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Cystic Hygroma of mediastinum presenting with Superior Vena Cava(SVC) Syndrome

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Abstract:
Cystic hygroma is a rare vascular cystic tumor of lymphatic origin. Mostly it occurs in neck. But rarely it can occur also in anterior or superior mediastinum. Presentation of tumor usually gets delayed as most of them are asymptomatic. We present a case of 9 year male child with complains of dyspnoea on exertion and swelling of face. X-ray chest revealed widening of the mediastinum. Computed Tomography(CT) scan showed a large heterogeneous mass with multiple cystic spaces in anterior mediastinum and was compressing superior vena cava. Surgical excision of the mass was performed. Histopathological examination of mass reported as cystic lymhangioma of superior mediastinum.

Key Words:*Cystic Hygroma, Mediastinum, Superior Vena Cava Syndrome.*

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I. Introduction

Cystic hygroma, also known as cystic lymhangioma is a rare vascular cystic tumor of lymphatic origin. Hygroma, derived from Greek hygros(moist) and oma(tumor) (1). Usually it is multiloculated and benign in nature. Most common site of origin is neck(75%),next most common is axilla (20%). Mediastinal cystic hygroma constitute 1% of this rare tumor (2).Though asymptomatic in most of times, it can present with dyspnoea, cough, dysphagia or vascular compression (3). Often it can be an extension of cystic hygroma of neck.

II. Case Report

A 9 year male child presented with complains of dyspnoea on exertion with respiratory distress. On examination neck and face were swollen. X-ray chest revealed widening of the mediastinum. Computed Tomography (CT) scan showed 8×6 centimeter large heterogeneous mass with multiple cystic spaces in superior mediastinum, compressing superior vena cava(Fig.1a) and extending to anterior mediastinum and towards diaphragm along left paracardiac boarder (Fig.1b). Fine needle aspiration cytology was inconclusive .AlphaFeto Protein(AFP) and Beta Human Chorionic Gonadotrophin(Beta-HCG) hormones were normal.

Surgery was planned under general anaesthesia. Midsternotomy was performed. Mass filled with multiple clear fluid cavities was identified(Fig.2a). Careful dissection from SVC and innominate vein was performed. Extension along left paracardiac boarder was also dissected out. Complete excision of the mass was performed (Fig.2b). Patient had an uneventful recovery and was discharged on 4th postoperative day.

Histopathology report revealed dilated lymphatic channels with eosinophillic material in the lumen and lymphatics with endothelial lining (Fig.3). Child was doing well with complete regression of facial swelling and no respiratory difficulty at six month post operative follow up.

III. Discussion:

Cystic hygroma is a developmental malformation of lymphatic system .The most common accepted theory is ,a portion of jugular lymph sac remain separated and fails to establish subsequent connection with venous system (4) . So mostly it occurs in neck. But it can occur anywhere in the path of lymphatic development. Rare incidence of cystic hygroma in posterior mediastinum (5),abdomen and retroperitoneum (6) has been reported.

Most of literatures about mediastinal cystic hygroma are case reports due to its rarity. Usually it gets diagnosed by two years of age, if it is on body surface. But in mediastinum , presentation may get delayed even

up to adulthood as it is asymptomatic in most of cases(2).Some of these may become very giant and may remain occult and present as massive effusion on rupture(7). Mediastinal cystic hygroma presenting with SVC syndrome is very rare and not reported in literature. Preoperative diagnosis is almost impossible though CT scan and magnetic resonance imaging (MRI) features are well described (8).

Other cystic masses considered for differential diagnosis include teratoma, thymic cyst, necrotic tumor, pericardial or bronchogenic cyst, often goiter (9).

Complete excision is possible in most of times though sometimes may be difficult. If completely not excised recurrence is very likely.

Other less invasive therapies like local irradiation, laser ablation and injection of sclerosing agents are described with suboptimal results (10).

IV. Conclusion

Mediastinal cystic hygroma is a very rare tumor of lymphatic origin. Most of times it remains asymptomatic, but can present with symptoms of dyspnoea or SVC syndrome. Preoperative diagnosis is sometimes difficult. Complete excision should be aimed at; otherwise recurrence chances will be there.

Consent from the patient was taken. All ethical compliances taken care of. There was no funding for this research.

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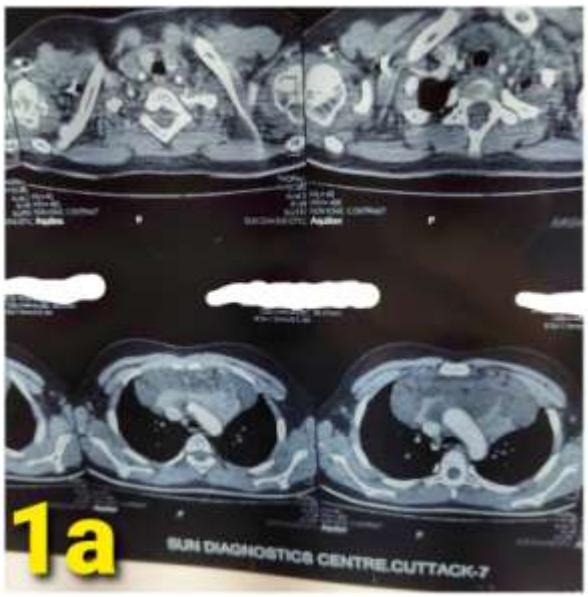

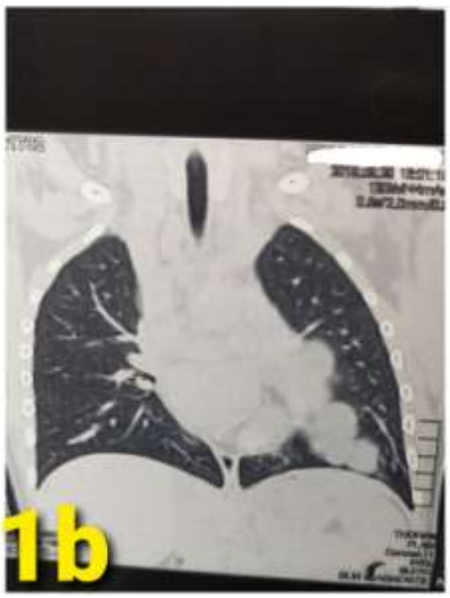

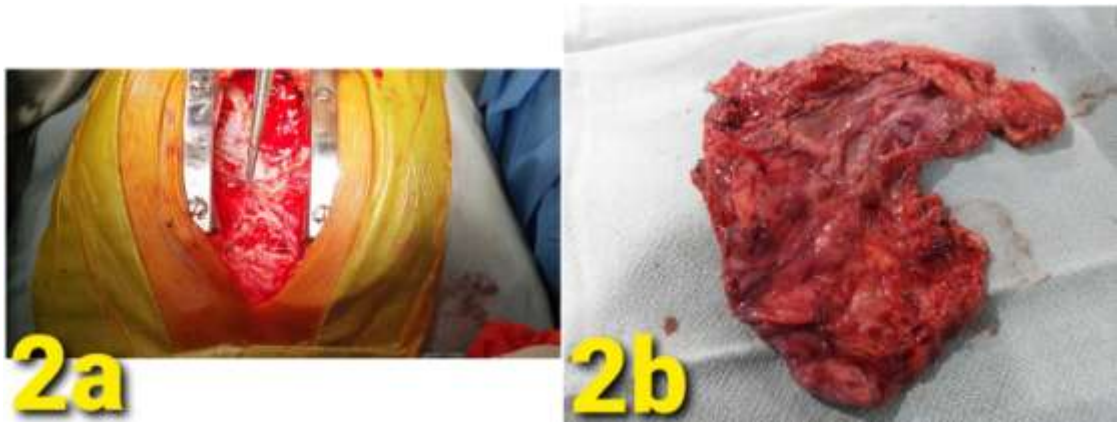

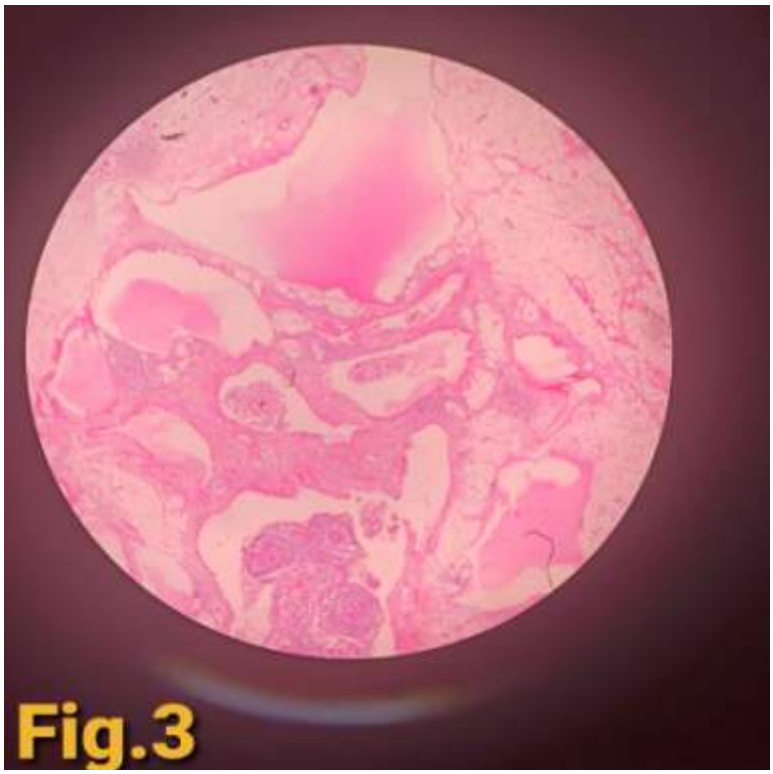

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