Primary intrathymic lymphangioma in an infant

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ABSTRACT

Lymphangiomas are relatively uncommon lesions of the lymphatic channels which can arise in virtually any part of the body. Although the most common site is the head/neck region, they could be found in the mediastinum. If mediastinal lymphangiomas are said to be rare, the thymic subentity is even scanty. We describe one case of mediastinal lymphangioma with a true intrathymic localization, which to our knowledge has been reported just once in the literature. This case report elucidates the surgical management of the first lymphangioma reported in an infant.

Keywords: Mediastinal cystic mass, pediatric mediastinal tumor, primary thymic lymphangioma

INTRODUCTION

Lymphangiomas are relatively uncommon lesions of the lymphatic channels, considered as true neoplasms by some and as malformations by others, mostly occurring during the first 2 years of life.[1] Although lymphangiomas can arise in virtually any part of the body supplied by the lymphatic network, the most common localizations are the head-neck and axillary regions, with only sporadic occurrences reported in other sites such as heart, lung, chest wall, breast, greater omentum, retroperitoneum, small-bowel mesentery, colon, cauda equina, and the extremities.[1] The mediastinum is a relatively rare location where around twenty cases have been documented in the literature.[2] Mediastinal lymphangioma is a rare neoplasm, accounting for 0.7%–4.5% of all mediastinal tumors.[3] While a majority of cystic lymphangiomas are diagnosed under the age of 5 years, they are rarely found in adulthood.[4] Most individuals with mediastinal lymphangiomas are asymptomatic or can present with chest pain, cough, dysphagia, or dyspnea. Here, we describe a case of successful surgical management of a true intrathymic lymphangioma.

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CASE REPORT

A 16-month-old male infant was brought to us with complaints of dyspnea on exertion and a history of recurrent bouts of fever since 2 months of age. Stable at presentation with normal vital signs, he was noted to possess a mass in the left hemithorax. Air entry on the left side of the chest was reduced. Chest roentgenogram showed opacity in the left hemithorax, with mediastinal shift to the right [Figure 1a].

A transthoracic echo assessment showed a mediastinal mass compressing the left ventricle. Normal biventricular function was noted at the same sitting. Computed tomography (CT) of the thorax revealed a large cystic lesion measuring 86 mm × 71 mm × 53 mm in the left hemithorax closely abutting the thymus gland and the left ventricle with two septations within the lesion causing a mass effect and mediastinal shift to the right [Figure 1b and c]. After routine preoperative evaluation, a working diagnosis of mediastinal mass was made and the patient was taken up for surgery.

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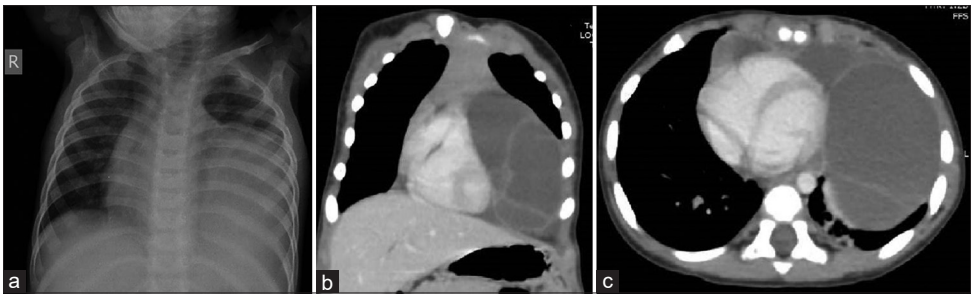

Figure 1: (a) Preoperative chest X-ray showing mass in the left hemithorax, (b) computed tomography thorax showing mass in the left hemithorax compressing the left ventricle, (c) computed tomography thorax showing two septations inside the mass

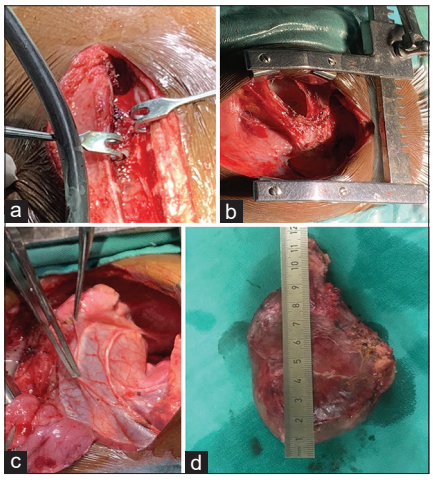

Figure 2: (a) Mass densely adherent to the posterior part of the sternum, (b) one of the ruptured locules on opening sternum, (c) mass densely adherent to the phrenic nerve, (d) mass after complete excision

Mediastinal access was achieved through midline sternotomy. While three large sacs of fluid were noted on examination, one sac (with sanguineous brown fluid content) was ruptured during sternotomy as it was strongly adherent to the posterior wall of the sternum. The remaining mass measured 95 mm × 50 mm × 70 mm. The mass was densely adherent to the surrounding structures including the phrenic nerve and the diaphragm [Figure 2]. The left lung was temporarily deflated and the cyst was dissected from the pericardiophrenic recess and the diaphragm.

RESULTS

The patient tolerated the procedure well and was extubated the same evening. With an uneventful postoperative stay, he was discharged on the third postoperative day. The final histopathology report revealed thymic follicular hyperplasia with flattened epithelium and eosinophilic material within the cystic lumen and lymphoid aggregates with germinal center formation in the wall suggestive of lymphangioma.

DISCUSSION

Lymphangiomas are unusual vascular lesions of uncertain origin, often a consequence from sequestrations of lymphatic tissue failing to communicate normally with the lymphatic system, although with a neoplastic or hamartomatous origin in some cases.[1,5] They may also represent embryologic remnants of lymphatic tissue which arise either from failure to connection to efferent channels or sequestration of portions of lymph sacs during development. They are usually noted at the sites of primordial lymph sacs in fetuses, neonates, or young children. The first detailed description of a lymphangioma was rendered by Redenbacker in 1828.[5]

Although the head-neck and axillary regions are the more common areas of localizations, a small number of lymphangiomas have also been noted in adult patients in other zones such as heart, lungs, chest wall, breast, greater omentum, small-bowel mesentery, retroperitoneum, colon, cauda equina, mediastinum, and other extremities.[2]

Primary lymphangiomas of the mediastinum are rare, with fewer than twenty cases reported in the literature so far. They are often found incidentally as slow-growing mediastinal masses detected on imaging modalities ranging from chest X-rays to sectional CT or magnetic resonance scans. Large lesions in symptomatic adult patients can present as chest discomfort, cough, hoarseness, angina, and dyspnea, which can grow rapidly as a result of acute hemorrhage within the lesion.[5]

The pathologic differential diagnosis of cystic lymphangiomas from cavernous hemangiomas is based on the features of the cyst contents and the presence of smooth muscle and lymphoid tissue in the walls of the cystic spaces of the former.

After a diligent literature search, our case represents the second description of true thymic lymphangioma with unequivocal intrathymic localization after Licci *et al.* reported the lesion in a 66-year-old male in 2014.[5]

This case of a true thymic lymphangioma in a child is the first of its kind.

Thymic lymphangioma should always be accounted for in the differential diagnosis of mediastinal cystic masses in all patients. It is a biologically benign vascular proliferation and may be asymptomatic in most presentations. Total surgical excision remains the gold standard in most cases to prevent complications which may arise from compression on vital structures, as lymphangiomas may grow and surround large blood vessels, airways, and other mediastinal structures. Histopathology is vital to ascertain the diagnosis, and proceed with any further intervention, in cases of malignancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published, and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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