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Haemopericardium and cardiac tamponade complicating pulmonary lymphangioleiomyomatosis

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Abstract

A case of pulmonary lymphangioleiomyomatosis complicated by haemopericardium and cardiac tamponade is reported. This was successfully managed by creating a subdiaphragmatic extraperitoneal window.

Pulmonary lymphangioleiomyomatosis is a rare disease of premenopausal women characterised by benign proliferation of smooth muscle along lymphatic routes in the thorax and abdomen. Complications include pneumothorax, chylous pleural effusions, chylous ascites, haemoptysis, and cystic lung disease with respiratory failure and death.[1-3] Pericardial effusion causing cardiac tamponade has not been described in established disease. We describe a patient with lymphangioleiomyomatosis complicated by haemopericardium and cardiac tamponade that was successfully managed by creating a subdiaphragmatic extraperitoneal window.

Case report

A 33 year old woman presented in August 1988 with a one week history of cough and dyspnoea. She had had six pneumothoraces (three on each side) since 1981 during the course of three pregnancies and a further left sided pneumothorax five months previously when not pregnant, which had neccessitated a left thoracotomy and chemical pleurodesis. Lung biopsy at that time showed classical features of lymphangioleiomyomatosis.

When she was admitted in August 1988 clinical examination indicated cardiac tamponade with raised venous pressure, impalpable apex beat, distant heart sounds, and tender hepatomegealy. The chest radiograph showed pronounced cardiomegaly and cystic lung disease. Echocardiography showed a large pericardial effusion. At pericardiotomy under general anaesthesia 1500 ml of heavily blood stained fluid was drained and a subdiaphragmatic extraperitoneal window was formed.

Analysis of the pericardial fluid for malignant cells and infection (including tuberculosis) gave negative results. The lipid content was not measured because of contamination with blood. Pericardial biopsy showed non-specific inflammatory change only. The patient made a rapid postoperative recovery and was discharged two weeks later. When she was reviewed seven months later clinical examination and echocardiography showed no evidence of reaccumulation of pericardial fluid or of constrictive pericarditis.

Discussion

We report a case of classical lymphangioleiomyomatosis complicated by a large haemopericardium that led to cardiac tamponade. This complication has not been described in established lymphangioleiomyomatosis.

The term lymphangiomyoma was introduced by Cornog and Enterline in 1966,[2] who analysed 20 cases and gave the first definitive description of what has become known as lymphangiomyomatosis or, more precisely, lymphangioleiomyomatosis. Corrin *et al* described 23 cases in 1975[1] and Carrington *et al* a further six cases in 1977,[3] and the physiological, pathological, and radiological correlations are now well documented. Of the 49 patients reviewed by these three groups, none had had cardiac tamponade. One patient was found to have a pericardial effusion of 150 ml of chylous fluid as an incidental postmortem finding.[2 4] Miller *et al* reported a case of fatal chylopericardium caused by "hamartomatous lymphangiomyomatosis" in 1959.[5] Cornog and Enterline, however, were unable to confirm the histological findings.[2]

Our report of pericardial complications of lymphangioleiomyomatosis extends the range of documented clinical manifestations of this disease. Echocardiography is not performed routinely in these patients and no large series of postmortem examinations has been reported, so possibly pericardial collections in lymphangioleiomyomatosis are more common than is suspected.

The management of cadiac tamponade complicating lymphangioleiomyomatosis raises important considerations. Because the effusion is likely to reaccumulate simple aspiration is unlikely to suffice. A pleuropericardial window with free drainage of irritant blood and chyle into the thoracic cavity could further exacerbate already compromised lung function. To avoid these problems we elected to perform drainage by creating a subdiaphragmatic extraperitoneal window and this approach has so far been successful in our patient.

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