Unusual region for pericardial malignant mesothelioma: cutaneous manifestation in a Turkish woman

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Abstract

Malignant mesothelioma is a disease that originates from mesenchymal cells. It is related to the occupational or environmental exposure to asbestos. The treatment remains controversial because it is commonly diagnosed at a very late stage, and the prognosis is very poor. In this report, we present a 37-year-old female patient who was admitted with shortness of breath, palpitation and inability to sleep on her back for the previous 10 days. A large pericardial effusion was detected on echocardiography. Pericardiocentesis was performed and the patient's symptoms were alleviated. However, approximately 7 months later, she was readmitted to the clinic with complaints of a mass at the incision site. Pathological examination of the mass yielded a diagnosis of pericardial malignant mesothelioma. Malignant mesothelioma is a rare occurrence, and to our knowledge, there are no reports in the English literature of pericardial malignant mesothelioma local invasion to an incision site.

Introduction

Malignant mesothelioma is an extremely rare tumor that arises from pleural, pericardial or peritoneal mesenchymal cells.[1] A male predominance has been described. The tumor is found in patients 15-60 years of age. Especially in Central Anatolia (Turkey), since the emergence of exposure to erionite fibers in some regions, a clinical disease may be seen between 20-30 years of age. Barış and colleagues reported that 62 people died due to mesothelioma from 1970-1981 in the Karain village of Konya.[2] The clinical manifestation of malignant pericardial mesothelioma is nonspecific. The diagnosis is usually achieved during autopsy or by evaluation of surgical materials. The prognosis is very poor and the median survival from diagnosis is 6 months.[3] The optimal treatment is still controversial, but surgery, radiotherapy, chemotherapy, or combination therapies are most frequently used in practice. Primary malignant pericardial mesothelioma is described in the literature with about 150 cases. Only a few cases with metastasis of malignant mesothelioma have been reported. In our study, an unusual case of pericardial mesothelioma presenting as a mass at the incision site is described.

Case Report

A 37-year-old female patient was admitted with a 10-day history of shortness of breath, palpitation and inability to sleep on her back. The plain chest X-rays showed extensive pericardial effusion. She was a lifelong nonsmoker, and there were no risk factors in her medical history. After the diagnosis was confirmed with echocardiography, the patient was operated under general anesthesia. Pericardiocentesis was performed with midline incision below the sternum. Approximally 500 mL of hemorrhagic pericardial fluid was aspirated. She had significant symptomatic relief after drainage. The mediastinal region was decreased in the follow-up chest radiograph, and echocardiography revealed minimal pericardial effusion. A cytological evaluation of the pericardial fluid was negative for malignant cell or infection. She was discharged with medical treatment follow-up. Approximately 7 months later, she was readmitted to the clinic with complaint of a mass at the incision site (Figure 1). Ultrasonography revealed a multiloculated collection (abscess) at the incision site. The patient underwent surgery under local anesthesia, and the mass was removed in its entirety and sent for pathological examination. Histologically, the tumor was composed of neoplastic epithelial cells arranged in irregular tubular and papillary structures that had extensively invaded the muscular and lipomatous tissue. Focal areas were also seen in which atypical cells were arranged in storiform or patternless pattern separated by dense collagenized tissue. Microscopic examination revealed malignant mesothelioma of epithelioid and desmoplastic type. The neoplastic cells were cuboidal and had distinct cell borders, a low-to-moderate nuclear-to-cytoplasmic ratio, pale-to-clear cytoplasm, and pleomorphic round-to-oval nuclei. The neoplastic cells were strongly positive for calretinin and focally positive for Ber-Ep4 and MOC-31. Thyroid transcription factor (TTF)-1 and thyroglobulin immunostaining, which was performed to differentiate from thyroid papillary carcinoma because of papillary proliferation of epithelioid cells, was negative (Figure 2). On the basis of these findings, the morphological diagnosis was malignant mesothelioma. Positron-emission tomography-computerized tomography (PET-CT) showed increased ^{18}F-fluorodeoxyglucose (FDG) uptake (SUV max: 17.80) in a 53×53×76 mm lobulated mass extending to the paracardiac area of skin beneath the sternum with pericardial thickening (Figure 3). Lymphadenopathy was found with increased FDG uptake posterior to the body of the pancreas.

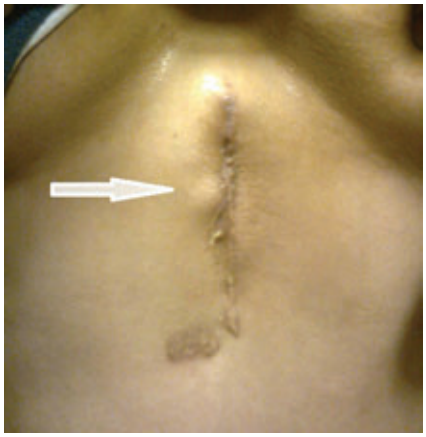

Figure 1. The mass is seen at the incision site (white arrow).

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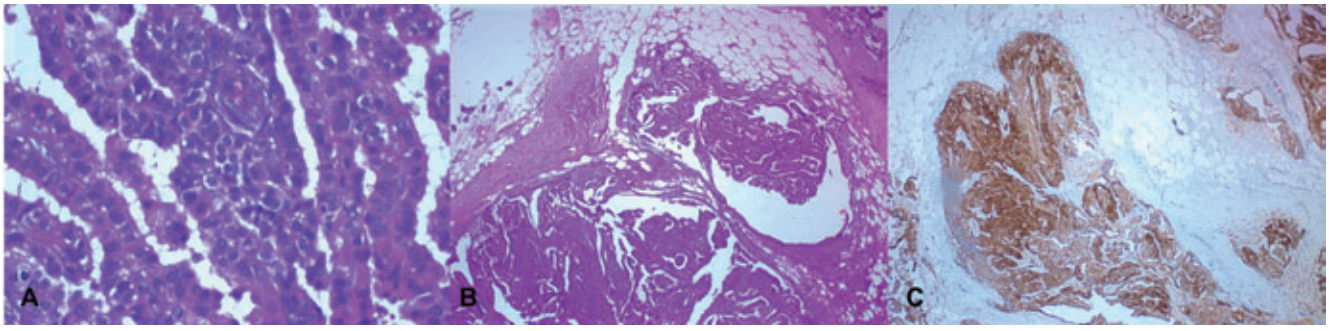

Figure 2. A) The tumor showed papillary growth pattern with pleomorphic round-to-oval nuclei [Hematoxylin & Eosin (H&E) stain, ×40]. B) Tumor tissue invading the muscular and lipomatous tissue (H&E stain, ×4). C) Tumor cells were strongly positive for calretinin.

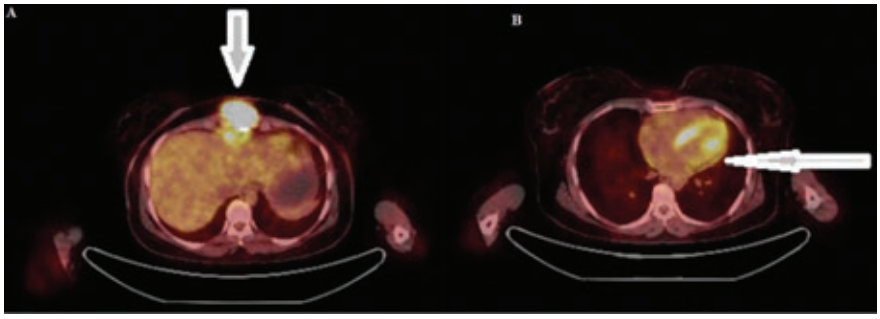

Figure 3. Positron-emission tomography imaging demonstrates increased FDG uptake of the mass (A: white arrow) and thickened pericardium (B: white arrow).

Discussion

Malignant mesothelioma is a very rare disease and carries a poor prognosis. The incidence of primary malignant pericardial mesothelioma was below 0.0022% in a necropsy study.[4] Most cases of mesotheliomas are related to asbestos exposure. Asbestos causes a chronic inflammatory and fibrotic reaction, which is managed by cytokines and activated macrophages. Tumors are classified as epithelioid, sarcomatoid, desmoplastic, or biphasic based on tissue biopsy. Any combination of patterns may be present. Epithelioid mesotheliomas show a wide range of morphologic patterns, such as tubulopapillary, adenomatoid, lymphohistiocytoid, deciduoid, and small cell variant. Recognition of these variants of epithelioid mesotheliomas is important in the diagnosis, but they have no clear prognostic significance. Spindle cell and desmoplastic mesotheliomas are associated with a shorter survival than the conventional type.

The clinical findings of pericardial mesothelioma are constrictive pericarditis, pericardial effusion, cardiac tamponade, and heart failure. Echocardiographic evaluation is the most commonly used diagnostic tool. A pericardial fluid drainage provides relief to the patient. However, the reliability of body cavity fluid cytology is low for malignant mesothelioma; its sensitivity is reported to be 33-84%.[5] Differentiation of mesothelioma from benign mesothelial hyperplasia with reactive atypia can be very difficult or impossible in cytologic specimens, so purely cytologic diagnosis of malignant mesothelioma is fairly low. For definitive diagnosis, histochemical, immunohistochemical and electron microscopic examinations are required. Magnetic resonance or PET-CT may be used to identify the presence of a pericardial mass. Duysinx *et al*. found PET scanning to have a 96.8% sensitivity and 88.5% specificity for malignant pleural disease.[6] Mesotheliomas mainly metastasize to the intrathoracic lymph nodes or lung, and distant metastasis of malignant pericardial mesothelioma is very rare. In the literature, intracardiac invasion into the right atrium was reported in pericardial mesothelioma.[7] Pleural mesothelioma with metastasis has been described in a wide variety.[8,9] Furthermore, tumor metastases from the parietal pleura to the skin surface following tracts from pleural procedures are known complications of mesothelioma.[10] However, pericardial mesothelioma metastasis to the skin surface is not a familiar situation.

Conclusions

Etiological research is very important in patients presenting with pericardial fluid, especially in endemic areas. In our study, while the patient had no known history of asbestos exposure, she may have been exposed due to her residence in a village area of Konya. No reports were found in the literature regarding pericardial malignant mesothelioma local invasion to an incision site. Although pericardial fluid cytology may be negative for these malignant diseases, the masses should be considered seriously because early diagnosis is very important.

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