RECURRENT CERVICAL LYMPHANGIOMA IN AN ADULT- A CASE REPORT

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PRESENTATION OF THE CASE

A 32 years old male presented in our hospital with swelling in right side of lateral aspect of lower neck for 6 months which was initially small in size, gradually increased over 6 months to present size of 6 x 5 cms. He complained of slight discomfort during neck movement. He had no pain over swelling, shortness of breath, dysphagia, dysphonia and had no other swelling in body. He was operated for similar swelling in right side of neck at same location 1 year back. He remained asymptomatic for 6 months; then he noticed a recurrence of present swelling in same location.

Swelling was soft, cystic, globular, non-fluctuant, situated in the posterior triangle of right side of neck above clavicle behind sternocleidomastoid with smooth surface; measured 6 x 5 cms with transillumination test being positive. Transverse scar was present. Swelling was non-pulsatile, no dilated neck veins were visible, no other swelling was seen in the neck, trachea is centrally placed.

CLINICAL DIAGNOSIS

Recurrent Cystic Lesion of The Neck.

DIFFERENTIAL DIAGNOSIS

- Neoplasms- metastasis from squamous cell carcinoma (HNSCC), lymphoma, papillary thyroid cancer or salivary gland cancer.
- Thyroglossal Duct Cyst
- Cervical Ranula
- Bronchial Cleft Cyst
- Laryngocele
- Parathyroid Cyst
- Hydatid Cyst
- Haemangioma

PATHOLOGICAL DISCUSSION

Blood investigations were normal. USG neck showed evidence of large loculated cystic lesion with internal septation measuring approx. 44 x 12 x 42 mm. CECT scan of neck revealed multiloculated cystic lesion in right side of neck measuring 32 x 20 x 50 mm in carotid space extending inferiorly lateral to jugular vein and carotid vessels, deep to sternocleidomastoid muscle (Figure 1a).

Gross- Received already cut opened cystic mass measuring 6 x 4 cms. On cut section seromucinous material was noted (Figure 2).

Microscopy- H&E stained multiple sections showed fibro connective tissue and tumour. Tumour is composed of numerous lymphatic channels lined by flattened endothelium filled with lymph and lymphoid cells (Figure 3). Also, few congested blood vessels and fibro fatty tissue was noted- lymphangiomatous malformation.

Lymphangiomas are rare congenital benign lesions that result from abnormal growth of lymphatic vessels. Lymphangiomas occur most commonly in children and rarely observed in adults. It is rare for lymphangiomas to make their initial presentation during adulthood and fewer than 100 such cases have been reported. The pathophysiology of adult lymphangiomas is not clearly understood. Herein we report a case of recurrent adult lymphangioma in a 32-year-old male localized in the neck.

Lymphangiomas are rare congenital benign lesions & results from abnormal growth of lymphatic vessels. Three theories have been proposed for its origin. (a) Blockage or arrest of normal growth of the primitive lymph channels occurring during embryogenesis (b) Primitive lymphatic sac does not reach the venous system (c) During embryogenesis, lymphatic tissue lays in the wrong area.[1]

Cervical lymphangioma are derived from lymphatic vessels with a progressive and painless growth. The incidence of lymphangiomas has been reported to range from 1.2 to 2.8 per 1000 newborns. They have no gender predilection. Lymphangiomas occur most commonly in children and rarely observed in adults. It is rare for lymphangioma to make their initial presentation during adulthood and fewer than 100 such cases have been reported. Most lymphangiomas are congenital, but later presentations may occur from infections, neoplasm, trauma and iatrogenic injuries in an adult.[2]

First case of lymphatic malformation was described by Redenbacher in 1828.[3] Imaging techniques aid in the precise mapping of the lesion and in defining its boundaries with the surrounding vital structures. So, it improves therapeutic

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success. Complete surgical excision considered to be best approach in adults.[4]

Lymphangiomas are rare benign lymphatic malformations regarded as an abnormality of morphogenesis rather than as a neoplasm.[5] They occur most commonly in children and rarely in adults. Approximately 60% of lymphangiomas are present at birth, and up to 90% are detectable by 2 years of age.[6] It is rare for lymphangiomas to make their initial presentation during adulthood and such presentations may occur as a result of trauma, infection, neoplasms or iatrogenic injuries.[7] The pathophysiology of adult lymphangiomas is not clearly understood, but may occur secondary to induction of dormant rests of embryonic lymphatic tissue that are stimulated to differentiate and grow.[8]

The most common sites are in the posterior triangle of the neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal space and pelvis.[9] Most lymphangiomas are asymptomatic and have no gender predilection. They present as a painless mass that progressively enlarges. Typically, the mass is soft, non-tender, and ill defined. Symptoms may develop when the lymphatic malformation enlarges to compress surrounding tissue. There may be obstructive symptoms like dysphagia, dysphonia.[10] USG and CT have been used to evaluate the anatomy of lymphangiomas.[11] FNAC findings include small and round lymphocytes with intermingling histiocytes without mitoses or atypia.[12] Grossly - well-circumscribed lesions made up of one or more interconnecting cysts to ill-defined, sponge like compressible lesions composed of microscopic cysts.

Microscopically these lesions appear as endothelial lined lymphatic spaces with intervening fibrous tissue and lymphoid aggregates.[13]

Differential diagnosis in adults include malignancy, hydatid cyst, bronchial cleft cyst, haemangioma, thymic cyst and plunging ranulas/mucocoele.[14,15] Cytologically absence of parasitic elements ruled out hydatid cyst, absence of blood ruled out haemangiomas, absence of squamous cells and cholesterol clefts ruled out bronchial cysts, absence of atypical cells ruled malignancy. Plunging ranulas show foamy histiocytes in large numbers and few lymphocytes and background shows mucinous material.[15] Histopathologically the tumour showed multicystic spaces lined by flattened endothelial cells. The plunging ranulas lack lining epithelial cells and are lined by rows of histiocytes.[16]

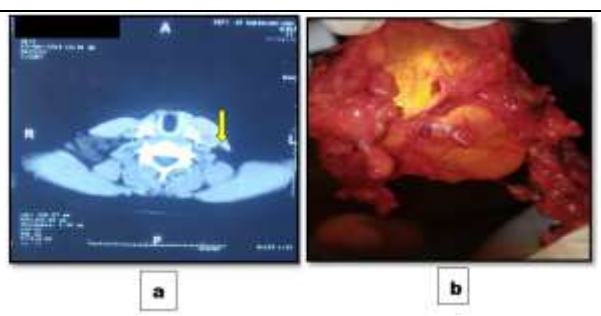

Figure 1. 1a) CECT Image Showing Cystic Masses in The Right Cervical Area. 1b) Transillumination Test- Positive

Figure 2. Gross Picture Showing Multiloculated Cystic Mass

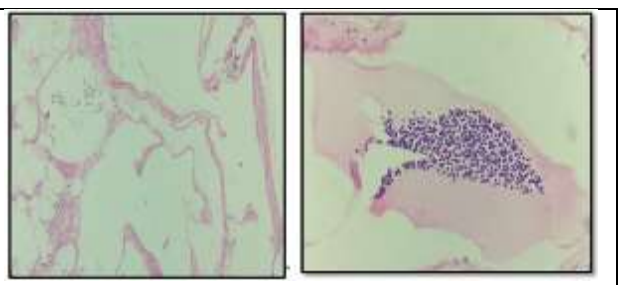

Figure 3. HPE Showing Lymphatic Channels Lined by Flattened Endothelium Filled with Lymph and Lymphoid Cells (H&E 10x, H&E 40x)

DISCUSSION OF MANAGEMENT

The swelling was approached with 6 cms transverse incision extending from midline to anterior border of trapezius 3 cms above and parallel to clavicle; sternocleidomastoid was retracted medially.

Clear fluid filled cystic swelling measuring approx. 6x4 cms deep to the midpoint of sternocleidomastoid identified (Figure 1b). This cystic swelling was circumferentially dissected and removed in its entirety. Care has been taken during dissection to prevent injury to vital structures. The excised lesion was submitted to histopathological examination.

For those with minimal symptoms, treatment can be delayed beyond infancy and include a combination of surgery and sclerotherapy with OK-432, a lyophilized mixture of group-A Streptococcus pyogenes.[17] The main stay of therapy is surgery. Complete excision of lymphangioma has been shown to have an 81% cure rate. When only part of the lymphatic malformation is excised, there is an 88% recurrence rate.

Although a rare possibility in adults, cystic lymphangiomas must be considered in the differential diagnosis of cystic lesions in neck of adult. Imaging techniques can aid in mapping of the lesion and in defining its boundaries with the surrounding vital structure. Complete surgical excision remains the treatment of choice for large, persistent or recurrent lymphangiomas in adult. This will

help in lowering the rates of recurrence. Histopathological diagnosis is gold standard for confirmation of diagnosis.

FINAL DIAGNOSIS

Recurrent Cervical Lymphangioma.

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