HEMANGIO-ENDOTHELIAL SARCOMA OF THE RIB IN A FOURTEEN YEAR OLD GIRL*

R. Cengiz Yalçın, M.D., Kemal Gavuzoğlu, M.D.,*** and İnci Köşeli, M.D.****

Hemangio-endothelial sarcoma is a rare disease of unknown etiology. Stout[1] gave a comprehensive description of the disease in 1943, based on a study of 18 cases from various sources and with the cooperation of several different surgeons and pathologists. Only two of these cases involved infants. Wollstein[2] in 1931, reported one case of a four month old infant with primary sarcomatous lesion in the lung. At Memorial Center in New York, up to 1960, only two pediatric cases of hemagio-endothelial sarcoma of the bone have been seen. McCarthy reviewed 1056 cases of angioma, and identified 20 of them as angiosarcomas, two of which occurred in children.[3] Andersen reported on 175 tumors seen in children at Presbyterian Center in New York, two of which were angiosarcomas originating in the liver.[4] Handy reviewed 1930 cases of malignant tumors in children in New York State (New York City excluded), Kiesewetter and Mason reported 404 cases in Pittsburg, and Campbell, et al, reported 361 cases of tumor in children in Manchester, England.[5–7] These reports do not identify separately the cases of hemangio-endothelial sarcoma.

Kauffman and Stout recorded only 129 cases of malignant hemangio-endotheliomas seen at the College of Physicians and Surgeons, Columbia University, during a 54-year period ending in 1961.[8] Nine of these involved children under 16 years of age. They found acceptable reports of only nine other pediatric cases in the literature. In most of these cases, the tumor originated in the ribs.

Hartman and Stewart reported ten cases of hemangio-endothelioma, primarily of the bone, in adults, but in only one case did the tumor originate in the ribs.[9]

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* From the Departments of Pediatrics, Chest Surgery and Pathology, Atatürk Sanatorium, Ankara, Turkey.

** Pediatrician.

*** Chest Surgeon.

**** Pathologist.

Case Report

This 14 year old girl was admitted to the children's division of the Atatürk Sanatorium on July 6, 1963, because of persistent pain in the left side of the chest and the left arm. She had been in relatively good health until three or four months prior to admission, when pain developed in the left lateral aspect of her chest. She did not see a physician until the pain became more severe and she could not move her left arm. Hospitalization was advised.

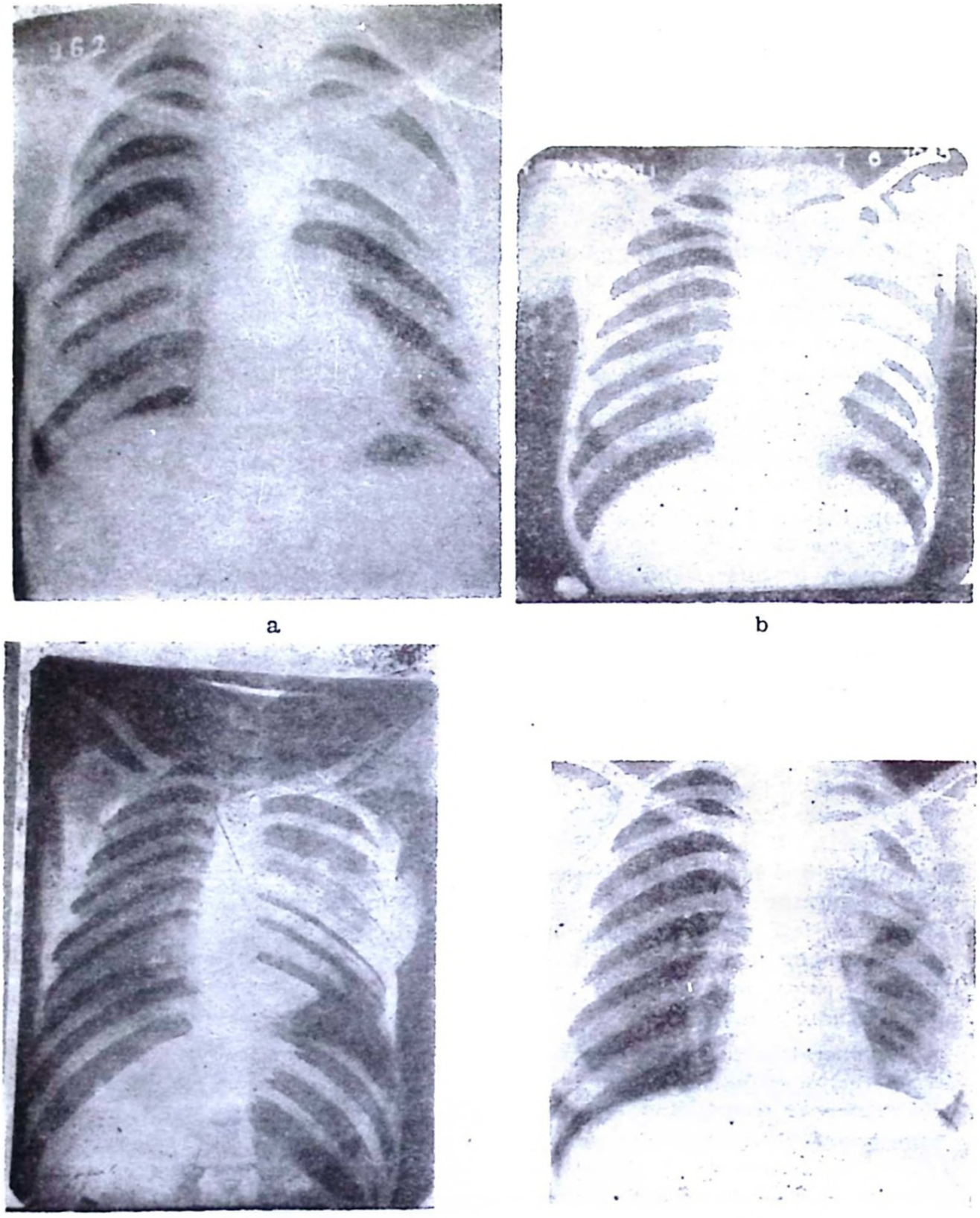
Her past medical history included the usual childhood diseases and an automobile accident six years prior to admission. The family history was noncontributory.

Physical examination: A hard mass was palpable at the left anterior axillary line over the fourth rib. The mass was visible at one point and palpable over the entire length of the rib. There was dulness in the upper and medial regions of the left chest and respiration was diminished in these areas. Blood pressure was 115/70 mmHg, and pulse rate was 84 per minute. Heart sounds were clear and the electrocardiogram was normal. Tests of lung function indicated vital capacity of 2200 cc (77.7 % of normal); breathing capacity was 70 liters (74.0 % of normal); minimum volume was 24.8 liters; and oxygen up-take 240 cc. Blood studies revealed hemoglobin, 75 %; white blood cells, 5800 with differential counts of polymorphs, 62; lymphs, 30; stab cell, 5; monocytes, 3; platelets, 163,000. Bone marrow studies revealed a slight shift of leucocytes to the left, but within normal limits. No pathological cells were observed in the different stains. Wasserman and Kahn tests were negative.

A chest x-ray showed an irregular enlargement along the course of the fourth rib, with two fractures. The third and fifth ribs were pushed out of place and a massive shadow was seen in the upper and medial regions of the left lung (Figure 1 B). Bone x-rays of the skull, extremities and spine were negative. The patient had had a microfilm of the chest the previous year during a tuberculosis screening test, but there was no follow-up (Figure 1 A). Bronchoscopy showed the left main bronchial lumen to be narrower than normal. During bronchiography it was impossible to fill the bronchus of the left upper lobe with radiopaque material, and the bronchial tree of the left lower lobe was only partially visualized.

An operation to remove the tumor was performed on July 11, 1963. There was a fusiform tumoral mass over the course of the fourth rib. The tumor was extremely fragile and bled easily. The upper lobe of the left lung was completely infiltrated by the tumor, with partial infiltration of the lower lobe. Approximately 400 cc of bloody fluid was aspirated from the thoracic cavity. A frozen section of the tumor was diagnosed as cancer. Considering the possibility of infiltration into the mediastinal and aortic areas, a radical removal of all diseased areas was not feasible. Only the original tumor of the fourth rib was resected.

The gross specimen was an 18 by 6 by 5 cm fusiform mass with pieces of the fourth rib at each end (Figure 2). One side of the tumor was rather smooth and covered with fibrotic tissue. The other side was uneven in appearance. It was filled with small soft yellowish-white necrotic and hemorrhagic nodules. The tumor itself was partly destroyed. One lymph node in the infiltrated lung tissue gave the impression microscopically that it had not been infiltrated by



a b c d

Figure 1. A. Film of the chest taken a year prior to admission during a screening test for tuberculosis, revealing a massive shadow in the left upper lobe and irregularity of the fourth rib. B. On admission, revealing the more massive and extensive lesion. C. Film of the bones of the chest and spine taken on admission, showing rib destruction more clearly. D. Film taken 8 months after operation and x-ray therapy.

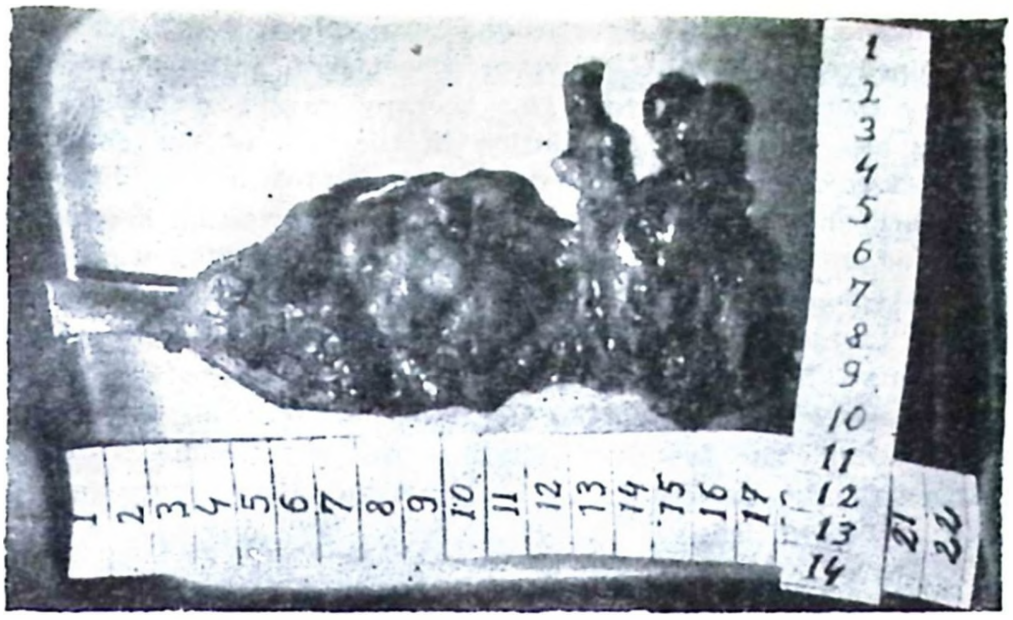


Figure 2. Photograph of the excised tumor lesion revealing an extremely fragile tumor with pieces of rib at both ends.

the tumor. Histological examination revealed that the tumor was very rich in capillaries. Tumoral infiltrations consisted mostly of oval shaped cells with a nucleus of loose chromatin. There was marked proliferation of the capillary endothelium, and capillary spaces were full of malignant epithelial cells, some of which were heaped up and some of which formed islands. Specimens of material from the rib also had the same microscopic appearance (Figure 3). The histological diagnosis was hemangio-endothelial sarcoma.

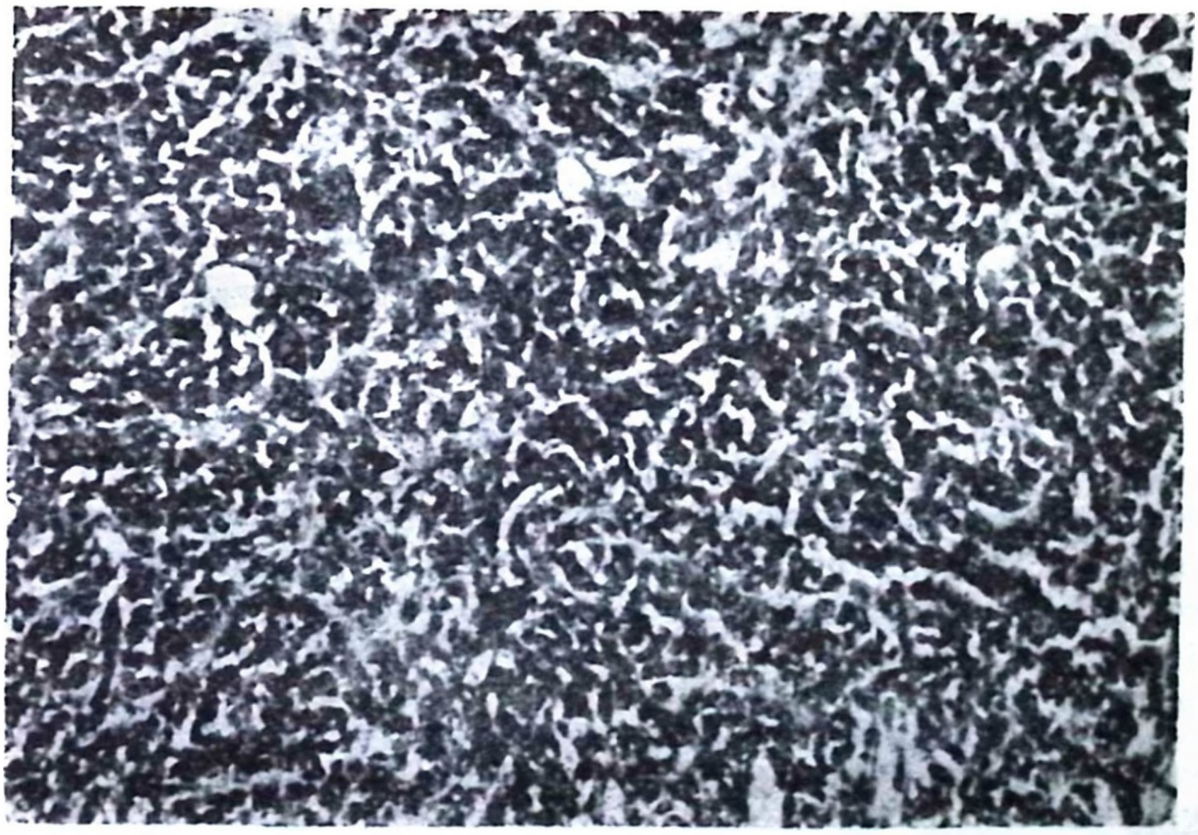


Figure 3. Photomicrograph (78X) of the tumor, revealing a marked proliferation of capillary endothelium and malignant epithelial cells forming islands or heaped in capillary spaces.

The patient's postoperative course was uncomplicated. She felt somewhat better and gained some weight. However, chest pain returned and a 31-day course of x-ray therapy was given. This therapy gave some relief from pain and there was also a marked diminution in the size of the shadow of the left lung on x-ray. The patient felt fairly comfortable for the next six months, but then she developed a dull pain in her lower back which became progressively more severe. A bone survey and flat plate of the abdomen revealed no pathological symptoms. X-ray therapy was begun eight months after the first course, and there was a dramatic relief from pain following the second course. The patient was comfortable for another six months, but thereafter x-ray therapy had to be re-instituted. One and a half years after the operation, she lost her vitality and her condition degenerated quickly. Although the patient is still alive, she is on increasing doses of narcotics for relief of pain.

Discussion

A diagnosis of hemangio-endothelial sarcoma cannot be made on the basis of clinical observations alone. Definite diagnosis can be made only after an exploratory operation and histological examinations.[2] The histology of these tumors is characterized by numerous capillaries composed of malignant cells which are heaped up (as in our patient) and sometimes simulate vascular cavernous sinuses.[1, 3, 10, 11] The cells can be fusiform, round or polygonal.[10, 11] Clinically and histologically the tumor may simulate Ewing's sarcoma,[1–3] and some authorities consider this type of tumor to be a variety of Ewing's sarcoma.

The etiology of this disorder remains a mystery. Although many hold that trauma plays an important role, McCarthy noted a history of trauma in only three of 20 cases he investigated, and Stout noted only two in his series.[1, 3] McCarthy believes that they may be of genetic origin, and some hold that congenital vascular defects are generally the cause of such tumors originating in the liver. Our patient had a history of trauma six years prior to hospitalization, but, although the trauma was on the left side, we have no evidence of any connection between the trauma and the tumor. Considering the histology, x-ray findings, and history, we believe that this tumor originated from the medullary capillaries of the fourth rib.

According to McCarthy, the average survival time of patients with this kind of tumor is approximately two and a half years.[3] Nine per cent of the patients survive for five years, and 17 per cent for three years. Two patients seen at New York Memorial Center survived for less than 25 months. Our patient is in poor

condition but is still alive after more than two and a half years following the first x-ray and diagnosis. The x-ray taken a year prior to admission in the course of a tuberculosis survey showed the tumoral lesion of the fourth rib, but lung infiltration had not proceeded very far.

Some believe that malignant cells spread along lymph channels, whereas others hold that local invasions occur. We believe that in our case, the tumor was a local growth.

In the majority of cases, the symptoms are recognized too late and the prognosis is generally poor. For this reason «hemangiomatosis of the lung should be considered as a rare possibility in the differential diagnosis of unexpected shadow in the lung in even the youngest infants».[12] We feel that had our patient's condition been diagnosed earlier her chances for survival would have been better. Early surgical removal of the tumor could have effected a complete cure. In the later stages of the disease, intermittent fractional x-ray therapy might be considered the best therapy for relief of pain and comfort of the patient.

Summary

The case of a 14 year old girl with hemagio-endothelial sarcoma of the rib is presented. Diagnosis was made by histological examination and the main lesion was excised. The authors review the literature on this disorder, and discuss the etiology, diagnosis, treatment and prognosis.

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