Large Intra-Pericardial Sarcoma Arising From Ascending Aorta- A Case Report

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Citation

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Abstract

A young male of 18 years admitted with a 1-week history of retrosternal discomfort, cough and exertional breathlessness and a 3-month history of anorexia and weight loss was found to have a large sarcoma arising from the intrapericardial ascending aorta. Surgery was performed by median sternotomy with complete excision. Histo-pathological examination revealed features of sarcoma. Intra-pericardial tumors arising from the ascending aorta are very rare and only five cases have been reported so far.

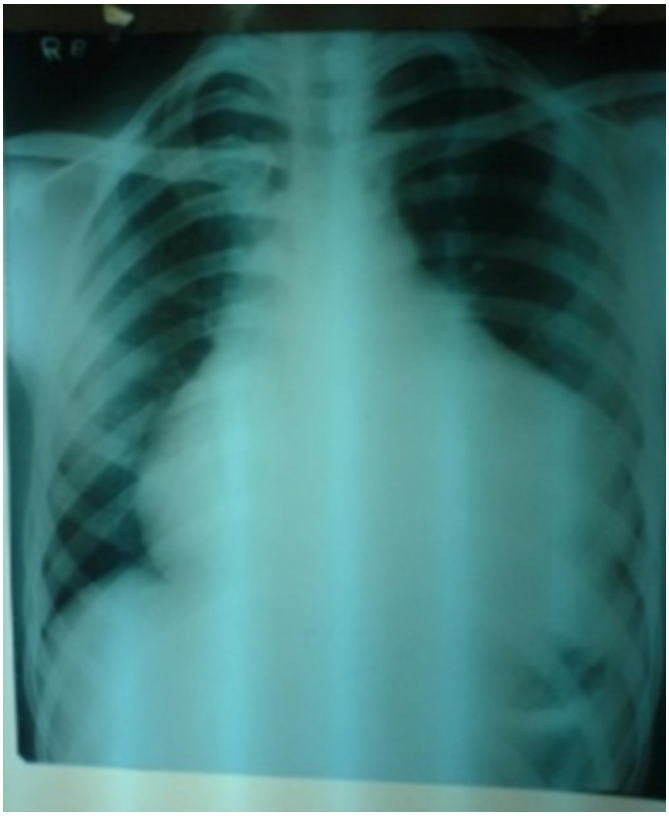
INTRODUCTION

Primary tumors of the aorta are very rare and correct preoperative diagnosis is uncommon. There are only 35 to 40 cases of primary aortic tumors reported in the literature so far ([1],[2]), most of them seen in the descending thoracic and abdominal aorta ([3]). Most of these are malignant and the most frequent histologic type is malignant fibrous histiocytoma ([4]). We present an unusual intra-pericardial tumor arising from the adventitia of the ascending aorta within the pericardium. The tumor was a sarcoma of spindle-cell morphology. The tumor was excised through a median sternotomy without cardio-pulmonary bypass.

CASE REPORT

An 18-year-old male was referred to us with a provisional diagnosis of massive pericardial effusion with a pericardial cyst. The patient had presented with a 1-week history of exertional breathlessness, retrosternal discomfort & cough. There was a history of gradual weight loss and anorexia for the last 3 month. On physical examination, he appeared well. His pulse rate was 90/min and his blood pressure 110/70mmHg. There was no pulsus paradoxus and the JVP was no raised. On auscultation, the heart sounds were masked in intensity without any murmurs. Chest x-ray showed an enlarged cardiac shadow with normal lung fields (Fig. 1).

Figure 1



Routine investigations were normal. ECG showed sinus rhythm with low-voltage complexes. Echocardiography revealed features of massive pericardial effusion with no features of tamponade and normal LV functions. A large cystic structure was seen within the pericardial cavity just lateral to the free right atrial wall. Cardiac MRI revealed

features of a large pericardial mass attached to parietal pericardium, with features consistent with pericardial tumor (Fig. 2 &3).

Figure 2

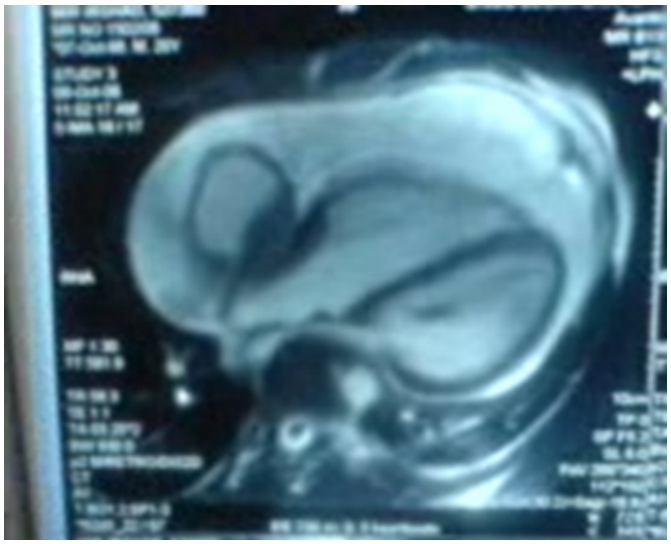
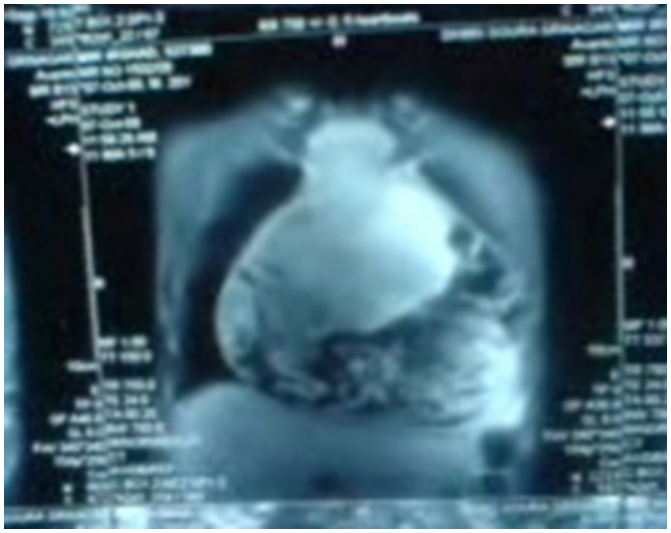


Figure 3



The heart was approached through median sternotomy. Peroperatively, a large, soft, friable and pale tumor with smooth surface was found in the pericardium. The tumor completely obscured the view of the heart. Flimsy adhesions were present between tumor and pericardium. The tumor was arising from the ascending aorta near to the right pulmonary artery fold. The tumor was engulfing all great vessels, right atrium, right ventricle and left ventricle. A cystic cavity was found within the tumor near the right atrium measuring 5 x 5cm. A dissection plane was created between tumor and ascending aorta which was carried down to over the right pulmonary artery, superior vena cava, right atrium, right ventricle and left ventricle. The tumor was seen to be arising from the adventitia of the ascending aorta involving the media by a thin stalk which was resected in continuity with the tumor and the defect was closed by purse-string suture using 3-0 prolene. The post-operative period was uneventful.

The tumor was sent for histopathological examination. Grossly, the tumor was measuring 15 x 15 x10cm and weighed 750g. Microscopically, it showed features of soft-tissue sarcoma of spindle-cell morphology.

COMMENT

The tumor was a challenge in terms of diagnosis. The clinical presentation of this tumor was unusual because of lack of signs of compression of the heart and the great vessels. It is remarkable that a tumor of such a dimension could not result in features of cardiac tamponade. Echocardiographic findings revealed massive pericardial effusion which was contrary to MRI and operative findings and also it could not be determined clearly whether the tumor is arising from pericardium, heart or great vessels.

Tumors of the ascending aorta are rare. Reviewing the literature, we found a number of cases of tumors of the aorta. So far, only five cases of intra-pericardial tumors arising from the ascending aorta have been reported in the literature: one is a spindle-cell sarcoma ($_5$); one is a malignant fibrous histiocytoma ($_2$); one is a lipoma ($_3$); one is ectopic thyroid & the last is a teratoma ($_6$). As such, this is the second spindle-cell sarcoma of the ascending aorta being reported. All other aortic tumors reported were located in the descending thoracic or abdominal aorta and the majority of them were sarcomas. There were a few case reports of aortic sarcomas that developed after replacement of the descending thoracic or abdominal aorta with vascular prosthetic grafts ($_4$).

There was technical difficulty in removing the tumor because of large size, but we successfully removed the whole tumor without the use of cardiopulmonary bypass. Although outlook in these patients remains guarded, the patient needs regular follow-up for detection of early recurrence.

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