50%. However, primary mediastinal hemangiopericytomas are distinguished by early recurrence-about 40% recurring within one year of resection [3]. So, long-term follow-up and frequently second look surgery is needed to prolong life expectancy.

Conclusion
Hemangiopericytoma is a potentially malignant vascular tumour with high incidence local recurrence in spite of adequate surgical resection. Since surgery is the mainstay of treatment, a low threshold should be maintained for multiple attempts to excirpate the tumour keeping adequate hemodynamic monitoring and management protocol at hand.

Authors’ Statements

Competing Interests

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. There were no financial support or relationships between the authors and any organization or professional bodies that could pose any conflict of interests.

Authors' contributions
RH, SB, DG, MN, SBH took part in the first operation. RH, MA, MRK took part in the second operation. RH led the operation in both times. MC did the pathological examination. All contributed to literature search and preparation of the manuscript. All authors approved the final manuscript.

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Written informed consent of the patient and her guardian was taken for publication of this case report.

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