CYSTIC LYMPHANGIOMA IN ADULT - A RARE CASE PRESENTATION

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Abstract: Cystic lymphangiomas are rare benign neoplasms that occur due to lymphatic malformations. Congenital variety occurs at birth and presents within two years of life, whereas acquired types in adults occur due to spontaneous cause, trauma, inflammation and lymphatic obstruction. They arise in neck, axilla, mediastinum, retroperitoneal space, etc. Prompt diagnosis and complete excision of the cyst is curative. A female of thirty one years, presented with left sided neck swelling of six months duration, occupying lower neck and just above the medial one third of left clavicle that was compressable and transilluminating. The case was evaluated and complete excision of the cyst was done. About less than 100 cases of Cystic Lymphangiomas in adults had been reported in literature. We present a case of adult type of Cystic Lymphangioma in neck of thirty one years female, producing mass effect over left jugular vein. Complete excision was done to prevent its complications and recurrence. We report this case, because of its rare occurrence in adults and the difficulties in its diagnosis.

Keyword: Lymphangioma, cystic, unilateral infrahyoid, complete excision.

INTRODUCTION
Cystic lymphangiomas are rare benign congenital or acquired malformations arising in the lymphatics. The congenital type that occurs at birth, and more than 90% of those cases presents within first two years of life. They occur either as an isolated case or with an associated chromosomal abnormalities. Acquired type in adults are rare, and occur due to various proposed etiologies such as spontaneous cause, trauma, inflammation, and due to lymphatic obstruction. They present as cystic swellings of lymphatics, involving head and neck, axilla, groin, retro-peritoneal space, small bowel, mesentry, gall bladder, etc. Till date, less than 100 cases of cystic lymphangioma in adults have been reported in the literature and they pose difficulty in the diagnosis. In this report, stage I unilateral infrahyoid and macrocystic type of cystic lymphangioma involving the neck of a thirty one year old female is presented.

CASE PRESENTATION:
A female of age 31 years presented with complaints of swelling in the left side of the neck of 6 months duration with discomfort and dull aching pain over left side of neck while swallowing of 3 months duration. There was no recent history of trauma, neck infections or surgery in the neck. No significant past illness or menstrual disturbances.

**Cystic Swelling Between the Two Heads of Sternocleidomastoid**

General condition of the patient and vitals were stable. On examination of the neck, an 8 x 6 cm single, discrete swelling occupying the left supraclavicular fossa, behind the both heads of sternocleidomastoid muscle and just above the medial one third of the left clavicle, that becomes prominent on valsalva manoeuvre. There were no dilated or prominent veins in the neck. The swelling is partially compressible, non pulsatile, soft and cystic in consistency and transillumination positive. No other palpable neck mass or regional lymphadenopathy. On FNAC, about 24 ml of haemorrhagic fluid was aspirated, showed mature lymphocytes in a haemorrhagic background without pleomorphic cells. The swelling disappeared after aspiration and reappeared within days. USG Neck showed, a cystic lesion of 8 x 4 x 2 cm occupying the left lower neck supported by MRI of Neck displaying a cystic lesion of 7.1 (TR) x 2.9 (AP) x 5.4 (CC) cm situated in the left lower neck, anterior to the carotid vessels and closely related to sternocleidomastoid and clavicular margin and consistent with features of Cystic Lymphangioma. Barium swallow and UGI Endoscopy were found to be normal.

**MRI Neck - Plain Study**

**MRI Neck - Contrast Study**

After obtaining Informed consent and Anaesthetic fitness she was posted for surgery. Under general anaesthesia, through a left skin collar incision over the neck, the thin walled cystic lesion of size 7 x 4 x 2 cm, containing clear fluid was excised completely. Wound was closed in layers. Post-operative period was uneventful.
HPE confirmed the Cystic Lymphangioma.

DISCUSSION In 1909, Sabin described the derivation of lymphatic tissue from the five primitive buds of developing venous system, namely a paired jugular sacs, a paired posterior sacs and a single retro-peritoneal sac. Sequestration of the lymphatic tissue that has lost its potential to grow into any of these sacs results into lymphangioma. In 1976, Whimster using lymphangiographic and radiographic studies, revealed that the large multi lobulated cisterns arising from the primitive lymph sac fails to connect to the rest of the lymphatic system, and the cause for this failure is unknown. Few lymphangiomas represent vascular malformations during embryonic development, rather than as true neoplasms.

Vascular Endothelial Growth Factor (VEGF-C) and VEGF-receptor -3 are active in the formation of lymphangioimas. Based on MICROSCOPIC characteristics they are classified as Capillary (lymphangioma circumscriptum) with small capillary sized epidermal lymphatics, Cavernous with dilated lymphatic channels invading surrounding tissues, Cystic lymphangioma (cystic hygroma) that are large cysts filled with straw coloured protein rich fluid, and Hemangio lymphangioma with vascular component.

According to their SIZE AND VOLUME, they are Microcystic of volume less than 2 cm³, Macrocystic of volume more than 2 cm³ and Mixed with both microcysts and macrocysts. By their location and extent of neck cysts they are STAGED into, Stage I – Unilateral Infrahyoidal Stage II - Unilateral Suprahypoidal Stage III – Unilateral, supra and infrathyroidal Stage IV – Bilateral Suprahypoidal Stage V - Bilateral Supra and Infrahyoidal Cystic lymphangioniomas arise as single or multiple cysts involving the lymphatics of skin, subcutaneous and loose areolar tissue. They are mostly asymptomatic and have no gender predilection. They are soft, slow growing, doughy mass, and multi loculated cysts with clear or yellow lymph fluid. They are fluctuant, transilluminating and fixed to deeper tissues.

They may cause cosmetic disfigurement, minor bleeding, recurrent cellulitis, and lymph fluid leakage. They rarely lead to intrathoracic extension, create mass effects on vital structures, dysphagia, respiratory distress, and severe neck infections. As their is no malignant transformation, it has excellent prognosis. Incomplete excision results in recurrence. USG Neck helps to know the shape, size, extension and its cystic nature. CT Neck determines the extent of the cyst, cystic or lymphatic nature of the mass. They produce few characteristic features, such as well circumscribed lesion.
adjacent structures that have been enveloped or displaced, absence of calcifications, and varied attenuation within the lesion. MRI of Neck complimates the findings obtained by either USG or CT in delineating the extent of involvement and relationship to adjoining vital structures, especially in the neck. OROPHARYNGEAL ENDOSCOPY to rule out oropharyngeal extension. IMMUNOHISTIOCHEMISTRY with Factor VIII related antigen it is negative or weakly positive (positive in hemangioma), and with Laminin it shows discontinuous basal lamina of the vessel (multilayered basal lamina in hemangiomas). DERMOSCOPY in cases of capillary (cutaneous) lymphangiomas.

HISTOLOGICALLY Cystic lymphangioma is indistinguishable from cavernous lymphangioma. In DIFFERENTIAL DIAGNOSIS, due to its cystic nature; Thymic cyst, bronchogenic cyst, cystic teratoma or lymphangiectasias should be considered. ADJUVANT THERAPIES such as Sclerosant therapy using OK-432 (picibanil), bleomycin, fibrin tissue sealant, sodium tetradecyl sulphate, doxycycline, ethanol, 10% Hypertonic Saline, etc are tried that induce fibrosis and producing ablation of the endothelial cells of the disrupted lymphatics which feed the lymphocele. Intravenous cyclophosphamide for recurrence had been tried in paediatric cystic hygromas. Aspiration causes re-accumulation of the cyst. Laser treatment for oropharyngeal extension, Cryotherapy, and Radiotherapy are also tried. Complete SURGICAL EXCISION gives excellent prognosis without recurrence. Anatomically inassessable lesions needs staged removal. SUMMARY Till date, less than a hundred cases of Cystic Lymphangiomas in adults had been reported in the literature. The various site of occurrence in the reported cases were lymphatics of head & neck, mediastinum, axilla, groin, retroperitoneum, small intestine, mesentry, gall bladder, and extremities. They are usually asymptomatic, except cosmesis. As the normal lymphatics are always in relation to vascular and neural elements, on progression these pathological cystic lesions produce mass effects on these vital structures. This is evident in this case producing mass effect on the left jugular vein. Adjuvant therapies such as aspiration, sclerosant injections, Cryotherapy, radiotherapy, and laser therapy doesn’t produce uniform and expected results. Complete excision of the cyst is curative. CONCLUSION In adults, Cystic Lymphangiomas presents rarely. Clinical evidence supported by imaging studies and biopsy are diagnostic. Aspiration causes recurrence and the various adjuvant therapies produced invariable results in the previous studies. Complete surgical excision prevents recurrence and its complications. This case is reported because of its rarity in adults and difficulties in its diagnosis and management.

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