

Primary pericardial mesothelioma presenting as pericardial constriction

Ansari MA, Thapa R, Pradhan S

Department of Radiology and Imaging, Tribhuvan University, Teaching Hospital, Maharajgunj, Kathmandu, Nepal
International Clinic, Kathmandu, Nepal

Correspondence to: Dr. Mukhtar Alam Ansari, Dept. of Radiology and Imaging, Tribhuvan University, Teaching Hospital, Maharajgunj, Kathmandu, Nepal.

Email: mukhtar077@hotmail.com

Abstract

A 48 year-old-male presented with a six month history of shortness of breath and cough with low grade fever. Chest x-ray shows enlarged cardiac size. Echocardiogram revealed pericardial effusion. ESR was raised and blood count showed lymphocytosis. Anti-tubercular treatment was started and four months after completion of treatment there was no significant improvement. Pericardial biopsy was done which revealed findings of chronic pericarditis. Echocardiogram was repeated which revealed thickened pericardium with few foci of calcification and pericardial effusion. Diagnosis of constrictive pericarditis was made. Patient was planned for pericardiectomy. During operation fleshy vascular mass was seen arising from pericardium along with nodular pericardial thickening. Biopsy was taken and closer was done. Post operatively contrast enhanced CT scan of chest was done which revealed malignant mesothelioma. Histopathological analysis showed a primary malignant mesothelioma. Post operative period was uneventful for initial 2-3 days. Patient was taken by his party against medical advice on 3rd post-operative day. Patient did not returned to the hospital again.

Key words: Pericardial mesothelioma, echocardiogram, CT scan

Introduction

Primary pericardial mesothelioma is a rare malignancy with a very poor prognosis. Its incidence is estimated to be around 0.0022%.¹ It is a highly aggressive tumour with global survival under 6 months. Surgery can be curative in localized cases detected early in the course of the disease, but early detection is unfortunately very rare. It responds poorly to radiotherapy. Management is usually only palliative with pericardiectomy and chemotherapy to reduce the tumour mass.

Case Report

A 19-year old male presented with a five month history of shortness of breath, cough and low-grade fever. The rest of his medical history was unremarkable. Blood count showed lymphocytosis and an elevated ESR. A chest X-ray was done which revealed an enlarged cardiac silhouette.

Echocardiography was done which revealed pericardial effusion.

Anti-tubercular medication was started. However, four months later there was no improvement in the patient's symptoms. A pericardial biopsy was done which revealed evidence of chronic pericarditis. The echocardiography was repeated which showed thickened pericardium with a few foci of calcification along with pericardial effusion. So the diagnosis of constrictive pericarditis was made and pericardiectomy was planned for the patient.

During the surgery a fleshy vascular mass was seen arising from the pericardium, along with nodular pericardial thickening. Biopsy of the mass was taken and closure was done. A post-operative contrast enhanced CT scan of the chest revealed malignant mesothelioma (Fig. 1A, 1 B; 2). Histopathological examination of the biopsy specimen also indicated primary malignant mesothelioma (Fig. 3).

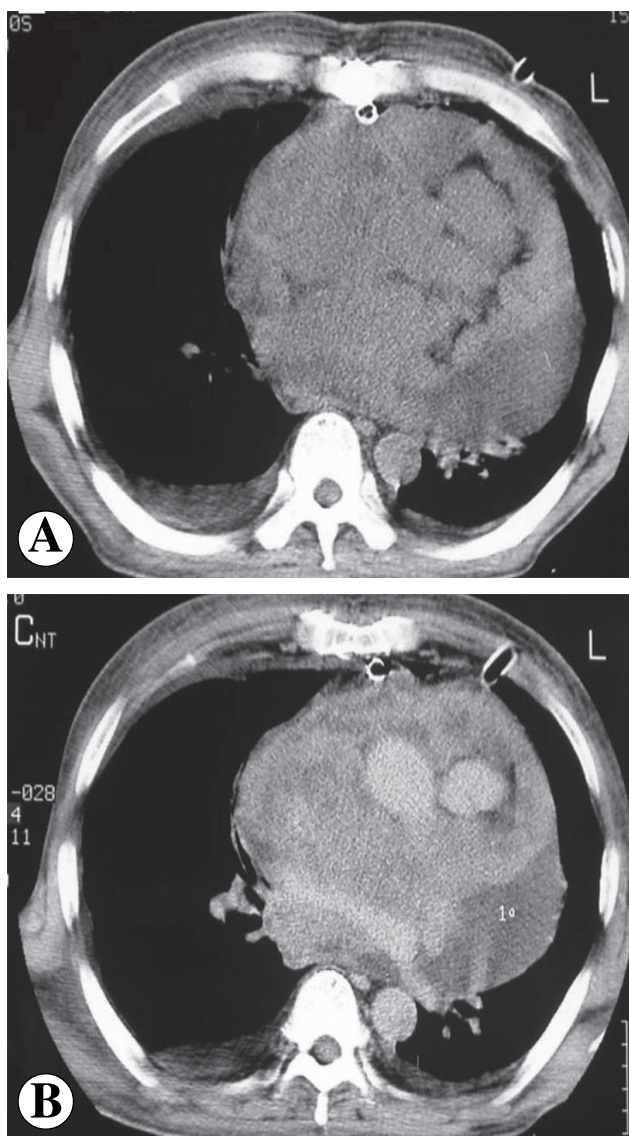


Fig. 1: The plain (Fig. 1 A) and contrast enhanced chest CT (Fig. 1 B) scan shows thick irregular non-enhancing soft tissue density mass lesion surrounding the heart and root of great vessels. There is evidence of fluid collection in mediastinum posteriorly on the left side. Bilateral pleural effusion is also seen.

The initial post-operative period was uneventful. The patient was taken away from the hospital on the third post-operative day against medical advice. He did not return to the hospital after that.

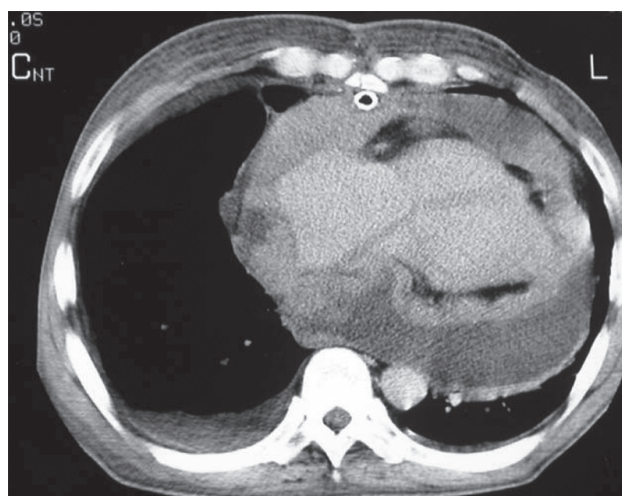


Fig. 2: Contrast enhanced chest CT scan demonstrates extensive pericardial involvement of the tumour that encases the ventricles and atrium.

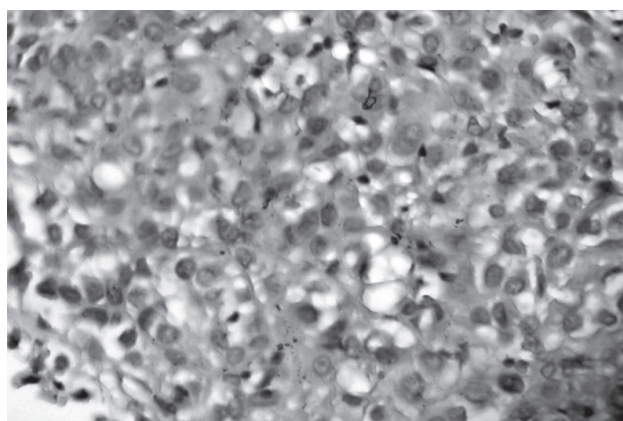


Fig. 3: Histology shows cohesive atypical mesothelial cells with vacuolated cytoplasm and prominent nucleoli.

Discussion

Pericardial mesothelioma is a rare malignant neoplasm that arises from the cells of the pericardium. Although primary pericardial mesothelioma represents less than 1% of all mesotheliomas, it accounts for 50% of all primary pericardial tumours.² The incidence of malignant pericardial involvement has been reported to be 0.15-21% of all patients with an underlying malignancy.⁷ Its incidence has a male-to-female ratio of 2:1.³ While many sources claim asbestos exposure to be a cause of pericardial mesothelioma, others

Primary pericardial mesothelioma

still believe that there is no definite association between the two. Pericardial mesothelioma might also occur post-radiotherapy although its preponderance in males makes radiotherapy a less likely risk factor since more females have breast cancer and are exposed to subsequent chest radiotherapy.⁴

Presentation can be at any age but the mean age is 46 years. Patients usually present with dyspnea, chest pain, cough, palpitations, fever and night sweats. Diffuse involvement of the pericardium may lead to symptoms of pericarditis, pleural effusion or cardiac tamponade. It may also cause heart failure due to myocardial infiltration. Primary mesothelioma may also resemble tuberculous pericarditis or intra-atrial myxomas. Distant metastasis, conduction block due to myocardial infiltration, and tumour embolism causing neurological deficits have also been reported⁷.

Histological type may be epithelial, spindle cell or mixed. Cytological examination of pericardial fluid is frequently non-diagnostic due to the difficulty in differentiating malignant mesothelioma cells from reactive cells.² Therefore, primary pericardial mesothelioma should be suspected in cases of hemorrhagic pericardial effusion even if the cytological evaluation of the fluid is negative for malignant cells.⁵ Chest radiography in patients with **pericardial** mesothelioma show cardiac enlargement, evidence of **pericardial** effusion, an irregular cardiac contour, or diffuse mediastinal enlargement. Chest CT demonstrates irregular, diffuse **pericardial** thickening and **pericardial** effusion. Magnetic resonance imaging has now emerged as the best test for demonstrating the nature and extent of the constrictive process, as well as its infiltration into the cardiac wall and great vessels.² It demonstrates encasement of the heart by a soft-tissue **pericardial** mass, along with an associated **pericardial** effusion.⁶

Pericardial mesothelioma is usually discovered late in the course of the disease and only palliative care is possible. Fine-needle aspiration to remove excess pericardial fluid and associated symptoms can be done. Surgical intervention with pericardiectomy and resection of the tumour is indicated for symptomatic relief in patients presenting with pericardial constriction. Radiotherapy is used as adjuvant treatment in patients with incomplete removal of the tumour. Chemotherapy with a combination of cisplatin or carboplatin plus gemcitabine or paclitaxel is used. Pemetrexed is a new anti-folate drug whose use in combination with a platin-drug has shown increased survival rates.⁷

Generally the prognosis of pericardial mesothelioma is poor with survival after diagnosis ranging from 6 weeks to 15

months.⁷

Preliminary management with photodynamic therapy and attempts at inhibiting the effects of growth factors, such as vascular endothelial growth factor and platelet-derived growth factor, and vaccine treatments are being explored. Early diagnosis and multidisciplinary patient care is essential for improved outcome. In the future, combined therapeutic strategies involving radical surgery, radiotherapy, adjuvant chemotherapy, and immunomodulation may have a role in the treatment of pericardial mesotheliomas.⁵

References

1. Eren NT, Akar AR. Pericardial Mesothelioma. Current treatment options in oncology. 2002 Oct;3(5):369-73.
2. Grebenc . Primary cardiac and pericardial neoplasms: radiologic-pathologic correlation. Radiographics. 2000;20:1073-103.
3. Kainuma S, Masai T, Yamauchi T, et al. Primary malignant pericardial mesothelioma presenting as pericardial constriction. Ann thoracic cardiovasc surgery. 2008;14396-8.
4. Kralstein J, Frishman W. Malignant pericardial diseases: diagnosis and treatment. Am Heart J. 1987;113:785-90.
5. Santos C, Montesinos J, Castañer E, et al. Primary pericardial mesothelioma. Lung Cancer. 2008;60:291-3.
6. Small GR, Nicolson M, Buchan K, et al. Pericardial malignant mesothelioma: a latent complication of radiotherapy? European Journal of Cardio-Thoracic Surgery. 2008;33:745-7.
7. Suman S, Schofield P, Large S. Primary pericardial mesothelioma presenting as pericardial constriction. Heart. 2004;90(1):e4.