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Pleurectomy for chylothorax associated with intestinal lymphangiectasia

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We report an unusual case of bilateral chylothorax associated with intestinal lymphangiectasia, a rare condition characterised by dilated lymphatics in the small bowel and associated with a protein losing enteropathy. Bilateral pleurectomy in addition to thoracic duct ligation was required to control this problem satisfactorily. We have been unable to find a previous report of pleurectomy performed to control chylothorax. A case is made for early surgical intervention in this rare condition.

Case report

A 39 year old woman presented in March 1984 complaining of a six month history of weight loss, weakness, and abdominal distension. On examination she appeared anaemic and had asymmetrical lymphoedema of the lower limbs with abdominal proptosis.

Four years previously chylous ascites had been noted at hysterectomy for fibroids. After removal of varicose veins a lymphocele had developed in the left groin. Lymphangiography of the left leg at that time showed a paucity of lymphatics. Intestinal lymphangiectasia was suggested after a barium meal and confirmed by small bowel biopsy.

Over the year after presentation a chylous effusion developed, first on the left and then on the right side also. Control of the subsequent shortness of breath required frequent bilateral thoracocenteses. Over this period about 19 litres of chyle were removed. Despite a fat free, high protein, high carbohydrate diet, the chylothorax was not controlled and the serum protein concentration fell to 45 g/l.

In view of her failure to respond to conservative treatment over 12 months left thoractomy was undertaken. At operation lymph was noted to be oozing from multiple pinpoints on an abnormally thickened pleura. An apparently normal thoracic duct was identified. Pleurectomy was therefore undertaken. Three litres of chyle drained from the intercostal drains during the first four postoperative days but none subsequently. After removal of the chest drains there was no recurrence of the chylous effusion on the left.

Persistence of the right chylothorax led to right thoracotomy one month later. At operation the right pleura was similar in macroscopic appearance to the left, but the changes were less noticeable. Thus in addition to right pleurectomy the thoracic duct was ligated. Postoperative recovery was uneventful after minimal lymph drainage from the chest drains. Histological examination of both specimens of pleura showed abnormal, dilated lymphatics similar to those observed in the small bowel biopsy specimens.

There has been no recurrence of the chylous effusion and the patient remains symptom free 18 months after operation with normal serum protein values.

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Discussion

Primary intestinal lymphangiectasia is a rare condition of uncertain aetiology, characterised by dilated small bowel lymphatics and often complicated by anomalous lymphatics elsewhere, typically in the limbs.[1] Patients usually present before the age of 30 years with a protein losing enteropathy and asymmetrical lymphodema.[2] Barium meal appearances are characteristic, with mucosal odema and "puddling" of the radiocontrast material around the enlarged lymphatics of the small bowel. The diagnosis is confirmed by identifying characteristic dilation of mucosal lacteals in jejunal biopsy specimens. Dilatation of the thoracic lymphatics, as in this case, is unusual, complicating only two of a series of 18 cases reported by Strober.[3] Chylothorax from any cause is rare, probably most commonly seen after thoracic surgery,[4] complicating 0·6% of all thoracic procedures.[2] Blunt trauma occurring in thoracic injury may cause rupture or avulsion of the duct.[5] Obstruction of the duct and subsequent chylothorax may arise as a result of extrinsic compression by bronchial tumours and tuberculous nodes or by intrinsic lesions such as tumours of the duct, congenital malformations, or thrombosis of the great veins.[6] Chylothorax resulting from abnormal lymph drainage of the pleura, as in this case, is rarely observed.

Untreated, chronic chylothorax has a mortality rate of 50% over a period of months.[7] Persistence of the effusion is associated with protein malnutrition and immune deficiency.[3] Apart from measures directed towards diagnosis and treatment of any primary cause, treatment usually rests on intercostal drainage of the chylous effusion coupled with dietary regulation to replace protein and carbohydrate losses and reduce fat intake. Intravenous feeding may be required to reduce intestinal absorption of fat and thus decrease thoracic duct flow.[2]

Occasionally, pleurodesis may be undertaken. Persistence of the effusion despite conservative treatment requires surgical intervention. It is not clear whether pleurectomy countered the chylous effusions by obliterating the pleural space or by removing the primary cause, an abnormal pleura. Coupled with thoracic duct ligation (first performed by Lampson in 1948[7]) this certainly would be expected to

worsen the protein losing enteropathy, but it did not have this effect here or in other cases.[3] It has been proposed that chest wall lymphatics dilate to accommodate the accumulating lymph, providing an alternative route for lymph drainage.

The case reported here demonstrated many of the features of intestinal lymphangiectasia complicated by chylothorax. Twelve months of conservative management failed to solve the problem. Surgical intervention was therefore necessary. The appearance of generalised lymphatic leakage from abnormal pleural lymphatics led to pleurectomy. This was adequate on the left, but on the right the pleural changes were less severe and therefore pleurectomy was coupled with the more standard procedure of thoracic duct ligation. In the light of this experience, we would suggest that a short and defined period of conservative treatment is worthwhile but that surgical treatment should follow promptly if this fails.

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