Primary pericardial mesothelioma presenting as pericardial constriction: a case report

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CASE REPORT

A 19 year old male student presented with a six month history of chest pain, dyspnoea, and lethargy and was found on an echocardiogram to have a dilated left ventricle with a small pericardial effusion. Ramipril and a course of steroids were tried but serial echocardiograms showed a persistently thickened pericardium and slowly developing features of constriction. On computed tomography, a large mediastinal mass encasing the heart, along with para-aortic and paratracheal lymphadenopathy, was found. Right heart catheter studies showed equal pressures in all four chambers. His deteriorating clinical condition led to a pericardiectomy. Histology confirmed primary pericardial mesothelioma. The patient died soon after surgery.

Pericardial mesotheliomas are extremely rare and have an overall poor prognosis. Common clinical features are constrictive pericarditis, cardiac tamponade, and cardiac failure. Surgery can be curative in localised cases. Response to radiotherapy is poor. Pericardiectomy to alleviate symptoms and chemotherapy to reduce tumour mass are commonly used palliative measures.

DISCUSSION

Primary pericardial tumours are rare and can be benign (teratoma, fibroma, angioma, lipoma) or malignant (mesothelioma, sarcoma). Secondary tumours are more common, metastasising mostly from the lung, breast, melanomas, lymphoma, or leukaemia. The incidence of malignant pericardial involvement has been reported in the literature from 0.15–21% of all patients with an underlying malignancy. It is estimated that of all patients with malignant cardiac involvement, about 85% have pericardial involvement. Prognosis is poor with survival after diagnosis ranging from six weeks to 15 months. Pericardial mesothelioma is extremely rare, although it is the most common primary malignant pericardial tumour. Its incidence was < 0.0022% among 500 000 cases in a large necropsy study. Approximately 200 cases have been reported so far and only 25% of these were antemortem diagnoses. Pericardial mesothelioma can present as a localised or as a diffuse mass. Three histological types have been described: epithelial, spindle cell, and mixed.
relation to asbestos exposure but this is seen only in cases with coexistent asbestos related pleural disease. The onset of symptoms is usually insidious. Common clinical manifestations of pericardial mesothelioma are constrictive pericarditis, pericardial effusion, cardiac tamponade, and heart failure caused by myocardial infiltration. Further symptoms may arise due to compression of coronary arteries and local spread into the surrounding great vessels. Primary mesothelioma can also mimic tuberculous pericarditis or intra-atrial myxomas. Distant metastasis, conduction block due to myocardial infiltration, and tumour embolism causing neurological deficits have also been reported.

Pericardial fluid in pericardial mesothelioma can be difficult to aspirate. Echocardiography is the most commonly used initial investigative tool. Magnetic resonance imaging and CT are useful in showing the extent of involvement of contiguous structures and the degree of constriction. Other investigations such as immunohistochemistry, cytological examination, and high pericardial hyaluronic acid content of the pericardial aspirate can be diagnostic. The abnormal pericardium has also been known to take up gallium 67 extensively at scintigraphy.

Pericardial mesothelioma responds poorly to radiotherapy. Cyclical combination chemotherapy with doxorubicin, vincristine, and cyclophosphamide may reduce the tumour mass. Surgical resection can be curative in localised cases. Palliative treatment includes pericardectomy, which prevents cardiac tamponade and relieves constriction. Alternatively, a pericardial window can be formed to introduce chemotherapeutic agents. Areas of ongoing research include intracavitary chemotherapy and irradiation. Photodynamic treatment, inhibition of growth factors, vaccines, and adenoviral molecular chemotherapy are also being explored.

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