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ADULT CYSTIC LYMPHANGIOMA AND ITS FEATURES: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT **Background:** Lymphangiomas are benign malformations of lymphatic system uncommon in adults. We present a rare case of adult cystic lymphangioma in the neck region. **Case presentation:** A 26 year old female presented with a large swelling on the right side of the neck. The patient was uncomfortable due to the cosmetic impact of the swelling. Clinical diagnosis of cystic lymphangioma was made based on history and physical examination. An impression of lymphangioma was given on FNAC and radiological examination. The cyst is surgically removed by raising a skin flap and the cyst wall specimen was sent for histopathological examination for confirmation. The histopathological sections confirmed the diagnosis. The postoperative period is uneventful. There was no recurrence till the current follow up. **Conclusions:** In a case of painless and progressive neck swelling, lymphangioma should be considered as a differential diagnosis despite the fact that they occur rarely in adults and appropriate treatment should be initiated to prevent pressure symptoms due to the progressive nature.

KEYWORDS : Cystic Lymphangioma, Neck swelling, Cystic hygroma, FNAC, Case report, Surgical excision

INTRODUCTION:

Cystic lymphangioma is a benign malformation of lymphatic system, which is common in the lymphatic rich head and neck region, axilla and groin during the first 2 years of life [1-4]. They can occur in various sites such as mediastinum, retroperitoneum, orbit, tongue, lip, base of oral cavity, breast, cervical region, abdomen, inguinal region, spleen, colon and at the level of spermatic cord [1-7]. There are various classifications for lymphangioma. It may be congenital or acquired [3]. Congenital lymphangiomas occurs due to improper connection of lymphatic channels to the main drainage ducts. Acquired lymphangiomas occur when there is any disruption of previously normal lymphatic channels due to surgery, trauma, malignancy and radiation therapy. Histopathologically lymphangiomas can be classified into cavernous, capillary and cystic. Cavernous lymphangiomas are well defined and consists of large dilated lymphatic channels. Capillary lymphangiomas composed of small thin walled capillary sized lymphatics. Cystic lymphangiomas or cystic hygromas are composed of large lymphatic spaces with lymphocytes, collagens and smooth muscles. Two theories of pathogenesis available are mechanical theory and congenital theory [2]. The mechanical theory defines lymphatic obstruction or contusion, while the congenital theory explains the improper connection of lymphatic channels [2]. The prevailing theory of adult cystic lymphangioma is due to delayed proliferation of the lymphatic channels [10]. The differential diagnosis for cystic lymphangiomas are lipoma, cystic choriostoma, thymic cyst, pericardial cyst, bronchogenic cyst, tumors of major salivary gland, carotid body tumors, soft tissue sarcomas, thyroid tumors, mucocele and cystic teratoma [8-11]. Clinical diagnosis is based on previous medical history and physical examination.

Other diagnostic approaches are FNAC and radiological examination [8, 11]. Cystic lymphangioma can be differentiated from others by histopathological examination [8, 11]. Before surgical excision, the extent was confirmed by radiological examination as intrathoracic extension can occur in some cases. Treatment of choice is complete surgical excision of the affected lymphatic channels [12] with good cosmesis and preservation of vital anatomical structures. Other modalities of treatment are sclerotherapy, laser surgery, cryotherapy, electrocautery, intralesional steroids, embolization, aspiration and drainage by mediastinoscopy, chemical sclerosity with IV cyclophosphamide, video thoracosopy, scannopuncture, guided echo and radiotherapy [2, 3]. The effective treatment for Multiloculated masses and complete resolution is found to be better with complete surgical excision. The complications of surgical excision are haemorrhage, haematoma, seroma, injury to adjacent structures during the procedure, wound infection, flap infection & necrosis [3].

Herein, we report a case of large cystic lymphangioma in the neck region in a 26 year old female. Diagnosis was made by clinical examination along with simple investigation by FNAC. We also discuss the features of radiological and pathological features of Cystic Lymphangioma.

Case Presentation:

A 26 year old female presented with a large swelling on the right side of the neck region as shown in Figure 1 (Fig.1a.Gross picture shows swelling in the neck region.) for the past 3 months, which is gradually increasing in size without associated systemic symptoms, dysphonia or pain. No personal or family history in relation to the current disease and no history of trauma, radiation exposure, infection of head and neck region were described. On physical examination, the swelling was non-tender, soft and cystic with a smooth margin and bosselated surface. Fine needle aspiration cytology from the large cystic swelling on the right side of the neck was done and yielded 5 mL of straw colored fluid. Cytological smear shows plenty of scattered lymphocytes. Background shows proteinaceous material as shown in Figure 1(Fig.1b.FNAC smear showing numerous mature lymphocytes in a protein background). Ultrasound shows large septate cystic lesion (Figure 1c.Ultrasound shows large septate cystic lesion.). MRI of the neck region showed evidence of well-defined cystic lesion with internal separation of size 11.9×11.4×11.3 cm (Volume=1.53 L) along the right posterior triangle of the neck between lower two-third of sternocleidomastoid muscle and trapezius muscle in intermuscular plane insinuating into subcutaneous plane extending from C3 to D3 level [Fig.2 MRI shows well defined T1 hypointense (a), T2 hyperintense (b) multisepated cystic lesion.]. The lesion is seen displacing trachea and midline structures left laterally. The patient's complete blood count and other biochemical parameters were within normal limits. Radiological impression of Cystic lesion probably cystic lymphangioma was consistent with the FNAC findings. A complete surgical excision of the cyst was done and the specimen was sent for histopathological examination [Fig.3 a.Intraoperative shows raised skin flap and other structures preserved;b.Excised cyst wall section sent for histopathological examination;c.Postoperative image showing wound healing with healthy skin flap;d.Histopathological examination shows Large, irregular vascular space lined by flattened, bland epithelial cells(i.e.,lymphatic channel) with collagenous stroma containing lymphocytes]. The sections sent confirmed the diagnosis of cystic lymphangioma. The skin and the anatomical structures are

preserved and the postoperative period is uneventful with no complications and there is no signs of recurrence till the last follow up.

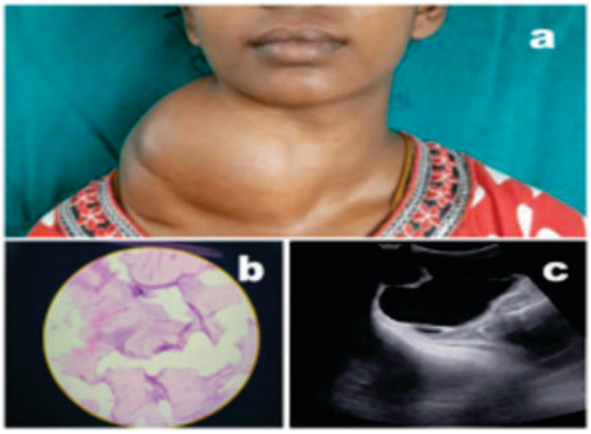

Figure 1 a.Gross picture shows swelling in the neck region; b.FNAC smear showing numerous mature lymphocytes in a protein background;c.Ultrasound shows large septate cystic lesion.

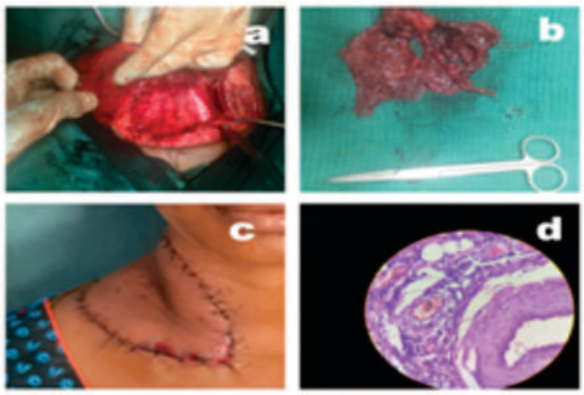

Figure 2 a.Intraoperative shows raised skin flap and other structures preserved;b.Excised cyst wall section sent for histopathological examination;c.Postoperative image showing wound healing with skin preservation;d.Histopathological examination shows Large, irregular vascular space lined by flattened,bland epithelial cells(i.e.,lymphatic channel) with collagenous stroma containing lymphocytes.

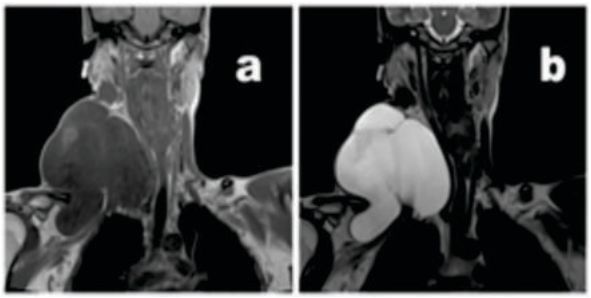

Figure 3 MRI shows well defined T1 hypointense (a), T2 hyperintense (b) multisepated cystic lesion.

CONCLUSIONS:

The patient underwent a complete surgical excision of the lesion as soon as the MRI results were received with preservation of skin and other structures. As removing the cyst being a concern for a surgeon, excision along with preservation of skin and other anatomical structures should be the ideal one for the patient and so a skin flap is raised to ensure proper cosmetic result. We emphasis that the patient's concern for cosmetic effect of the swelling and occurrence of complications and insinuation into adjacent structures, early complete surgical excision should be made as early as the diagnosis is confirmed along with MRI findings to know the extent of the cyst. This will prevent the complications of the growth of the cyst. Despite the limitations of a single case presentation, this will present a complete picture of a large, multiloculated cystic lymphangioma in adults and an insight into the surgical excision.

Patient Perspective:

The patient was concerned about the cosmetic effect of the swelling. Patient also feared about the postoperative complications. After explaining the procedure and complications, the patient gave consent for surgery. The skin and other anatomical structures around the swelling were preserved. The postoperative period was uneventful. After the surgical excision, the patient was feeling well.

Competing interests: The authors declare that they have no competing interests.

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