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CASE STUDY

CERVICAL CYSTIC LYMPHANGIOMA IN AN ADULT: A CASE REPORT OF RARE ENTITY WITH REVIEW OF LITERATURE

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ABSTRACT

Lymphangiomas are rare benign hamartomas that result from maldevelopment of primitive lymphatic sacs. They are most frequently found in the neck and axilla, while intra-abdominal and mediastinal lymphangioma are uncommon. Lymphangiomas are rare pediatric tumors and very fewer cases reported in adults according to the literature. Therefore this article discusses a case report of cystic lymphangioma of 22 year old male patient with a brief review on its clinicopathologic presentation, classification, diagnostic techniques and its management.

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INTRODUCTION

Lymphangiomas are developmental defects of the lymphatic channels that belong to a large spectrum of vascular malformations (Emmanuel A. Ameh *et al*., 2011). They constitute 6% of all benign tumors of childhood and most of the cases (90%) diagnosed exclusively before the age of two years (Kraus *et al*., 2008). Lymphangiomas occur exceedingly rarely in adults and very few cases are described in the literature (Morley *et al*., 1999). Their diagnosis is generally based on previous medical history, clinical presentation followed by ultrasonography and histopathology. Despite their benign nature, surgical management is difficult, especially for cavernous/cystic lymphangiomas because of their tendency to spread along the vital structures and the subsequent high incidence of recurrence within 6 months to 1 year postoperatively (Kalsotra *et al*., 2001). Hence this article discusses a rare case of cervical cystic lymphangioma of 22 year old male patient with a brief review on its clinicopathologic presentation, classification, diagnostic techniques and its management.

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Case report

A 22 year old male patient presented with a chief complaint of swelling in left side of the face in front of the ear since 5 years. The swelling was slow growing but showed a rapid growth since last 2 months. On extra oral examination the lesion was extending superiorly 0.5 cm in front the pinna of the ear, inferiorly 4 cm below the inferior border of mandible and 2 cm anterior to the angle of the mandible and posteriorly covering the posterior border of ramus of mandible (Figure 1). The swelling was present on the left side submandibular region which was ovoid with well defined borders, soft, fluctuant in nature and was not compressible. The cervical ultrasonographic findings revealed large cystic mass in the soft tissue of the left cheek region filled with turbid, haemorrhagic fluid, measuring 4.5X2.5 cm in diameter with septae in between. No extension or involvement seen in parotid gland. Small enlarged nodes are also seen in upper part of neck. Magnetic resonance imaging (MRI) findings showed a well defined T2 hyperintense lesion with lobulated outline, noted in the submandibular region and extending along the medial as well as lateral aspect of the posterior portion of ramus of mandible. Fine needle aspiration cytology (FNAC) revealed macrophages on a haemorrhagic background, no malignant cells were seen. Depending on clinical and radiological assessment, we came to a provisional diagnosis of

Lymphangioma. Considering a gold standard treatment plan, surgical excision was performed for the lesion. Nasal endotracheal intubation was done under general anesthesia (GA) and strict aseptic conditions. A submandibular incision was placed and tissue dissection was carried out in subplatysmal plane to reach the lesion. The invaded lesion was separated from adjacent structures like muscle, tissues, nerves and excised in toto (Figure 2A/B). A watertight closure was achieved using three layered closure and a pressure dressing was applied with drain in place. The received specimen was soft large cystic sac (5 cm X 4cm in diameter) with external pebbly surface. The cut surface showed multiple blood filled macro cysts with intersecting thick septae (Figure 3).

On histopathological examination, haematoxylin and eosin (H and E) stained section showed multiple large irregular cystic luminal spaces which were lined by widely spaced thin endothelial cells. The stroma supporting the cystic spaces was dense fibro cellular with proliferation of small variable sized vascular channels filled with lymphocytes and occasional red blood cells (RBC). Many lymphocytic aggregates were also noted along with muscle bundles and adipose tissue at the periphery (Figure 4). Correlating the clinical, radiological and histopathological investigation, a final diagnosis of Cystic lymphangioma was given. The patient was advised postoperative short term and long term follow up visits. In postoperative follow-up, he was asymptomatic during the following 12 months period and MRI was unremarkable.

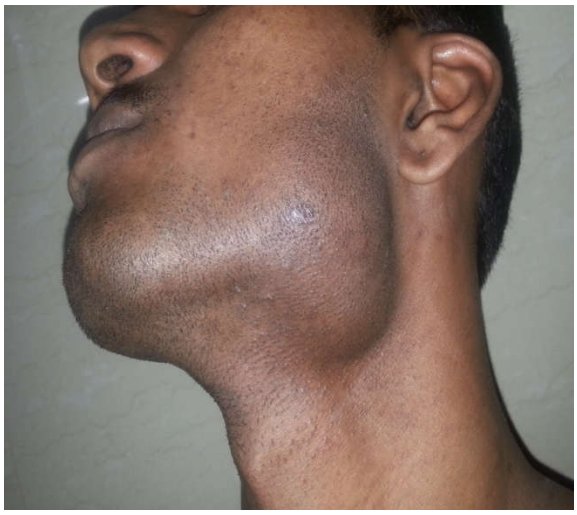


Figure 1. Clinical picture showing extra oral nodular swelling in front of pinna of the ear

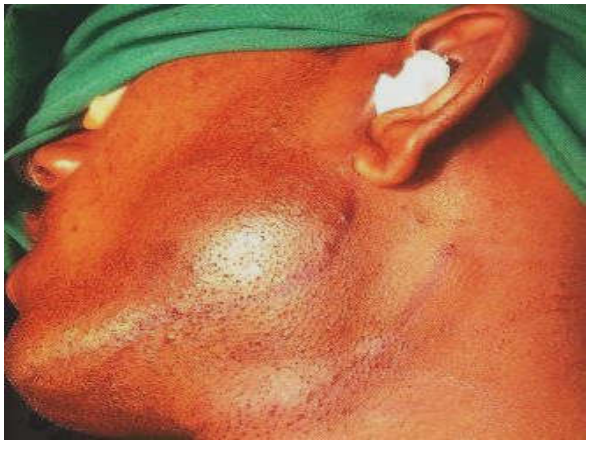
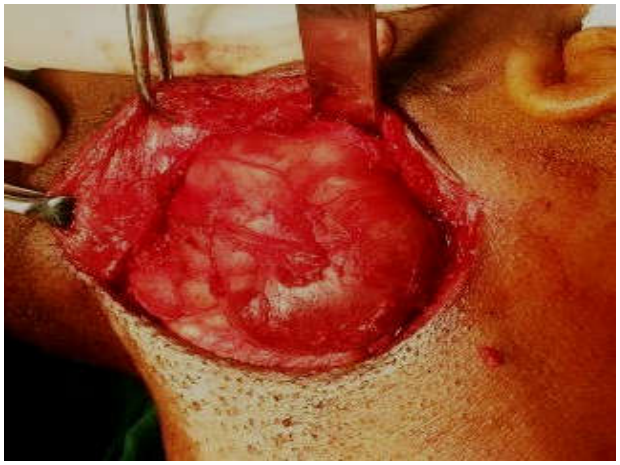


Figure 2. A: A submandibular incision was marked in subplatysmal plane to reach the lesion



B: The lesional tissue on surgical exposure

Figure 3. The excised specimen in toto. [Inset cut surface showing multiple macro cysts with intersecting thick septae]

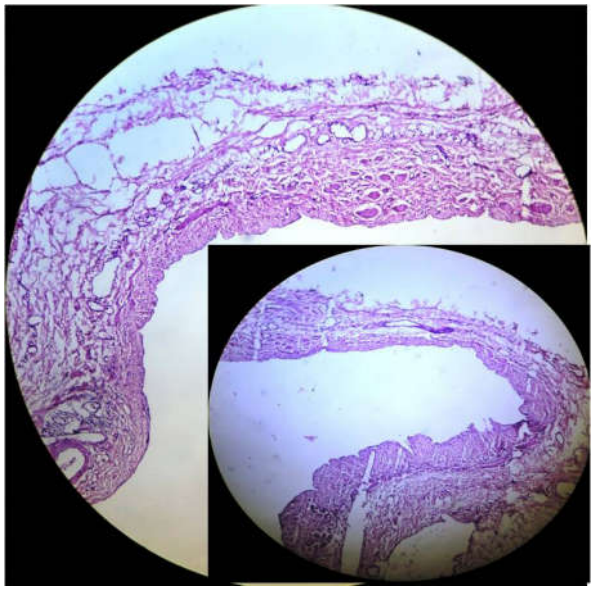


Figure 4. Cystic lumen lined by widely spaced thin endothelial cells with the supporting fibro cellular stroma with proliferation of small variable size vascular channels filled with lymphocytes [H &E stain, 10X] [Inset Multiple large irregular cystic luminal spaces] [H &E stain, 4X]

DISCUSSION

Lymphangioma is a benign congenital malformation of the lymphatic system (Güner *et al*., 2006). Some authors described cystic lymphangiomas occurring in the cervical area as cystic hygromas, a term first used by Wernher in 1843 (Emmanuel A. Ameh *et al*., 2011; Rolekar and Shah, 2014). The majority of lymphangiomas arise from parts of lymph sacs that are pinched off during development of or that fail to establish connections with the main lymphatic or venous channels 1843 (Emmanuel A. Ameh *et al*., 2011). Three theories have been proposed to explain the origin of this abnormality: (1) Blockage or arrest of normal growth of the primitive lymph channels occur during embryogenesis. (2) Primitive lymphatic sac does not reach the venous system. (3) During embryogenesis, lymphatic tissue lays in the wrong area (Kandakure *et al*., 2012). In adults, lymphangiomas may occur from delayed proliferation of lymphoid cell rests, either spontaneously or in response to infection, trauma, neoplasm or any iatrogenic injuries (Rolekar and Shah, 2014). Lymphatic malformations were formerly classified by Landing and Farber (1956). Furthermore histologic classification of lymphangiomas categorized into capillary (microscopic cavities), cavernous (cystic cavities upto 1cm in size) and cystic (cystic hygroma with cavity size larger than 2cm) which has no clinical usefulness (Rolekar and Shah, 2014; Derin *et al*., 2014). This nomenclature has been replaced by unifying classification of Mulliken, was adopted by the International Society for the Study of Vascular Anomalies in 1996. They categorized lymphangiomas into Macrocystic (formerly cystic hygroma), Microcystic (formerly cavernous lymphangioma) and Mixed variety (Emmanuel A. Ameh *et al*., 2011). The incidence of lymphangioma has been reported to range from 1.2-2.8 per 1000 newborn (Kandakure *et al*., 2012). This condition is approximately 50-60% appear before the end of 1st year of life and 80-90% before the end of 2nd year of life (Singh *et al*., 2014). However presentation may occur as late as adolescent or adult. There is no ethnic predisposition and male to female equal predilection reported in the literature but some reports stated male predominance (Emmanuel A. Ameh *et al*., 2011) which was seen in our case report. In head and neck region, cervical area is the predominant site for occurrence particularly the posterior triangle (75-80%), due to presence of extensive lymphatic system. Due to infiltrative nature of hygromas within the soft tissues of the neck, these may extend from posterior cervical area into the anterior compartment of neck which was in accordance with our case. It may cross the mid line, may reach into the cheek or down into the mediastinum and axilla (Kalsotra *et al*., 2001; Güner *et al*., 2006). The various sites reported are the head (especially tongue), chest wall, intraabdominal, groin, retroperitoneal space, pelvis, thigh and extremities (Emmanuel A. Ameh *et al*., 2011; Güner *et al*., 2006; Mathe and Dil, 2012). The major sign and symptom given by the patient is the presence of mass which is soft, non tender and ill defined. Most of them discovered early on account of their size and association of respiratory obstruction, feeding problem and sometimes pain associated with it (Kandakure *et al*., 2012). The present case was not having any symptoms apart from enlargement of swelling.

On clinical examination, differential diagnoses of cervical masses are hemangioma, lymphangioma, inclusion cyst (sublingual, submandibular salivary cyst), subaceous cyst and branchial cleft cyst, mycobacteria tuberculosis, salivary gland tumors, mucocele, carotid body tumor (Singh *et al*., 2014). Although the diagnosis of most lymphangiomas is clinical, various modalities like ultrasonography, plain radiography, computed tomography (CT) and MRI, needle aspiration and culture are required for confirmation of the diagnosis, planning of treatment and follow up (Emmanuel A. Ameh *et al*., 2011). Radiological investigations greatly aid in the preoperative diagnosis of lymphangioma with ultrasound scan (USS) being used as the initial mode of investigation, detecting the lesion. Macrocystic lymphangiomas typically appear as thin walled multiseptate, multicystic, hypoechoic masses (Kandakure *et al*., 2012). However, CT scan is significantly better at determining tumor content. It is especially helpful in determining the relations to major vessels and other surrounding structures; hence is essential in planning the surgery (Mathe and Dil, 2012). Lymphangiomas appear as low attenuation and fluid-filled masses on CT. Fluid-fluid levels can occasionally be seen representing acute or subacute bleeding into the cyst. MRI is an excellent modality to assess lesion extent in terms of tissue planes, airway compression, mediastinal extension, potential solid organ and bone involvement (Prashant *et al*., 2014). All these investigations had been performed in the present case report for better assessment of the lesion

On fine needle aspiration (FNAC), small round lymphocytes with intermingling histiocytes without mitosis or atypia is evident. In adults, FNAC may be therapeutic as well as diagnostic (Rolekar and Shah, 2014). Microscopically, the cysts are lined by endothelium, supported by stroma of varying thickness and containing smooth muscle elements and lymphoid tissue. The endothelial lining is quite vulnerable to infection and chemical irritants. The main histologic differential diagnosis is cavernous hemangioma in which blood filled large cystic spaces described similar to lymphangioma. But the endothelium of hemangioma gives positivity for pancytokeratin and Factor VIII immunohistochemically. All histologic and FNAC features were observed in our case that fulfills the criteria for cystic lymphangioma. The treatment of cervical hygromas has varied from neglect to complete excision. Many children with lymphangiomas present with preoperative complications, which can be life threatening, particularly in the cervical area like respiratory obstruction, rapid size increase, infection, ulceration, feeding and speech difficulty, mortality (Emmanuel A. Ameh *et al*., 2011). Therefore the mainstay's treatments for lymphangioma are sclerotherapy and surgical excision. OK 432 is one of the most commonly used sclerosing agents. It is produced by group A Streptococcus pyogenes. It produces an inflammatory reaction when applied with intracystic injection. Then it causes the destruction of endothelium, sclerosis, and cicatricial contraction of cyst wall. OK 432 should be used as the primary form of treatment for lymphangiomas especially in children due to good results and minimal complication rate (Mathe and Dil, 2012). Fever and local inflammatory reaction are the common side effects of OK 432. But hypopharyngeal edema was reported especially on administration for cysts located near the airway (Kalsotra *et al*., 2001). Surgical excision is known to be an ideal standard treatment for all types of lymphangioma. However, incomplete surgical resection is linked with recurrence. Recurrence rate accounts for 10% of cases due to difficulty in resecting the entire cyst wall (Prashant *et al*., 2014).

In our case, the cyst was fairly adherent to adjacent tissue, which included the sternocleidomastoid muscle, spinal

accessory nerve, and posterior cervical spinal nerves. Therefore, we preferred piecemeal excision to avoid adjacent tissue damage. Total excision is important for the prognosis, because cyst remnants usually cause tumor recurrence. In addition, it should be kept in mind that there are important complications of surgical therapy of cystic hygromas in the head and neck region such as cranial nerve injury, infection, bleeding, hematoma and postoperative sarcomas (Kandakure *et al*., 2012). Other treatment modalities like Laser therapy reduce cyst size but it has significant risk of damage to the overlying skin. Finally, interferon alpha is used on lymphangioma and hemangioma by means of antiangiogenic effect. The most common side effects of interferon alpha are fever, neutropenia, and diarrhea (Alani and Warren, 1992; Reinhardt *et al*., 1997). Radiation therapy, although occasionally used in the past, carries a risk of growth retardation and malignancy which is not recommended sarcoma (Kandakure *et al*., 2012).

Conclusion

Lymphangioma of the head and neck are benign neoplasms which are most common in children. Cervical cystic lymphangioma in an adult is a rare lesion in its clinical presentation. For proper management and to avoid complications, proper pre-operative evaluation, informed consent, proper workup should be done. This paper suggests surgical excision of such lesion is the best treatment modality as compared to laser therapy. However surgeons should ensure that total resection of the lesion without any remnant should be performed with the vital structures to be conserved during surgery.

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