

Localized isolated intrapericardial mesothelioma: a case report

Lokalize izole intraperikardiyal mezotelyoma: Olgu sunumu

Mohammed Alaa Nady,¹ Hussein Elkhayat,¹ Mohammed Mahmoud Mostafa,¹ Maged Abdelfattah²

¹Department of Cardiothoracic Surgery, Assiut University Hospitals, Egypt

²Department Oncology, South Egypt Cancer Institute, Assiut University, Egypt

There are several cancers and diseases which affect the pericardial space around the heart, and pericardial mesothelioma is one of the rarest pathology. It is the most common primary malignant pericardial tumor. The clinical presentation of malignant pericardial mesothelioma includes chest pain, shortness of breath, coughing and severe nocturnal sweating. In this article, we report the fifth case of the localized isolated pericardial mesothelioma in the literature and the longest survival at two years reported so far.

Key words: Mesothelioma; pericardium; thoracotomy.

Primary tumors of the pericardium are extremely rare, and this was proven by one of the largest necropsy series, which showed an incidence rate of 0.0022 in 500,000 cases. Mesothelioma is the most frequently seen form of this tumor, followed by sarcoma, teratoma, and fibroma.^[1-3]

Malignant mesothelioma is the most common primary malignancy of the pleura. It not only arises from the mesothelial surfaces of this serous membrane, but it can also come from the peritoneum and the pericardium. The recognition of an association between asbestos exposure and malignant mesothelioma dates from the work of Wagner^[4] in the 1960s, and the simian virus 40 (SV40) was recently identified as an additional significant carcinogen.^[5]

CASE REPORT

A 49-year-old female presented to our clinic with shortness of breath and chest pain that was retrosternal and stitching in character. On examination, the patient

Kalbin etrafındaki perikard boşluğunu etkileyen çeşitli kanser ve hastalıklar mevcut olup, perikardiyal mezotelyom bunların en nadir görülen patolojilerinden biridir. Perikardiyal mezotelyom, en sık görülen primer malign perikardiyal tümördür. Malign perikardiyal mezotelyomanın klinik tablosu göğüs ağrısı, nefes darlığı, öksürük ve geceleri şiddetli terlemeden oluşur. Bu yazıda, bugüne kadar bildirilmiş iki yıllık en uzun sağkalım süresi ile birlikte, literatürdeki beşinci lokalize izole perikardiyal mezotelyom olgusu sunuldu.

Anahtar sözcükler: Mezotelyom; perikardiyum; torakotomi.

was orthopneic, and there were congested neck veins, diminished cardiac sounds, and mild lower limb edema (ankle edema). There was no history of asbestos exposure.

A chest X-ray showed an enlarged cardiac silhouette that was suggestive of pericardial effusion. Echocardiography revealed moderate to severe pericardial effusion, and a mass was seen relative to the left atrium. In addition, the parietal pericardium was thickened, and the cardiac dimensions were normal (Figure 1). Pericardiocentesis revealed reddish fluid with occasional small-sized balls of atypical epithelial/mesothelial cells scattered in the hemorrhagic background, which was indicative of malignancy.

Chest computed tomography (CT) showed a left superior mediastinal mass, a thickened pericardium, and a moderate to massive amount of pericardial effusion. However, an abdominal CT scan showed no abnormalities. A preoperative CT-guided biopsy



Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2013.5740
QR (Quick Response) Code

Received: July 4, 2011 Accepted: November 13, 2011

Correspondence: Hussein Elkhayat, M.D. Department of Cardiothoracic Surgery, Assiut University Hospitals, 71515 Assiut, Egypt.

Tel: +0020105549653 e-mail: dr_khayat@hotmail.com



Figure 1. Echocardiographic finding of the mass in direct relation to the left atrium.

of the mass revealed a core of tissue with solid sheets of malignant epithelial cells that were surrounded by markedly hyalinized fibrous tissue. This suggested poorly differentiated carcinoma, and immunophenotyping was negative for cytokeratin (CK)7 and CK20 but positive for pan-CK.

The patient received two cycles of chemotherapy with VP-16 (etoposide) and Platinum (cisplatin) and was scheduled for surgery three weeks later.



Figure 2. Intraoperative view of the mass after opening the pericardium.

To begin the operation, the patient was placed in the supine position, and a left anterolateral thoracotomy was performed. The pericardium was markedly thickened. It was opened, and serosanguinous fluid was drained from the intrapericardial cavity. Further exposure of cavity revealed a reddish-brown mass (7x5 cm) with a nodular surface. The mass was attached to the left atrial appendage, and it was dissected using both blunt and sharp dissection. There was no infiltration to the myocardium. A partial pericardiectomy was performed, and the chest was closed in layers in a standard fashion after inserting pericardial and pleural drains (Figures 2 and 3).

A microscopic examination revealed irregular sheets of small, monotonous, rounded cells arranged around small vascular slits in a hemangiopericytoma-like pattern. The tumor cells had scanty cytoplasm, relatively large hyperchromatic nuclei with coarse chromatin, and inconspicuous nucleoli. In addition, the investing stroma was abundantly hemorrhagic and desmoplastic in nature.

Immunohistochemistry showed that the tumor cells were immunoreactive with CK (an epithelial tissue marker) and non-reactive with either neuron-specific enolase (NSE) (a marker of neuroendocrine cells), cluster designation (CD)34 (a marker of malignant vascular tumors), thyroid transcription factor-1 (TTF-1), CK7, and CK20. Calretinin, Wilms' tumor protein 1 (WT1), the monoclonal antibody D2-40, mesothelin (MSLN), and thrombomodulin (TM) along with CK5 and CK6 are not available in



Figure 3. Mass after excision.

our institute. The patient was then diagnosed with a small-cell variant of malignant mesothelioma.

The pericardial and pleural drains were removed according to standard protocol, and the patient was referred to the oncology department where she received three cycles of chemotherapy with Gemzar (gemcitabine) and Platinum (cisplatin). The patient was followed up with a chest CT scan that showed a stationary course, and she then received another three cycles of chemotherapy. After this course of chemotherapy, the patient unfortunately refused to come for further follow-up visits.

The patient had an uneventful postoperative period and was followed up via chest CT and echocardiography for 11 months postoperatively. No stationary course of the disease was reported, and no recurrence was detected. The patient refused to come for the next scheduled visits due to psychological and social problems, but follow-up was conducted by telephone interviews in which no further complaints regarding any of her previous symptoms were reported. At last report, the patient was continuing to do well with no complaints of any kind related to her surgical procedure.

DISCUSSION

There are several types of cancer and various diseases which affect the pericardial space around the heart, but pericardial mesothelioma is one of the rarest. However, it is the most common primary malignant pericardial tumor and can also be secondary to malignant pleural mesothelioma. Pericardial mesothelioma presents either as a localized tumor associated with pericardial effusions^[6] or as an encasing mass involving the whole pericardium.^[7] The degree of invasion varies, but local infiltration into the cardiac chambers has been reported.^[8] Chest pain, shortness of breath, coughing, and severe sweating at night have all been recorded as symptoms of malignant pericardial mesothelioma. When pericardial effusion is present, pericardiocentesis can temporarily improve the symptoms, but it often fails to provide a cytological diagnosis. To further complicate matters, a CT scan can interpret it as fluid rather than the malignant tumor that it is.^[9] Surgery was the only reasonable therapeutic strategy to establish the diagnosis and remove the mass in our patient. Chemotherapy and radiotherapy would have been only palliative.^[6,10] A review of the literature yielded four other cases of localized pericardial mesothelioma besides ours. Most cases of this rare cancer are seen in women and are of the epithelial type.^[11]

In our case, due to the lack of positron emission tomography (PET)-CT in our institute, we depended

only on a conventional chest CT scan with echocardiography for the diagnosis and follow-up of this patient. Intraoperatively, after we were confronted with the tumor, we thought it was a mistake to not have performed magnetic resonance imaging (MRI) to check for infiltration of the myocardium and adjacent great vessels.

In conclusion, we did not expect to find an intrapericardial mass and thought that the issue in our patient could be the result of an invasion from a superior mediastinal tumor, so video-assisted thoracic surgery (VATS) was not considered preoperatively. However, after we opened the pericardium to evacuate the effusion, we were confronted with the tumor. Hence, the superior mediastinal mass as seen by CT was in fact entirely intrapericardial. A diagnosis of pericardial mesothelioma was made based on the clinical findings and available immunohistochemical investigations. In our opinion, the main difficulty with regard to pericardial mesothelioma is that because it is so rare, it is almost never considered as the primary diagnosis.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Kralstein J, Frishman W. Malignant pericardial diseases: diagnosis and treatment. *Am Heart J* 1987;113:785-90.
2. Okura Y, Kato K, Hanawa H, Izumi T, Kamishima T, Yamato Y, et al. Pericardial mesothelioma secreting thrombomodulin. *Am Heart J* 1996;132:1309-11.
3. Darsee JR, Braunwald E. Diseases of the pericardium. In: Braunwald E, editor. *Heart disease, a textbook of cardiovascular medicine*. Philadelphia: Saunders; 1980. p. 1517-82.
4. Wagner JC. The sequelae of exposure to asbestos dust. *Ann N Y Acad Sci* 1965;132:691-5.
5. Carbone M, Rizzo P, Pass H. Simian virus 40: the link with human malignant mesothelioma is well established. *Anticancer Res* 2000;20:875-7.
6. Gössinger HD, Siostrzonek P, Zangeneh M, Neuhold A, Herold C, Schmoliner R, et al. Magnetic resonance imaging findings in a patient with pericardial mesothelioma. *Am Heart J* 1988;115:1321-2.
7. Ohnishi J, Shiotani H, Ueno H, Fujita N, Matsunaga K. Primary pericardial mesothelioma demonstrated by magnetic resonance imaging. *Jpn Circ J* 1996;60:898-900.

8. Lund O, Hansen OK, Ardest S, Baandrup U. Primary malignant pericardial mesothelioma mimicking left atrial myxoma. Case report. *Scand J Thorac Cardiovasc Surg* 1987;21:273-5.
9. Andersen JA, Hansen BF. Primary pericardial mesothelioma. *Dan Med Bull* 1974;21:195-200.
10. Nambiar CA, Tareif HE, Kishore KU, Ravindran J, Banerjee AK. Primary pericardial mesothelioma: one-year event-free survival. *Am Heart J* 1992;124:802-3.
11. Val-Bernal JF, Figols J, Gómez-Román JJ. Incidental localized (solitary) epithelial mesothelioma of the pericardium: case report and literature review. *Cardiovasc Pathol* 2002;11:181-5.