

Sarcoidosis presenting with chylothorax

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Abstract

A patient in whom chylothorax was the presenting feature of sarcoidosis is reported. Mediastinal lymphadenopathy was shown by computed tomographic scanning. Obstruction of the thoracic duct by enlarged lymph nodes or fibrosis is the probable cause of chylothorax in this case. The association of chylothorax and sarcoidosis is extremely rare.

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Chylothorax denotes a collection of milky chylous fluid in the pleural space with a fat content higher than that of plasma. It arises from obstruction of, or leakage from, the thoracic duct. The commonest causes are thoracic surgery, trauma, or malignancy obstructing the duct. There are numerous less common causes, including tuberculosis, filariasis, the yellow nail syndrome, and lymphangioleiomyomatosis.^{1,2} Only one case of sarcoidosis presenting with chylothorax has previously been reported.³

Case report

A 28 year old white man was referred from the diabetic clinic with a six month history of non-specific malaise and night sweats. He had developed increasingly severe exertional dyspnoea over the month preceding referral. He had suffered from insulin-dependent diabetes since the age of 14, and this was well controlled with no complications except mild background retinopathy.

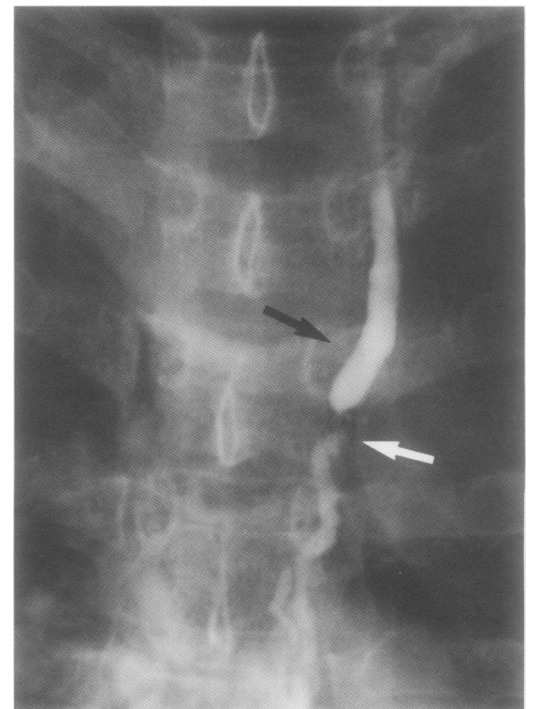
Physical examination revealed signs of a large right pleural effusion. Several small lymph nodes were palpable in the supraclavicular fossae, the anterior and posterior cervical areas, and the right axilla. A chest radiograph confirmed a large right subpulmonary pleural effusion. Thoracocentesis was performed and one litre of chylous fluid was aspirated. This contained predominantly mature lymphocytes with no evidence of malignancy. The triglyceride content of the fluid was 28.4 mmol/l with a plasma triglyceride concentration of 1.29 mmol/l, confirming that this was chyle. Culture for conventional organisms and mycobacteria was sterile. A full blood count was normal apart from moderate lymphopenia, and the biochemical profile (including serum levels of calcium and C-reactive protein) was also normal. Serum angiotensin converting enzyme

(ACE) activity was increased at 74 U/l (reference range 16-53 U/l).

Thoracic computed tomographic (CT) scanning with intravenous contrast medium demonstrated a large fluid collection in the right pleural space with partial atelectasis of the right lower lobe. Multiple lymph nodes were noted in the upper mediastinum, pre-azygous, pre-carinal, and para-aortic positions which were up to 15 mm in size. There was no hilar lymphadenopathy. An abdominal CT scan was normal. A lymphangiogram showed irregularity of the thoracic duct at the level of the carina and a segment of dilated duct above the aortic arch beyond which there was restricted flow of contrast medium (figure). There was some retrograde flow of contrast medium into the lymphatics around the posterior aspect of the right hemidiaphragm, but no leakage of contrast medium from the thoracic duct was seen.

Considerable nodularity and oedema of the mucosa was seen throughout the trachea and bronchi at bronchoscopic examination. Bronchial biopsy samples showed subepithelial granuloma formation with some necrosis. A supraclavicular lymph node biopsy was performed which yielded a lymph node almost entirely replaced by non-caseating granulomas. Acid fast bacilli were not seen in either specimen. A Heaf test produced a grade 1 reaction. A gallium scan showed abnormal uptake within the submandibular salivary glands, cervical, and inguinal lymph nodes. There was also an ill defined area of abnormal uptake within the left side of the posterior mediastinum.

He was treated initially with antituberculous chemotherapy, a low fat diet, and medium



Detail of lymphangiogram showing contrast medium within the thoracic duct flowing from bottom to top. There is irregularity and narrowing of the duct (white arrow), with a segment of dilated duct superiorly (dark arrow) below the clavicular head, beyond which there is restricted flow of contrast medium.

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chain triglyceride supplements. The chylothorax was drained. Four weeks after injection of the Kveim reagent, a skin biopsy sample showed granulomatous inflammation of the dermis, confirming the diagnosis of sarcoidosis. Mycobacterial cultures were negative after four weeks and prednisolone at a dose of 20 mg daily was substituted for Rifater (isoniazid, pyrazinamide, rifampicin) with a good clinical response (resolution of malaise and decrease in size of cervical nodes). After drainage of the pleural fluid a small reduction was seen in vital capacity (85% predicted) and carbon monoxide diffusing capacity (72%), which increased by 5% and 10%, respectively, after corticosteroid therapy. There was no further reaccumulation of chylous fluid and serum ACE activity returned to the normal range (29 U/l). After four months the prednisolone dose was reduced to 10 mg daily; two months later the patient remained in good health.

Discussion

Sarcoidosis is not a recognised cause of chylothorax, and the association has been reported in only three cases. There is only one previous description of a patient with chylothorax and mediastinal lymphadenopathy as a presenting feature of sarcoidosis.³ Chylothorax has also been reported in two patients with chronic sarcoidosis; it was recognised only at necropsy in one,⁴ and occurred with chylous ascites in the other.⁵ In both cases enlarged mediastinal lymph nodes with fibrosis and calcification were found at necropsy. The thoracic duct was clearly obstructed in one of these patients.

The thoracic duct is a posterior mediastinal structure lying between the aorta and the azygous vein. It passes behind the arch of the aorta, crossing to the left side of the spine before draining into the venous system at the confluence of the left subclavian and internal jugular veins. It is intimately associated with mediastinal lymph nodes which were found to be enlarged by CT scanning in our patient. We

also found increased uptake of gallium in the left side of the posterior mediastinum in the region of the thoracic duct. The mechanism of chylothorax formation in this patient is not certain, however. Compression of the thoracic duct by enlarged lymph nodes or obstruction due to fibrosis are possible mechanisms, supported by the lymphangiographic findings. However, ligation of the duct at a single point does not usually produce chylothorax.⁶ Extensive infiltration of the duct and lymphovenous anastomoses may be necessary to produce complete obstruction and accumulation of chyle in the pleural space.

Chylothorax complicates sarcoidosis considerably less frequently than other conditions involving intrathoracic nodes such as malignancy or tuberculosis. This may be due to lack of perinodal inflammation and the non-invasive nature of the disease in sarcoidosis, resulting in less extensive lymphatic obstruction.

Pleural effusions are uncommon but well described features of sarcoidosis and occur in fewer than 5% of patients.⁷ Pleural fluid collections in patients with sarcoidosis are therefore more likely to be due to an effusion than chylothorax. In patients with apparently idiopathic chylothorax (up to 25% of cases¹), sarcoidosis should be considered as a possible diagnosis. In a patient presenting with chylothorax and mediastinal lymphadenopathy, however, other causes of this combination such as lymphoma or tuberculosis should be ruled out before attributing it to sarcoidosis.

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