VASCULAR HAMARTOMA OF THE MEDIASTINUM*

A Case Report

*Murat Güvener MD***, Rıza Doğan MD***, Metin Demircin MD****
*Can İlyas MD*****, İlhan Paşaoğlu MD****

SUMMARY: Güvener M, Doğan R, Demircin M, İlyas C, Paşaoğlu İ. (Departments of Thoracic and Cardiovascular Surgery and Pathology, Hacettepe University, Faculty of Medicine, Ankara, Turkey). Vascular hamartoma of the mediastinum: a case report. Turk J Pediatr 1999; 41: 133-137.

Vascular hamartoma of the mediastinum is a rare benign vascular tumor. A 13-year-old girl presented with back pain, persistent coughing, palpitation, and angina pectoris. Preoperative investigations demonstrated an enlarging mass involving the superior mediastinum extending posteriorly (T6-T8). An encapsulated, 6x5x3 cm dark purplish mass adherent to the aortic wall was found. The main mediastinal mass was totally excised but limited resection was carried out in the paravertebral region. Microscopic examination revealed a vascular hamartoma. *Key words: hemangioma, mediastinum, vascular hamartoma.*

Vascular hamartomas have been reported as a rare pathological entity. They consist of hemangiomas that fall in a gray area between hamartomatous malformations and true neoplasms. Hemangiomas are bingn vascular tumors and are rare developmental vascular hamartomas that, by definition, do not group by mitotic activity[1, 2]. They occur more commonly in women between 30 and 50 years[2] of age[1]. Localization of vascular hamartomas can involve different parts of the body such as mediastinal, retinal, dermal, epidural, and testicular tissue[3-6].

Various vascular tumors, both benign and malignant, occur in the mediastinum. Many mediastinal tumors in infants and children are vascular. Only one-third of 38 mediastinal vascular tumors reviewed by Shields, Attar and Cowley[7, 8]. (1964) were found in persons younger than 20 years. The other two-thirds occurred in older persons mostly in their fourth and fifth decades of life. The incidence in men and women is approximately equal.

The tumor is generally solid and discrete, although diffuse infiltrative lesions have been reported. Calcification (phlebolith) occasionally occurs. Bony erosion or enlargement of an intervertebral foramen may also occur.

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* From the Departments of Thoracic and Cardiovascular Surgery and Pathology, Hacettepe University Faculty of Medicine, Ankara.

** Specialist in Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine.

*** Associate Professor of Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine.

**** Professor of Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine.

***** Research Assistant in Pathology, Hacettepe University Faculty of Medicine.

Although various histological types are seen, most of these tumors are benign. The benign lesions have been classified as hemangioendotheliomas or capillary hemangiomas. The malignant lesions are designated as angiosarcomas or hemangiosarcomas. Although malignant lesions occur in all age groups, they are most common in the fourth decade of life.

A case of vascular hamartoma involving the superior and posterior mediastinum is here in reported.

Case Report

A 13-year-old girl was admitted to our hospital with complaints of back pain in the subscapular region, persistent coughing, palpitation, and angina pectoris. The severity of pain increased when lying or coughing. Routine physical examination was normal. The chest roentgenogram revealed a double contour in the aortic arch (Fig. 1). Echocardiographic examination suggested aortic aneurysm. On angiocardiographic examination no true aortic aneurysm was found. Magnetic resonance imaging (MRI) examination of the lesion revealed a round mass extending along the aortic wall from the superior mediastinum posteriorly to the sixth and eighth thoracic vertebral bodies; septation was present in the solid hemorrhagic mass (Figs. 2a, 2b). The abdominal ultrasonography (USG) was normal. Complete blood count, biochemical analysis and electrocardiogram were found within normal limits.

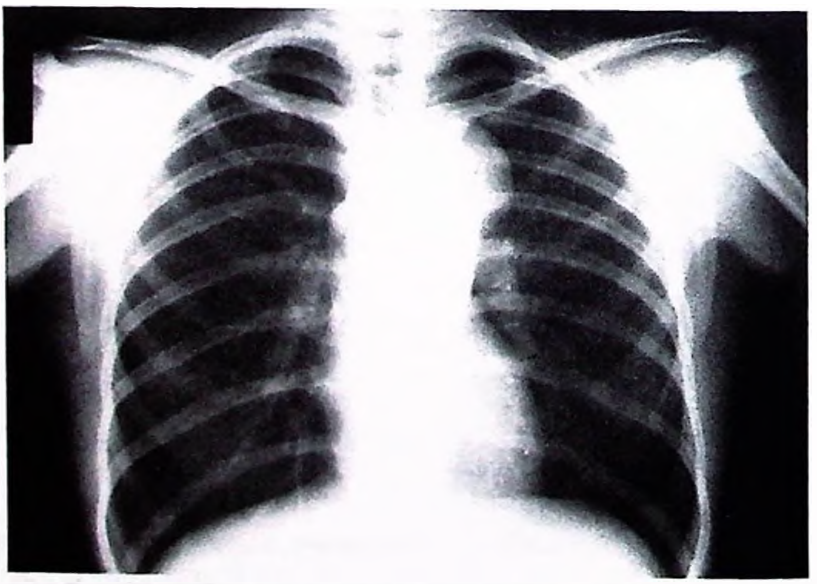
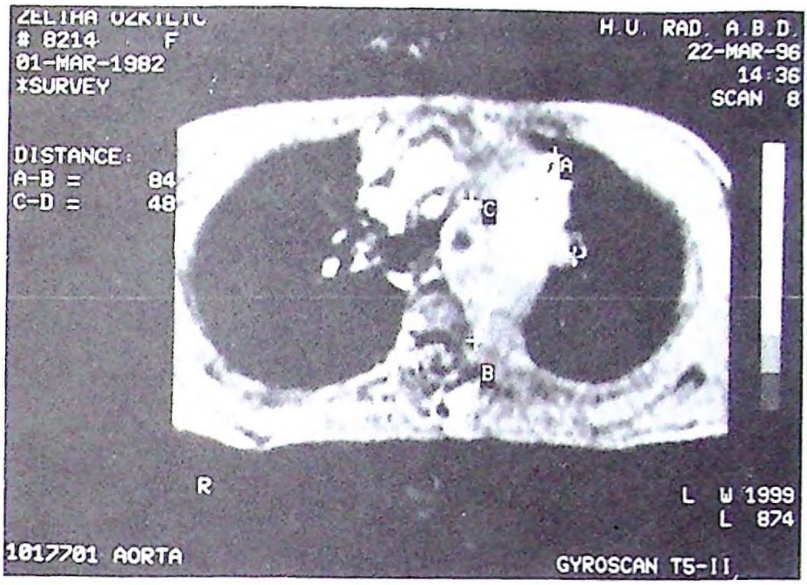
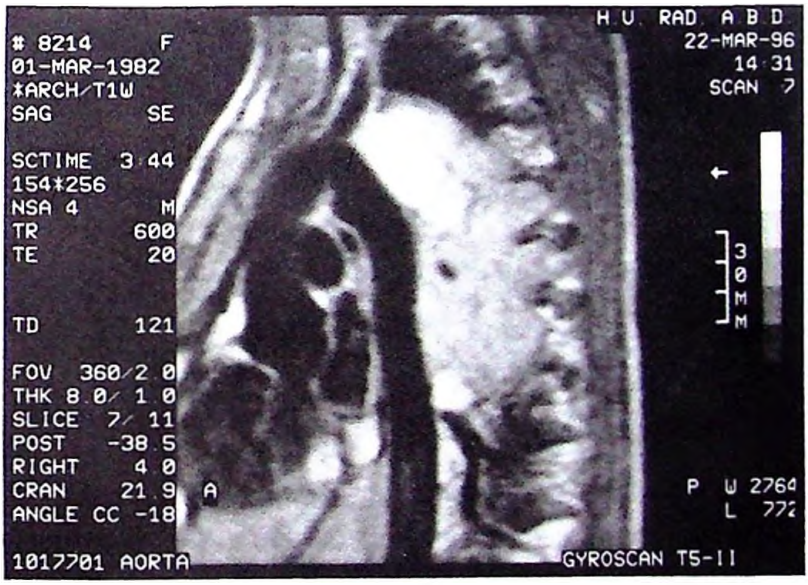


Fig. 1: Preoperative posteroanterior chest roentgenogram showing a double counter in the aortic arch.



(a)



(b)

Fig. 2: Preoperative MRI examination. (a) Transverse section showing a round mass extending posteriorly along the aortic wall from the superior mediastinum. (b) Sagittal section showing the relation between the mass and paravertebral region.

Left posterolateral thoracotomy was performed. A 6x5x3 cm dark purplish encapsulated mass adherent to the aortic wall was found. On exploration, the tail of the mass was found to be extending to the paravertebral region (T6-T8). The main mediastinal mass was totally excised but limited resection was performed in the paravertebral region.

Microscopic examination revealed a vascular hamartoma characterized by an irregularly lobulated cystic structure with coagulated blood in the cysts (Fig. 3). Postoperative irradiation was not applied. Postoperative course was uneventful and the patient was discharged on the sixth postoperative day.

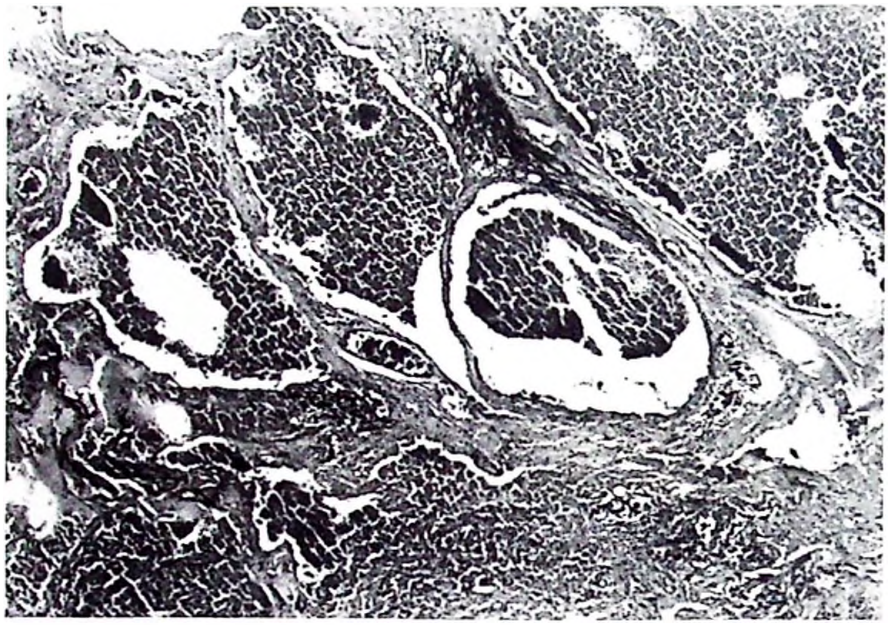


Fig. 3: Histological sections of the mass. The lesion is composed of large dilated blood-filled vessels lined by flattened endothelium (hematoxylin-eosin; original magnificantion x 100).

In the control MRI examination on the 24th day, the appearance of the excision area was evaluated as a hematoma. In the follow-up examinations, six months after the operation, the patient was well.

Discussion

Vascular hamartoma of the mediastinum is a benign tumor rarely found in the mediastinum. Thirty-eight mediastinal vascular tumors (benign or malignant) were reviewed by Shields, Attar and Cowley (1964)[7,8], and 18 benign mediastinal hemangiomas were reported by Moran and Suster (1995)[9]. The others in the literature are case reports[10,11].

Most vascular masses occur in the anterior mediastinum and adjacent visceral compartments. The remaining ones are present in the posterior aspect of the visceral compartment and paravertebral sulci. In our case the mass in the superior mediastinum was extending like a tail to the paravertebral sulci. Approximately 30 percent of these masses produce no symptoms; large and malignant lesions are more likely to be symptomatic. In this case, back pain, angina pectoris and coughing were the major symptoms. Concurrent vascular

masses may be present in the neck or elsewhere; however, in our patient there were not any accompanying lesions. Magnetic resonance imaging (MRI) is very useful in establishing a diagnosis of mediastinal vascular hamartomas[12].

At thoracotomy, the vascular masses are often soft, encapsulated, and dark purplish in color, and may or may not be pulsatile[7,8]. Both benign and malignant tumors may infiltrate adjacent structures[7,8]. In our patient, the mass had infiltrated the aortic adventitia and the epidural layer of the spinal cord. The choice of surgical treatment is enucleation when possible; however, sometimes only a partial resection can be accomplished because of infiltration of vital structures. Even with a benign mass such as this one, limited excision could be carried out in the paravertebral region. Blood loss may be extensive. In our patient blood drainage was less than expected. Postoperative irradiation appears to have little value. When the mass is benign, as in our patient, the prognosis is favorable.

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