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PERICARDIAL MESOTHELIOMA*

A Pediatric Case Report

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Pericardial mesothelioma is an extremely rare tumor originating from mediastinal cells. A total 120 cases were reported with this tumor by 1985.[1] This tumor is most commonly seen in the third and fourth decades of life; males outnumbering females by a ratio of two to one.[2] Anorexia, weight loss, fatigue, fever and cardiac symptoms related to the localization of the tumor comprise the clinical picture which closely resembles pericarditis. In 75 percent of cases, correct diagnosis has been established at postmortem examinations[1]. Morphologically, these tumors may be either localized or diffuse. Dawe et al[3] have defined three histological subtypes of pericardial mesothelioma: pure epithelial, spindle cell, and mixed. Metastases to the pleura, adrenal glands and skin have been noted in 25 percent of cases[4]. The prognosis of the disease is poor. Surgical excision is usually impossible, and treatment with irradiation and chemotherapy generally lead only to temporary improvement[4]. The mortality rate is 60 percent within six months following the appearance of clinical symptoms[5].

We decided to present our case because the number of childhood cases of this type of tumor diagnosed preceding death is extremely rare.

Case Report

A nine and a half year old boy was admitted to the Istanbul University Children's Hospital with symptoms of cough, dyspnea and mild fever of one month's duration. The history did not reveal any pertinent information such as exposure to asbestos fibers. Physical examination revealed dyspnea, and crepitations were heard in the lower lobes. Cardiac dullness was found to be enlarged. The heart sounds were muffled.

Laboratory studies which included peripheral blood counts, serum glucose, urea, creatinine and electrolytes were all within normal limits except for an elevated

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erythrocyte sedimentation rate of 90 mm/h. The chest teleradiogram revealed a cardio-thorax index of 0.7, and the left cardio-phrenic angle was obscured. The ECG showed negative S – T waves in the left precordial leads. Echocardiography revealed a significant collection of fluid and a large cystic mass located on the left side and posteriorly within the pericardium (Fig. 1). Axial computerized tomograms of the thorax and abdomen revealed a normal abdomen but a mediastinal tumoral mass occupied a substantial part of the left hemithorax (Fig. 2). Cytologic

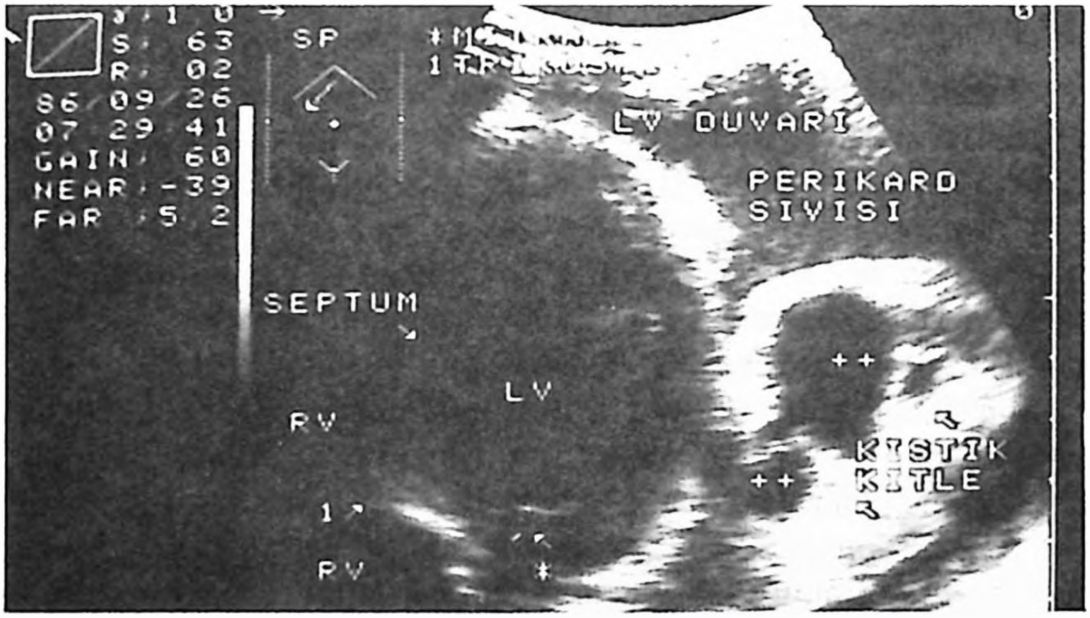

Fig. 1: The appearance of pericardial fluid and cystic mass on parasternal short axis examination

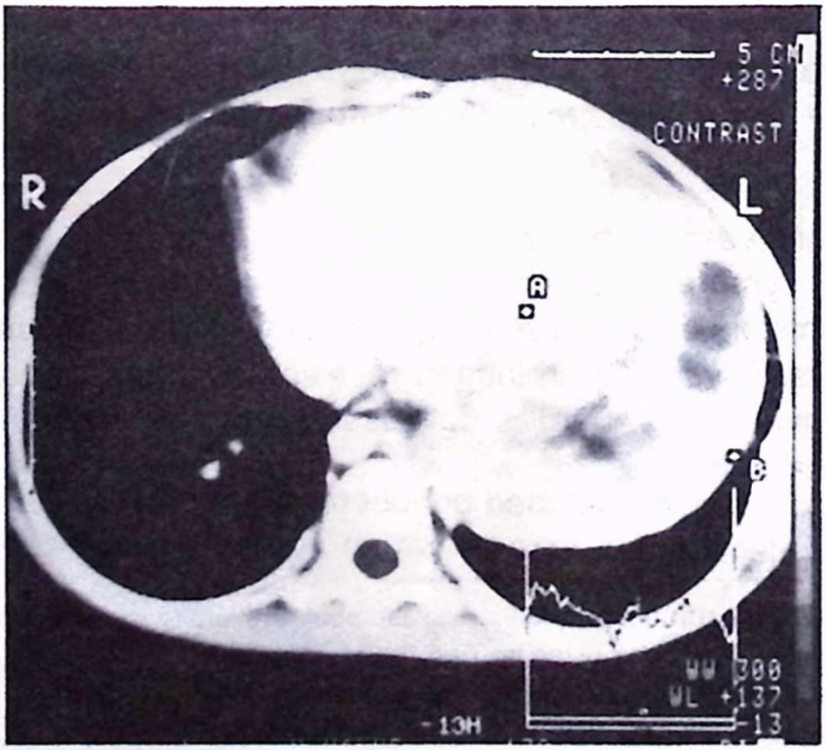

Fig. 2: Computerized axial tomogram of the thorax showing solid tumoral matter starting from the left upper mediastinum and covering a substantial portion of the front and central sections of the left hemithorax, extending down on to the left diaphragm, and containing necrotic areas

examination of the pericardial fluid disclosed copious atypical mesothelial cells. An open pericardial biopsy showed a diffuse, well-differentiated, tubulopapillar, malignant epithelial mesothelioma. Combination chemotherapy with cytoxan actinomycin D and dacarbazine was initiated[6]. No apparent clinical improvement was obtained and the patient's condition continually deteriorated. He died fifteen months after his admission. At postmortem examination a pericardial tumor weighing 2460 g and measuring 22 x 19 x 14 cm. was detected. The tumor also involved the heart. The mass was soft, hemorrhagic, greyish-yellow and could easily be separated from the adjacent tissues. It was partially necrotic and contained many cysts (Fig. 3). A small section of the tumor had invaded the

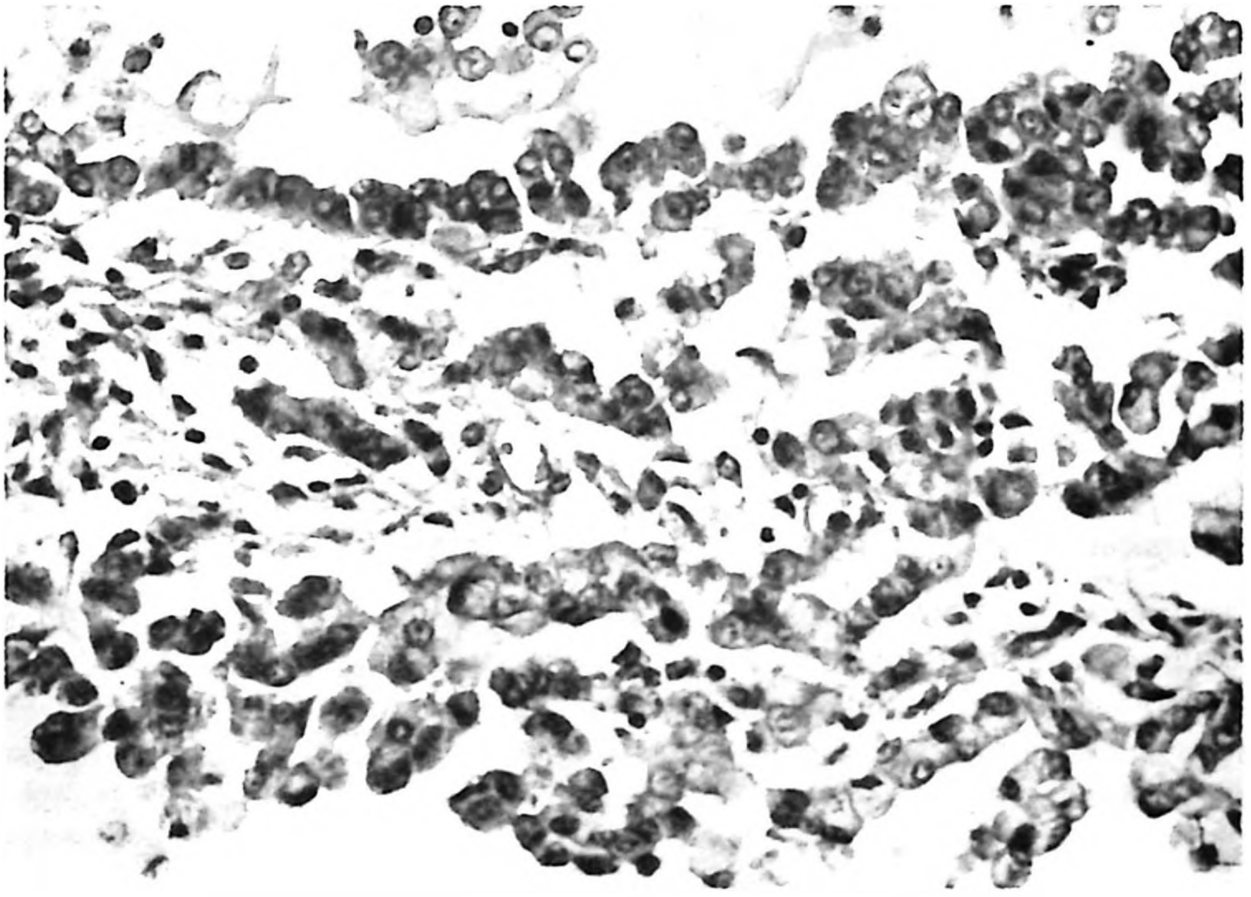

Fig. 3: Epitheloid type mesothelioma of tubulopapillar structure

myocardium. No tumoral formation was seen in the hilar, bronchial or mediastinal lymph nodes. Light and electron microscopy confirmed the diagnosis of a malignant epitheloid mesothelioma, and tubulopapillar in type, as identified in the biopsy (Fig. 4).

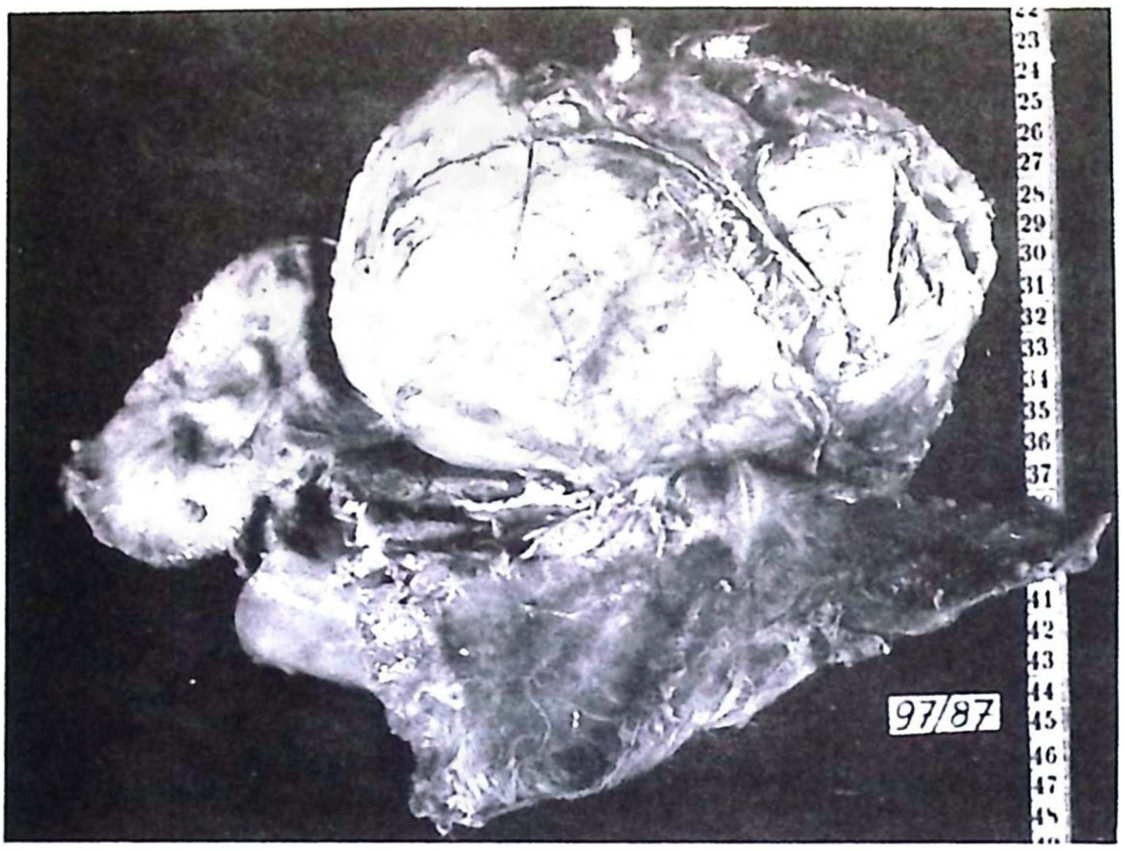

Fig. 4: Tumoral matter which can easily be separated from the pleura, and with no penetration to the parietal pericardium

Discussion

Primary pericardial mesothelioma is a very rare neoplastic disease. Pleural mesotheliomas are reported usually to be associated with pulmonary asbestosis[7]. The etiologic factors in pericardial mesotheliomas are yet unclear. The family and personal histories of this case disclosed that there had been no previous exposure to materials containing asbetos fibers. Besides, it has been reported that a period of latency ranging from 12 to 38 years after exposure to asbetstos fibers is necessary for this tumor to develop.[8]

Primary pericardial mesothelioma is an occasional diagnosis in all age groups but it is still rarer in children. In a review of the relevant literature, we were able to find only six pediatric cases reported to date[8-13].

The majority of the cases present as pericarditis[4]. Telecardiographic and ECG findings of our patient were also highly suggestive of pericarditis, but the echocardiographic examination revealed a retrocardiac cystic mass. It is also sometimes difficult to distinguish between malignant mesothelial cells and reactive cells on cytologic examination. Since a homogenous pericardial mass may be mistaken for fluid in echo imaging, CAT examination is more valuable in clinical diagnosis.[5]

It should also be noted that in 74 percent of cases pleural mesotheliomas involve both the pericardium and myocardium, and they must be carefully excluded in diagnosing primary pericardial mesothelioma[14]. In our case, postmortem examination revealed that the pleura was free of tumoral involvement. Our findings were in complete agreement with the criteria devised by Andersen and Hansen[9] which include limitation of the disease only to the pericardium, no tumoral penetration of the parietal pericardium, only regional lymph node involvement, if at all, and the verification of these criteria at postmortem examination.

Summary

A nine and a half year old boy was brought to the hospital because of a cough, dyspnea and mild fever. He was well-nourished and had an uneventful history. His chest X-ray and electrocardiographic findings suggested pericarditis but further examinations and an open pericardial biopsy revealed a mass histologically diagnosed as pericardial mesothelioma, a very rare tumor in this age group.

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