Case Report

ISSN: 1735-0344 *Tanaffos 2015; 14(2): 153-155*

TANAFFOS

Lymphangiomatosis: The Cause of Refractory Pleural Effusion in a Patient with Lupus Erythematosus

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Received: 21 November 2014
Accepted: 20 January 2015

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Mediastinal cavernous lymphangioma is a rare mediastinal lesion and its association with lupus erythematosus has not yet been reported in the literature. We present a 25 year-old female with lupus erythematosus who had bilateral massive refractory and recurrent pleural effusion as well as ascites for a long period of time. During surgery, a huge multicystic lesion with a thick wall, covering the entire parietal and visceral pleura was found, which was subsequently proven to be a cystic cavernous lymphangioma.

Key words: Pleural effusion; Mediastinum; Lupus Erythematosus

INTRODUCTION

Although pleuritis is a common manifestation of lupus erythematosus, the effusion is rarely massive and only few cases of refractory massive pleural effusion have been reported and even in such cases systemic or local therapy has been proven successful (1). Mediastinal cavernous lymphangioma is a rare mediastinal lesion and its association with lupus erythematosus has not been previously reported. Herein, we report a case of a young female with lupus erythematosus who suffered bilateral massive refractory pleural effusion for a long period of time. During surgery, she was found to have a huge mediastinal cavernous lymphangioma.

CASE SUMMARIES

A 29-year- old female presented to our center with bilateral refractory pleural effusion and ascites with a previous diagnosis of lupus erythematosus characterized by arthralgia, anemia, leucopenia, conjunctivitis, hepatosplenomegaly, strongly positive anti-nuclear antibody and elevated level of anti-ds DNA. She had developed massive pleural effusion of the left pleura seven years earlier, which extended to the right pleura subsequently causing significant dyspnea and non-productive cough. The patient had also developed ascites in the peritoneum as well as mild pericardial effusion five years earlier.

Pleural effusion was exudative at both sides and showed low C3 and C4 levels. It was positive for ANA and anti ds DNA and negative for adenosine deaminase. Cytology report revealed numerous mixed inflammatory cells with predominance of histiocytes and lymphocytes. No malignant cells were observed. Ascites fluid was semiturbid and exudative with 161 mg/dL triglycerides, low C3 and C4 levels and was positive for ANA and anti-ds DNA.

Her previous medications including prednisolone, azathioprine, chloroquine, cyclophosphamide and IV immunoglobulin caused no relief. Her medical record indicated that she had first undergone thoracentesis every three weeks for dyspnea, which had been increased to every week within a few months. She also had a history of left side chest tube insertion and pleurodesis with bleomycin 5 years ago; however, the effusion was found to be refractory and about 250 cc of fluid was collected with every attempt from the left side and 700-800 cc from the right side.

Due to having a 6-year history of refractory pleural effusion at the time of admission, we decided to perform video assisted thoracoscopic surgery (VATS) and Talc pleurodesis. A multicystic lesion like a honeycomb was seen on camera. For more exploration, thoracotomy was carried out and a huge multicystic lesion with a thick wall, covering the entire parietal and visceral pleura was found. There were several septations and different sized cavities. The largest cavity was in the subpulmonic position and was retrospectively found to be the site of numerous aspirations during the past seven years. The lesion was completely excised.

The pathologic report was a benign cystic lesion with thick-wall structures having many variable small to large sized lymphatic channels lined by one layer of thin endothelium as well as small capillary sized blood vessels with hemorrhage, inflammation and reactive fibrosis. The diagnosis of cystic cavernous lymphangioma (hygroma) was then confirmed.

After surgery, the patient's respiratory symptoms ameliorated significantly and there was no need for further fluid aspiration even from the right side. At the last follow-up 4 years after thoracotomy, there was no recurrence.

Six months later the patient was operated for her recurrent chylous ascites, which required aspiration of approximately 1500 cc of peritoneal fluid every other day. Upon laparotomy, multiple cystic lesions were observed in the lesser sac and one in omentum, which were all excised. The pathologic report was consistent with mesothelial cysts.

On follow-up, although some fluid still existed in the peritoneum there was no need for further peritoneocentesis.

DISCUSSION

Serositis is common in lupus erythematosus and pleural effusion is seen in almost 50% of patients with systemic lupus erythematosus. The effusion is usually exudative and ANA, Anti ds DNA and LE cells can be observed in the pleural fluid (2). It is usually bilateral and mild to moderate. Massive recurrent refractory pleural effusion is rare in lupus erythematosus (3). Although different systemic and local therapies have shown to be successful in managing refractory effusion (1), Talc pleurodesis is thought to be the most effective modality in controlling the pleural effusion (1).

In cases of ascites, systemic therapies have also been shown to be successful (4). In our patient, the effusion was primarily thought to be only due to the underlying disease. However, upon presentation with refractoriness to both systemic and local therapies and frustration with her weekly thoracentesis, we chose to perform VATS for thoracoscopic Talc pleurodesis and surprisingly other pathologies were observed, a cavernous lymphangioma in her chest as well as mesothelial cysts which were consistent with her systemic lymphangiomatosis.

Lymphangiomas, particularly the cavernous type, are rare mediastinal lesions and usually present in childhood. In adults, the diagnosis is usually made postoperatively as the diagnosis by radiographic imaging can be difficult especially when not suspected (5). On CT scan, the most common presentation is a homogenous cystic lesion, although the mass is known to be multiloculated, the septates are not visualized (5, 6). Lymphangiomas are dilated lymphatic channels, which are lined by endothelium and can contain fluid, which can be clear or chylous. They are divided into three groups of capillary, cavernous and cystic types (5, 7). Among the three pathologic types, the cavernous type is quite rare.

Mediastinal lymphangiomas are usually discovered incidentally on X-rays; however, when local growth occurs they can compress the adjacent structures and cause dyspnea, dysphagia, cough, hemoptysis or chest pain (8). In order to prevent their local growth, complete surgical excision of these benign lesions is recommended; although it can be difficult in cavernous type lymphangioma as it dose not have a discrete margin (7) and cases of recurrence have been seen when complete removal has not been achieved.

To our knowledge, this is the first reported case of cavernous lymphangioma in a patient with lupus erythematosus. The diagnosis was difficult as the patient's dyspnea and recurrent effusions were attributed to her underlying disease particularly due to the presence of low complement levels and also anti ds DNA and ANA in the pleural fluid. However, it is important to emphasize that in cases where no amelioration is observed with systemic and local therapy, presence of other rare lesions (which can be masked by the effusion) should be considered. We completely excised the lymphangioma in our patient with no recurrence after 4 years.

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