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Case Presentation and Imaging Review of an Adult Mediastinal Cavernous Lymphangioma; Case Report

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ABSTRAST

Adult mediastinal cavernous lymphangioma is a benign rare lesion originating from lymphatic system. It is usually asymptomatic. We have presented a 28-year-old man with dyspnea on exertion and palpitation. A mediastinal mass was discovered on CXR. CT findings revealed "non enhancing", "smoothly marginated", and "multilocular" mediastinal mass which had extended to superior mediastinum. Finally, pathological examination of the surgical sample indicated "cavernous lymphangioma". ***(Tanaffos 2003; 2(8): 71-76)***

Key words: *Adult, Mediastinal lymphangioma, Imaging, Cavernous lymphangioma*

INTRODUCTION

Mediastinal lymphangioma is a histologically benign proliferation of interconnecting lymphatic vessels and sacs; it may grow in an infiltrative fashion (1, 2). Mediastinal lymphangioma is rare, (less than 1%) and when confined to the mediastinum, they are usually asymptomatic (3). Fewer than 10% of cases occur in the posterior mediastinum. Lymphangioma usually appears during childhood. (90% of cases present at< 2 yr. of age) (4). Above all, cavernous type of mediastinal lymphangioma is very rare (5). We have presented a case of an adult symptomatic and pure mediastinal cavernous lymphangioma involving all the mediastinal compartments.

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CASE PRESENTATION

A 28-year-old nonsmoker man was admitted with dyspnea on exertion and palpitation for 1.5 month. Exacerbation of dyspnea had occurred in the last 2 weeks before admission. He did not have weight loss, chest pain, cough, and sputum .He had a history of vertigo since 5 years before and, therefore, had used phenobarbital and propranolol according to the physician's prescription. Social, occupational, and family history were unremarkable and no history of malignancy was found in his family as well. Physical examinations were normal. In laboratory findings, complete blood count (CBC), differentiation and

arterial blood gas (ABG) were within normal limits. Pulmonary function test was in normal range.

He was admitted in our center for more evaluations.

Posteroanterior chest radiographs obtained on admission showed a large mediastinal mass (Fig 1).

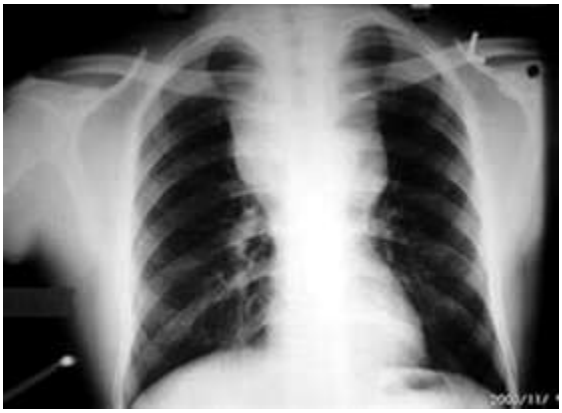

Figure 1. Posteroanterior (PA) view Chest x- ray shows a mediastinal widening and mass.

Thorax CT-scan with and without contrast showed non-enhancing, smoothly marginated, multilocular mass extending to superior mediastinum with lower attenuation than soft tissue involving all mediastinal compartments, enveloping mediastinal structures with mild displacement, and compression of vessels (Fig 2A and Fig 2B).

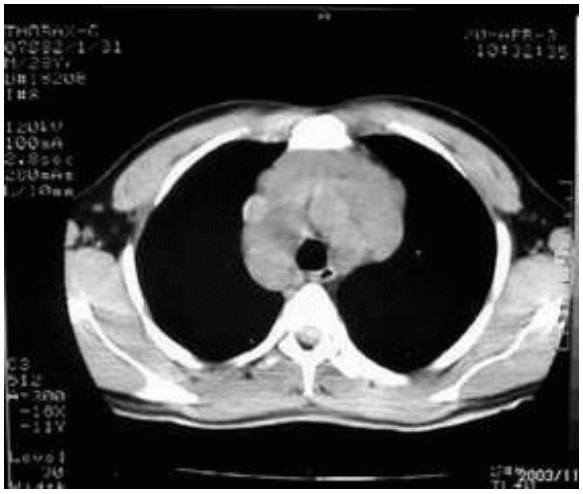

Figure 2 A. Chest CT scan without contrast shows mediastinal mass with lobulated margin.

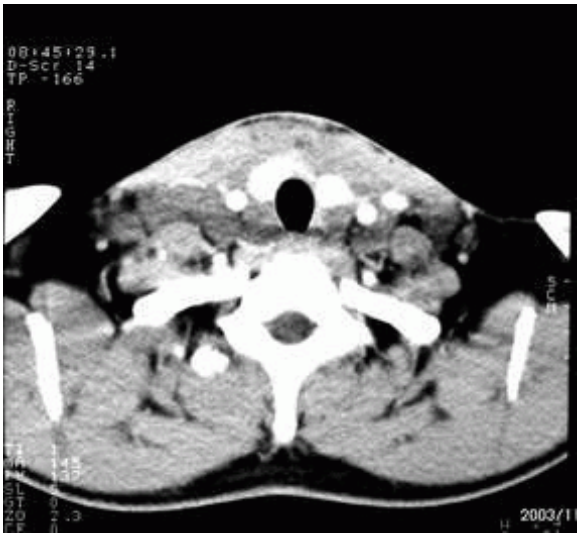

Figure 2 B. CT scan without contrast at basal neck level. No extension into neck is shown.

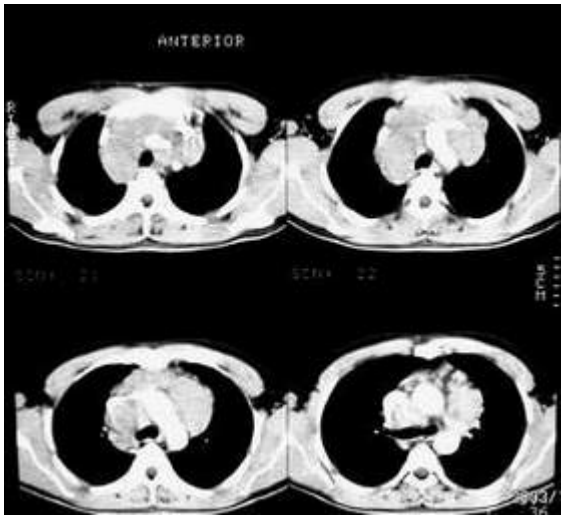

Figure 2 C. Chest CT scan with contrast. Mass involves all mediastinal compartments and mediastinal structures are enveloped. Also, mild displacement and compression of vessels is seen.

Right posterolateral thoracotomy was performed; the mass resected and sent for pathologic examinations.

Pathologic examination reported variable sized spaces with intervening lymphoid tissue and smooth muscle cells. The spaces were lined by a flat uniform lining. The lymphoid tissue was composed of small

lymphocytes and occasional follicles. Immunohistochemistry showed positive reaction for CD31 and thrombomodulin and negative reaction for cytokeratin cocktail. Smooth muscle cells revealed positive reaction for SMA.

At last, the pathological diagnosis was "Cavernous Lymphangioma".

DISCUSSION

The clinical course, radiological findings and pathological confirmation of the case described are compatible with mediastinal cavernous lymphangioma. Lymphangiomas are benign tumors of mesodermal origin consisting of proliferating lymph vessels of varying size. Pathologically they have been classified as simplex (capillary lymphangioma), cavernous, and cystic (cystic hygroma)(6). Cavernous type is microscopic thin-walled lymphatic channels with associated stroma that consist of varying amount of spindle-shaped smooth muscle cells ,collagen bundles, fibroblasts, and lymphocytes (5,7,8)similar to our pathological report. Cavernous lymphangiomas of the thorax are extremely rare (9).

Approximately, 1% of all lymphangiomas are confined to the chest (9). Unlike our case, lymphangioma is typically a tumor of very young children. Most lymphangiomas are discovered in fetuses, neonates, or young children. Fifty percent are present at birth, and 90% are discovered by 2 years of age (1, 10). Ninety-five percent involve the neck or axilla, and 10% extend into the superior aspect of the anterior mediastinum (11) or less commonly other mediastinal compartments (12). Rarely, lymphangiomas occur as primary mediastinal tumors in adults (2, 10).

To our knowledge, in world wide medical literature, few cases of adult mediastinal cavernous lymphangioma have been reported. From 5 cases of lymphangiomas of the mediastinum reported by Oshikiri et al., one case that was diagnosed by pathological examination was cavernous type (5). Also, from 9 cases in a retrospective 12-year period study conducted by Charruau et al., two cases were cavernous type adult mediastinal lymphangioma (13). In Japanese medical literature, only 7 cases of cavernous mediastinal lymphangioma were reported (14). Most of them were infants or young children.

Although the majority of lymphangiomas present in the first 2 years of life, there has recently been an increased recognition of lymphangiomas in adults (15, 16). Patients can complain of cough and dyspnea (from extrinsic compression of airways), steridor, hemoptysis, Horner's syndrome, dysphagia, superior vena cava syndrome, constrictive pericarditis, phrenic nerve palsy, or of symptoms related to a secondary infected lymphangioma (17-20).

Radiological imaging can help in differential diagnosis of mediastinal masses and, also, in determination of the exact location, extension and content of mediastinal lymphangiomas. There are few literatures describing the evaluation of imaging and radiological findings in mediastinal lymphangiomas especially cavernous type. Although, in 1990, Wernecke et al. in a study showed the high sensitivity of ultra sonography in diagnosis of mediastinal tumors in certain mediastinal regions, using CT-scan is still the golden standard to detect and evaluate mediastinal tumors (21). In 1997, Pui et al. in a study assessed ultrasound, CT, and MR images of 18 patients and concluded that ultrasound, CT, and MR imaging are valuable for evaluating lymphangiomas (22). Lymphangioma may invade nearby tissues and organs; moreover, computed tomography can determine the morphology of lymphangiomas, their exact location and their relationship to other organs (23). Miyake et al. presented CT results in three cases of mediastinal lymphangioma. One showed a

well-defined, multi-locular, water-density mass enveloping mediastinal structures, without their displacement or compression; one presented as well-defined, homogenous, water density mass in the right paratracheal region; and one was associated with hemorrhage into cysts and significant increase in size over 5 years. According to their comment, CT is useful for diagnosis and evaluation of extent or contents of mediastinal lymphangioma (24).

Shaffer and colleagues studied 19 cases of adult thoracic lymphangioma in three institutions in US. Their results showed that the most common CT appearance was a smoothly marginated cystic mass. Unusual features included calcification, speculated margins, and homogeneous soft tissue attenuation. The majority of cases were located in anterior or superior mediastinum. Unusual locations included the pericardium, pulmonary hilum, and pulmonary parenchyma. Signal characteristics on MR images varied.(11)

Finally, to detect the relationship between radiological findings and pathological features in any type of adult mediastinal lymphangiomas especially cavernous type like our presented case, in the year 2000, Charruau et al. in a retrospective study of adult's mediastinal lymphangiomas identified 9 cases over a 12-year period. The CT, MR, and pathological findings were reviewed. According to their results, they concluded that CT appearance of mediastinal lymphangioma was variable depending on the pathologic type. The less frequent cavernous type (2 cases) can be suggested based on a multiseptated and loculated mass on CT and/or MR examination (13).

Briefly, we presented this case because adult mediastinal lymphangiomas are rare tumors especially the cavernous type. Unlike the case presented above, and we mentioned earlier, they are usually asymptomatic. Imaging methods and its validity for diagnosis of mediastinal lymphangioma are still controversial. So we found it a considerable case to be reported.

In order to determine more exact radiological characteristics of mediastinal lymphangioma, more cases must be reported and reviewed considering the radiological findings.

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