CASE REPORT

Primary Lymphangiomyomatosis with Chylous Ascites

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Summary

Primary lymphangiomyomatosis is a benign tumour of lymphatic channels and lymph nodes, clinically manifested by chylous ascites. This disease is usually progressive and unresponsive to surgery, chemotherapy or irradiation. A case of a 36-year-old lady with chylous ascites due to underlying primary lymphangiomyomatosis is reported.

Key Words: Lymphangiomyomatosis, Chylous ascites

Case Report

A 36-year-old Indian lady was readmitted in April 1993 to the Universiti Kebangsaan Malaysia surgical unit of Kuala Lumpur Hospital because of complications associated with chylous ascites. When she was nine years old, the patient underwent an excision of cystic hygroma over the left supraclavicular region. She was apparently well until October 1989 (23 years later), when she presented with sudden onset of severe lower abdominal pain. She underwent emergency laparotomy in Kuantan General Hospital for suspected twisted ovarian cyst. At operation, the ovaries were found to be normal. However, there were numerous cysts in the mesentery, liver and spleen. A few cysts were biopsied and the abdomen was closed. Two weeks after the laparotomy, she started to develop progressive abdominal distension.

She was referred to Universiti Malaya Hospital in March 1990. Subsequently she underwent an exploratory laparotomy, with similar operative findings. A cystic spleen was removed and mesenteric lymph nodes excised for biopsy. The lesion was reported as a benign lymphangiomatosis with no evidence of malignancy.

In May 1990, she was first seen and admitted to the Universiti Kebangsaan Malaysia surgical unit at Kuala Lumpur Hospital. Clinically she had gross ascites and a cystic mass at the left supraclavicular region. She was neither dyspnoeic nor orthopnoeic. Diagnostic paracentesis performed yielded a milky white fluid with total protein of 3.9gm/100ml and globulin of 2.2gm/100ml. Cytology of the fluid revealed numerous lymphocytes.

Computerised tomography of thorax and abdomen was performed at this admission. It revealed an ill defined mass surrounding the left carotid and left subclavian arteries extending inferiorly to the level of the aortic arch with abdominal finding of ascites and numerous liver cysts.

She underwent local excision of the lymphangiomatous cysts of the left supraclavicular region without thoracotomy. The lymphangiomatous lesion in the abdomen was not suitable for complete resection. After that, she underwent two shunt procedures, a Denver peritoneovenous shunt (May 1990) and a right pleuroperitoneal shunt (Jan 1992). She had spontaneous resolution of ascites in June 1992.

Unfortunately, in February 1993 she presented to Tawakal Hospital with high grade fever and abdominal pain. Her problem was attributed to shunt complication manifested by peritonitis and septicaemia. Peritoneal fluid and blood culture and sensitivity results were unknown. She responded with a course of antibiotics and had both the shunts removed.

In April 1993, she was febrile again due to right empyema thoracis, and was admitted to Universiti Kebangsaan Malaysia surgical unit. She was treated with intravenous ceftazidime and a chest tube drainage. The pus which was sent for culture and sensitivity revealed no growth. A week after removal of chest tube, she was febrile again. She was treated with a course of intravenous imipenem based on the chest tube culture and sensitivity which grew *Pseudomonas* sp.

She improved clinically and radiologically. She is still on follow-up at the surgical outpatient clinic and is currently on a medium chain triglycerides diet. Since the ascites remains the same, no further drainage procedure was planned. However if she continues to complain of further distension with respiratory embarrassment, repeated paracentesis or shunt reinsertion may be helpful symptomatically.

Discussion

Chylous ascites is a clinical and pathophysiological manifestation of an underlying disease[1]. It is an uncommon clinical entity and has been recognised since the 17th century. There has been less than 200 reported cases.

Although there are many causes for chylous ascites, a cause can be found in less than 30 per cent of the cases. In adults, it is most commonly due to mechanical obstruction of the lymphatic system due to thoracic and abdominal neoplasms (such as Hodgkin's lymphoma or lymphosarcoma), inflammatory processes (such as tuberculous lymphadenitis and/or filariasis), or aortic aneurysm. Traumatic rupture of the lymphatic system can happen as a result of blunt trauma or surgery at the base of mesentery or retroperitoneum[2].

Lymphangiomyomatosis is a rare condition which can cause chylous ascites. This condition was first reported in 1955 by Laipply[3] and there has been 84 reported cases in literature. It is a benign tumour of lymphatic channels and lymph nodes. It has been postulated that it is a lesion of "hamartomatous" origin and metastases have never been reported. It occurs exclusively in women in their reproductive years and often involves lung, retroperitoneal and mediastinal tissues. This lesion is probably a result of the failure of a group of lymphatic vessels to communicate with the main lymphatic system.

The most prominent pathologic finding is the proliferation of atypical smooth muscles most often within the wall of the thoracic duct, lymphatics and lymph nodes near the skeletal axis. This clinical entity will not regress unless complete excison is performed.

Treatment includes supportive measure to relieve the symptoms caused by chylous ascites and treatment of underlying cause. Paracentesis is the most commonly used measure. Repeated paracentesis can result in serious complication such as pneumothorax, empyemas, peritonitis and incarceration of a hernia with small bowel obstruction.

Low-fat diet with medium-chain triglyceride (MCT) supplementation was popularised in 1964. The aim of this diet is to diminish the flow of chyle from the lymphatics of patients with chylous ascites. All patients with chylous ascites should be given this diet because a significant percentage of patients have been relieved with such a simple measure.

If the patient does not respond to the conservative management, peritoneovenous shunting is recommended. Some authors do not agree on using a shunt procedure because of the high complication rate such as disseminated intravascular coagulation, cardiac failure, arrhythmias, peritonitis, septicaemia and shunt blockage.

Surgery, irradiation and chemotherapy are not very effective in controlling the progressive nature of lymphangiomyomatosis. Once the chylous ascites has occurred, complete resection of the lesion is not feasible. Supportive measures available to relieve the ascites include paracentesis, medium chain triglycerides diet and shunt.

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