Cystic lymphangioma of the neck in an adult: A case report.

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Abstract:

Lymphangiomas are rare congenital malformations commonly seen in children. Its occurrence in adults is uncommon. The treatment of choice is complete surgical removal; however, the tumour tends to spread along vital structures therefore sometimes complete surgical removal is impossible. We describe the clinical and pathological features of cystic lymphangioma diagnosed in an adult with lateral neck mass. Clinical examination, ultrasonography, CT and fine-needle aspiration cytology were performed with complete resection. We report this case to emphasize the need to consider cystic lymphangioma in the differential diagnosis of lateral neck masses in adults, together with adequate imaging and cytological studies to corroborate the preoperative diagnosis.

Key words: Cystic lymphangioma, Neck mass, Surgery.

Introduction:

Lymphangioma is a benign congenital malformation of the lymphatic system that occurs as a result of sequestration or obstruction of developing lymphatic vessels. These lesions are usually discovered in infants or children younger than two years of age. Occurrence in adults is uncommon, and fewer than 100 cases of adult lymphangiomas have been reported in the literature.[1] The objective of this report is to discuss the clinical presentation, diagnosis, pathological findings and management of this malformation.

Case report:

A 32-year-old female presented in our department with diffuse swelling in the left supraclavicular region (Image 1) since four months. She did not have any obstructive symptoms like dysphagia, dyspnoea or any history of trauma or recent upper respiratory tract infection. Physical examination revealed a diffuse, soft, fluctuant, mobile swelling measuring 4 × 3cm in the left supraclavicular fossa, just above the medial one-third of the left clavicle extending into the posterior triangle of neck.

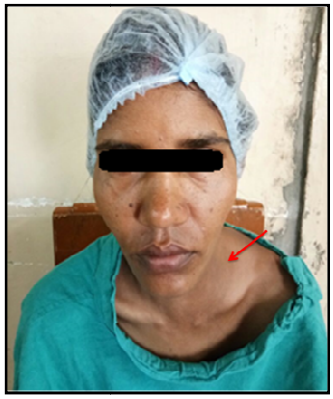

Image 1: Diffuse left supraclavicular swelling (red arrow)

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Suspecting the swelling to be a lipoma, the patient was referred to the cytopathology department where fine-needle aspiration cytology (FNAC) of the swelling was performed. Smears showed small lymphocytes, occasional macrophages along with RBCs in a proteinaceous background (Image 2 & 3). These features suggested a diagnosis of lymphangioma.

Image 2: FNAC Smear showing small lymphocytes & macrophages along with RBCs. [H & E, 4X]

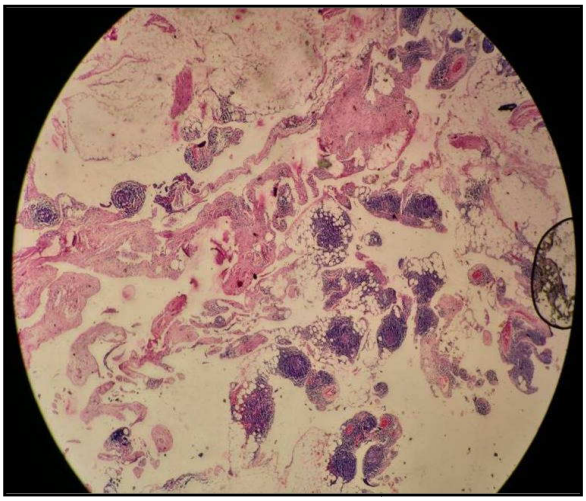
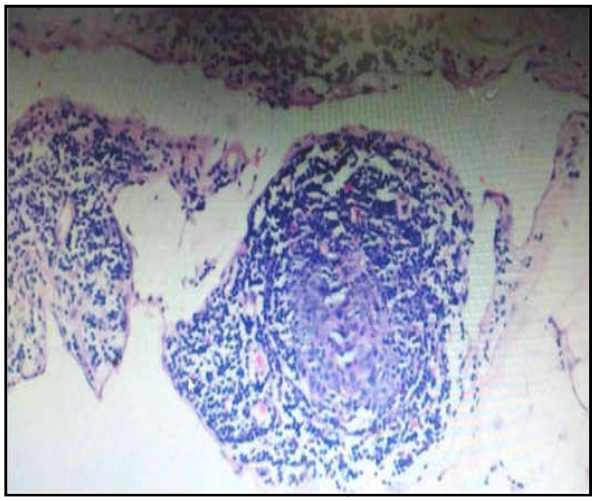


Image 3: FNAC Smear showing lymphocytes aggregates. [H & E, 10X]



CECT of neck was performed, which showed a well-defined, fluid density cystic lesion, measuring approximately 6 x 3.5 cm in left supraclavicular region located deep to strap muscles. Another similar characteristic cystic lesion of similar size was noted postero-laterally (Image 4). The lesions displaced left common carotid artery and internal jugular vein(without luminal narrowing) posteriorly, inferiorly, extending till manubrium sterni and superiorly till the thyroid cartilage (Image 5). The lesions caused mild displacement of trachea towards right side without any compression (Image 4). No post contrast enhancement was seen. These findings suggested benign cystic lesion of lymphatic origin (Image 4).

Image 4: Axial CECT scan through lower neck showing fluid density cystic lesion, displacing carotid vessels posteriorly.

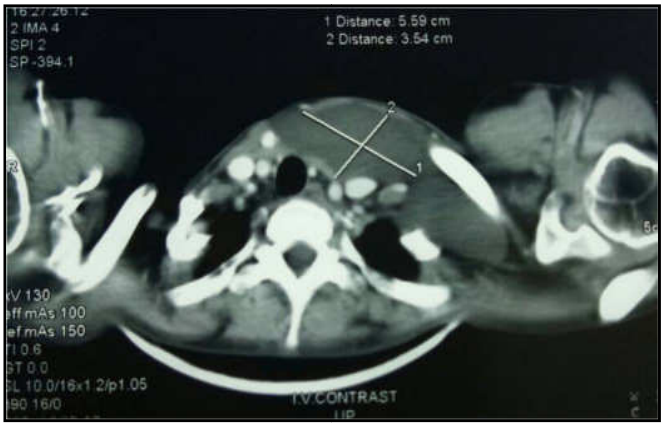
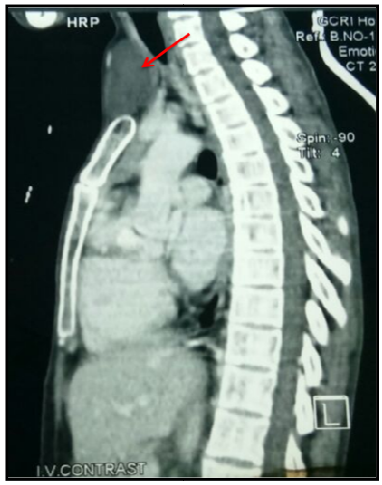


Image 5: Sagittal CECT showing superior and inferior extent of lesion. (Red arrow)



The lesion was reached through a 7 cm transverse skin incision, 2 cm above the clavicle on left side of the neck. The cystic lesions were found deep to middle layer of deep cervical fascia. The anteriorly located cyst got ruptured intraoperatively. Clear planes were found around the 2nd cyst and it was dissected from its medial end, behind the sternocleidomastoid muscle (Image 6)to its lateral end in the posterior triangle. It was touching the internal jugular vein and common carotid artery posteriorly. Total excision of cyst was done. The excised cyst was 4x3 cm size with thin glistening translucent cyst wall containing straw coloured fluid (Image 7).

Image 6: Intra-operative picture of lymphatic cyst deep to sternocleidomastoid muscle.

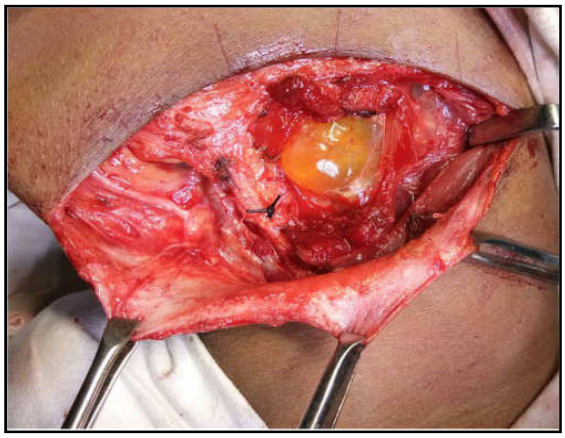
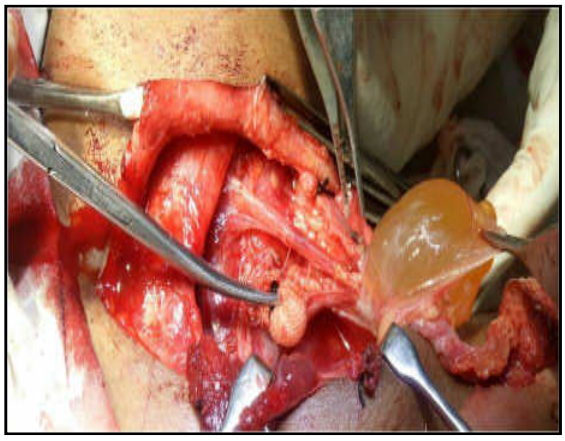


Image 7: Intra operative picture of excised lymphatic cyst showing thin translucent wall



There were no post-operative complications and the patient was discharged after one day of hospital stay. There was no evidence recurrence in subsequent follow up.

Discussion:

Lymphangiomas are a benign congenital malformation of the lymphatic system and consist of three histological subtypes. Capillary (composed of small lymphatics), cavernous (composed of larger lymphatics), and cystic lymphangioma (cystic hygroma- composed of large macroscopic lymphatic spaces with collagen and smooth muscle). Cavernous lymphangioma is the most common subtype. [2]

Cystic lymphangiomas occur in approximately 1 in 12000 births with 95% occurring by the second year of life. [1] It is uncommon in adults, as was the case with our patient. Although the lesion can occur anywhere, the most common sites are in the posterior triangle of the neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal space and pelvis[3]. Adult patients are usually asymptomatic.[1]

On physical examination these present as soft, painless, mobile, transilluminable cystic lesions. Lymphangiomas are best visualized by magnetic resonance imaging (MRI); the high water content allows lymphangiomas to appear hyperintense on T2-weighted images and the superior soft tissue contrast provides excellent anatomic delineation and extensions of the lesion.[4]. The other imaging methods are ultrasonography and computed tomography (CT).

This disease may be associated with Turner syndrome, Noonan syndrome, cardiac anomalies, trisomy syndromes and fetal hydrops.

Differential diagnoses vary with location and are usually hemangiomas, mucoceles and meningo-myeloceles.[5] Infection within the cysts (usually caused by streptococcus or staphylococcus species) may occur. This complication can cause rapid enlargement which may result in airway obstruction. Bleeding into the cyst is another complication. These lesions do not resolve spontaneously.

Percutaneous aspiration is not preferred because of the risk of bleeding, infection and recurrence. Injection of sclerosing agents like alcohol, bleomycin and OK-432 (a lyophilized mixture of streptococcus pyogenes and penicillin G potassium), have shown favourable results.[6]. Complete surgical excision is the preferred treatment. It can be performed under general or local anaesthesia. Sometimes, this may be impossible due to the infiltrating nature of the lymphangioma within and around neurovascular structures, muscles, blood vessels. In this condition, de-roofing, partial cystectomy and drainage of the contents can be performed. With this kind of treatment, recurrence rate of 10-15 % is reported. [7]

Conclusion:

The awareness of occurrence of cystic lymphangioma in adults is important for its proper management which includes complete surgical removal, to prevent recurrence. This report suggests the need to consider cystic lymphangioma in the differential diagnosis of lateral neck masses in adults, together with adequate imaging and cytological studies to corroborate the pre-operative diagnosis.

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