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Case Report

Cystic lymphangioma over the sternum: An extremely rare and atypical localization

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ABSTRACT

Cystic lymphangioma is a benign lymphatic system malformation that occurs usually in children. Most common site being cervicofacial region followed by axilla; the cases in the chest wall are very rare. We report a case of male infant with cystic lymphangioma occurring at an unusual site- over the sternum.

1. Introduction

Cystic lymphangioma is a congenital malformation that results from failure of primary lymphatic sac to establish drainage into the venous system leading to proliferation of lymphatic vessels and formation of cystic structure.[1,2] Rarely, it is an acquired malformation due to inflammation, injury or fibrosis.[2] It is the second most common benign vascular tumour of childhood, its incidence being 1/6000 live births.[3] This malformation is most often observed in the cervicofacial region (75%) and less frequently encountered in the axilla,[3–5] whereas mediastinal cystic lymphangioma is rare.[4,6] Other sites are mesentery, retroperitoneum, pelvis and lower limbs.[7,8] Its occurrence at sternal site is very rare and not many cases have been reported before.[7]

2. Case Report

A 10 months old male child presented with swelling over the sternum (Figure 1). The swelling was first noticed three months ago. It gradually increased to the present size. On clinical examination, the swelling was palpable, oval and of size 4x3 cm; over the sternum. It was fluctuant and non-pulsatile and nontender. Transillumination was not possible. Ultrasonography depicted a solitary subcutaneous cystic lesion, 4x3cm in dimension, without any intrathoracic extensions.

Surgical excision of the mass was done (Figure 2) and was subjected for histopathological examination.

Grossly, a globular specimen of size 4x3x2cm was received, with intact capsule. It was soft in consistency. On cut section, it was multicystic, filled with thin, whitish fluid (Figure 3). Microscopy revealed lymphatic spaces with irregular lumens lined by flattened epithelium and intervening stroma comprising of delicate meshwork of collagen intermixed with fibro adipose tissue (Figure 4). The characteristic histological features suggested the diagnosis of cystic lymphangioma. Considering its rarity and atypical localization, immunohistochemical study was done which revealed diffusely D2-40 and CD31 positive lining endothelium confirming the diagnosis (Figure 5).

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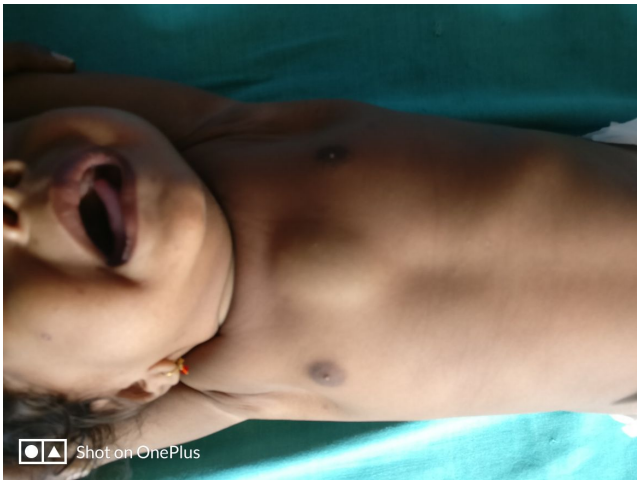

Fig. 1: 10 months old child with swelling over sternum

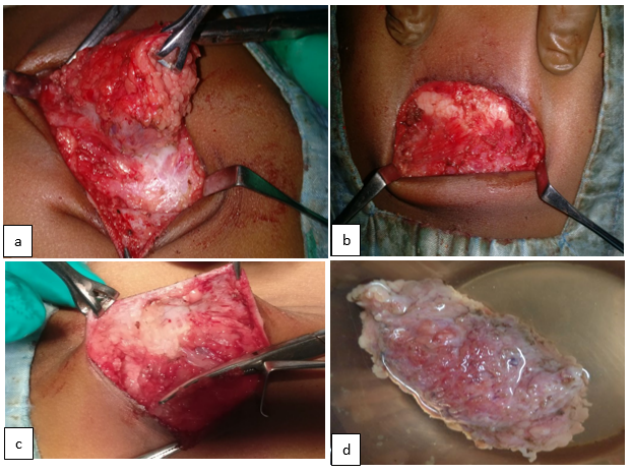

Fig. 2: a-d: Intraoperative view

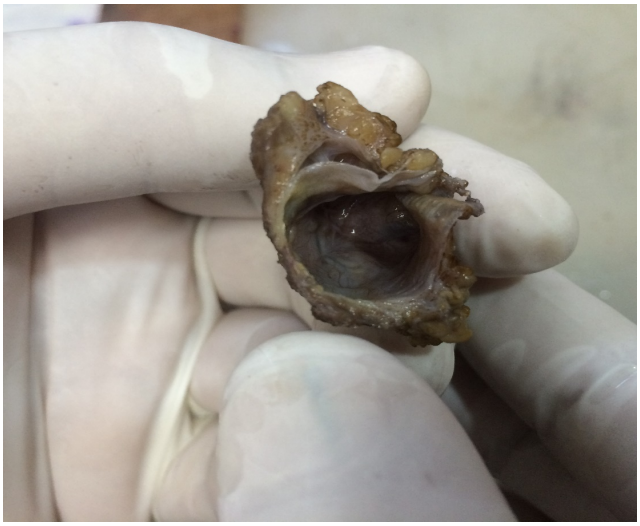

Fig. 3: Excised tissue on cut section. Note the multicystic nature of the lesion

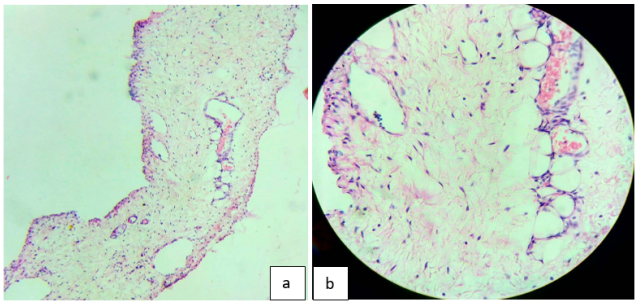

Fig. 4: Lymphatic spaces lined by flattened epithelium. Intervening fibroadipose tissue also seen; **a:** Low power view; **b:** High power view

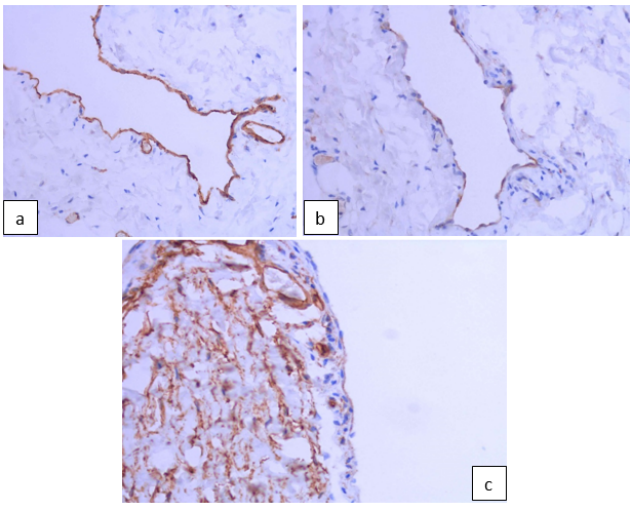

Fig. 5: a-b: Immunohistochemical examination with CD31 showing staining of the cells lining the inner surface of the cyst; (CD31, 10x).; **c:** Immunohistochemical examination with D2-40 showing staining in the endothelial cells; (D2-40, 40x)

3. Discussion

The lymphatic system plays an important role in human circulation and organ perfusion homeostasis. During the course of embryogenesis of lymphatic system, six primary lymphatic sacs are formed: two jugular, two iliac, one retroperitoneal and one dorsal to the retroperitoneal sac, the cisterna chyli. Later on numerous lymphatic channels from head, neck, limbs and body wall join the lymphatic sacs.[7,8] This lymphatic system starts to develop at around fifth gestational week and is expected to unite with the jugular vein in the neck at seventh gestational week. If this connection fails to develop by the 11^{th} to 12^{th} gestational week, cystic errors of lymphatic system are induced,[3] since the primary lymphatic spaces fail to join the central system.[1] The first detailed description of a lymphangioma is attributed to R Eden backer in1828.[9,10]

Smith et al[11] morphologically classified cystic hygroma into macrocystic, microcystic and mixed

type[3] whereas Mulliken et al.[12] proposed more widely accepted histopathological classification of lymphangiomas including three types : cystic type, simple (capillary) type and cavernous type.[3,8,13] Macroscopically, capillary lymphangiomas appear as wart-like or vesicular lesions that are unrelated to internal lymphangiomas. Cavernous lymphangiomas have discrete margins and insinuate themselves into surrounding structures. Cystic lymphangiomas, in contrast are multiloculated, fluctuant growths often enveloped in fibrous capsule.[9,14,15] The cyst may contain milky, serous, serosanguinous or straw coloured fluid. All histopathological types contain lymphatic channels lined by endothelial cells which are separated by a stroma composed of the connective tissue. Cystic lymphangioma consists of dilated lymphatic sacs with cyst walls lined by single layer of flattened epithelium, while cavernous and capillary lymphangiomas consist of smaller lymphatic channels.[3,8] In normal lymphatic vessels, endothelial cells express CD31 and usually do not express CD34 ; however, these cells do express the specific lymphatic marker D2-40. In contrast, in lymphangiomas, lymphatic vessels are more likely to express CD34.[4] Immunohistochemical studies in our case showed that the endothelial cells lining the inner surface of the cyst were specifically stained with D2-40 and CD 31.

Hancock et al. reported that 31.4% of all lymphangiomas are found in the cervical region, 18.9% in the scalp and the face (craniofacial), 9.2% in the trunk and 4.9% in the cervicoaxillothoracic region;[1,2] however can involve any body site where the lymphatic system originates.[3] The accurate anatomic location plays an important role in the management of lymphangioma, because the diagnosis is ultimately made postoperatively (after histopathological examination of resected tissue.[9] The differentials of cystic lesions of chest lying in midline or over sternum are dermoid cyst, lipoma, neurofibroma, ectopia cordis etc.[7] In addition to clinical findings the diagnosis of lymphangioma frequently requires aspiration of the lesion content for cytological, histopathological and radiological examinations. The management options are surgical excision, sclerosant therapy, radiotherapy, embolization, cyclotherapy, electrocautery, cryotherapy and laser therapy.[3,7,8] However, surgical excision is the recommended approach for lymphangioma treatment[3] and also the treatment of choice.[2]

4. Conclusion

Location of cystic lymphangioma over the sternum is very rare and should be considered as one of the differential diagnoses of midline cystic lesions of the chest or over the sternum.

5. Source of Funding

None.

6. Conflict of Interest

Authors declare no conflict of interest.

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