CASE REPORT

RECURRENT CYSTIC LYMPHANGIOMA OF THE NECK IN AN ADULT: A CASE REPORT OF RARE ENTITY

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ABSTRACT
In adult, cystic lymphangioma is uncommon cause of cervical swelling. A 35 years old woman presented with a slowly enlarging neck swelling for 2 years duration that was excised and identified by pathology as a lymphangioma. She had a similar neck swelling at same location excised 14 years back. We reported this case to emphasize the cystic lymphangioma in differential diagnosis of neck swelling in adults. The literature is reviewed and classification, symptoms, diagnosis, and treatment of lymphangioma are discussed. For small, non-expanding swelling observation is required. Complete surgical excision remains the treatment of choice for cystic lymphangioma of enlarge, persist, obstructed to vital structure in neck or recurrent lymphangioma. This will help in reducing the rate of recurrence.
Key-Words: Lymphangioma; Neck Swelling; Adult; Excision; Recurrence

Introduction

Lymphangiomas are benign lymphatic malformations that occur most commonly in children and rarely in adult.[1] They have derived from lymphatic vessels with a progressive and painless growth. Most lymphangiomas are congenital, but they have also been reported to arise from infections, neoplasm, trauma and iatrogenic injuries in an adult.[1-3] Imaging techniques can aid in the precise mapping of the lesion and in defining its boundaries with the surrounding vital structure. Complete surgical excision considered to be best approach.

Case Reports

A 35 years old woman presented in our hospital with swelling in right side of lateral aspect of lower neck for 2 years, which was initially small in size of 2 x 2 cm, gradually increased to present size of 10 x 8 cm. She had a complaint of slight discomfort in neck movement. She had no pain over swelling, shortness of breath, dysphagia, dysphonia, no other swelling in body. On past history, she was operated for similar neck swelling in right side of neck at same location 14 years back. She remained asymptomatic for 12 years then she noticed a recurrence of present swelling in same location.

On clinical examination, swelling was soft, cystic, non-mobile, situated in posterior triangle of neck extended anterior up to posterior border of sternocleidomastoid muscle and posterior up to anterior border of trapezius muscle and inferior up to upper border of clavicle measured 12 x 8 x 4 cm size with transillumination and fluctuation test positive. Transverse scar was present, non-pulsatile, no dilated neck veins visible, no other swelling in neck present, trachea is centrally placed.

On investigation, blood tests were normal. USG neck suggestive of 8 x 7 cm size ill-defined anechoic cystic lesion with multiple septation and loculation present in supraclavicular region extending up to right posterior triangle. CT scan of neck revealed multiloculated cystic lesion of 84 x 30 x 83 mm size with thin peripheral wall and thin septation is noted in intermuscular and subcutaneous plane in posterior triangle of neck on right side without evidence of any fat density or calcification is seen within the lesion. Minimal post contrast enhancement of peripheral wall and septa is noted. The lesion is situated posterolateral to right sternocleidomastoid muscle, lateral to right carotid and jugular vessel and right scalene muscle anterior to right trapezius muscle and posterosuperior to clavicle and subclavian vessel suggestive of possible lymphangioma.

After taking informed written consent, patient had been taken for surgery. The swelling was approached with 6 cm transverse skin incision with 3cm above and parallel to upper border of clavicle. A 9x4x8cm size cystic swelling was circumferentially dissected and removed in its entirety. The Care has been taken during dissection to prevent injury to vital structure and also removed the remaining 2 to 3 cysts, which were located anterior to trapezius muscle. Final pathology report revealed cystic lymphangioma with characteristics of dilated lymphatic spaces with lymphoid follicles. Postoperative period was uneventful and she was discharged from the hospital on postoperative day 5.

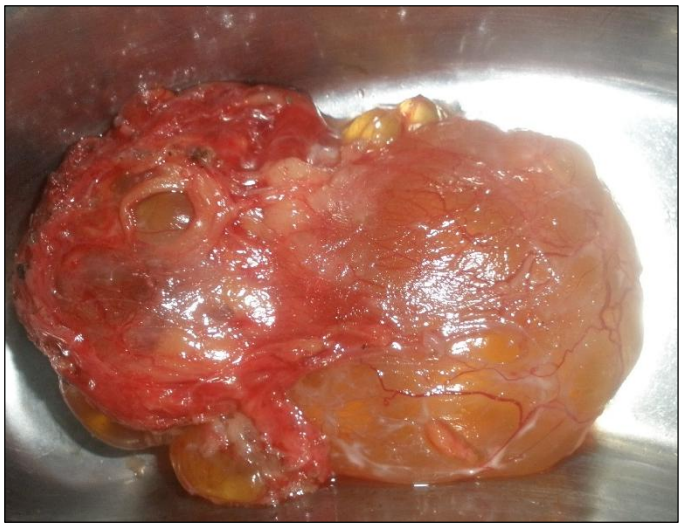

Figure-1: Cystic lymphangioma of the neck

Discussion

Lymphangiomas are benign lymphatic malformations that occur most commonly in children. Some authors refer to these lesions as cystic hygromas, a term first used by Wernher in 1843. Approximately 60% of lymphangiomas are present at birth, and up to 90% are detectable by 2 years of age.[1] Embryologically, these lesions are believed to originate from sequestration of lymphatic tissue from lymphatic sacs, during the development of lymphatico-venous sacs. These sequestered tissues fail to communicate with remainder of the lymphatic or venous system. Later on, dilatation of the sequestered lymphatic tissues results in the cystic lesions.[2] In adults, lymphangiomas may occur from delayed proliferation of cell rests, either spontaneously or in response to infection or trauma, neoplasm, or iatrogenic injuries.[3] It is rare for lymphangiomas to make their initial presentation during adulthood, but fewer cases have been reported. 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%).[4]

Lymphatic malformations can be categorized as superficial cutaneous lymphangioma, cavernous lymphangioma, cystic hygroma or diffuse systemic lymphangioma. They are also classified according to size: capillary lymphangiomas contain microscopic cavities, and cavernous lymphangiomas contain cystic cavities that are usually larger than 1 cm and cystic hygroma contain cystic cavities that are more than 2 cm.[5] Most lymphangiomas are asymptomatic. They have no gender predilection and 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 where it compresses surrounding tissue. There may be obstructive symptoms like dysphagia, dysphonia. Ultrasonography and computed tomography (CT) scanning have been used to evaluate the anatomy of lymphangiomas. On ultrasound examinations, these lymphatic malformations appear as thin-walled, multiseptate, multicystic, hypoechoic masses.[6] Lymphangiomas appear as multiloculated cystic lesions on CT scanning.[7] On T2-weighted MRI, lymphangiomas appear isointense to cerebrospinal fluid, whereas their intensity varies on T1-weighted images due to variable protein content.[8] Fine-needle aspiration (FNA) is routinely used to evaluate most neck masses, although it is almost never used in the paediatric population. In adults, where the risk of neoplasm is greater, there is no consensus in the literature concerning the use of FNA to diagnose these lesions. FNA findings include small and round lymphocytes with intermingling histiocytes without mitoses or atypia. In adults with compromised airways, FNA may be therapeutic, as well as diagnostic.[9] Microscopically, these lesions appear as endothelial-lined lymphatic spaces with intervening fibrous tissue and lymphoid aggregates.[10]

For small, nonexpanding lesions observation is done initially and those that persist, continue to grow, or present with obstructive symptoms should be resected. In adult, the main complaint is cosmetic unacceptability of mass. 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 hygroma within and around neurovascular structures, muscles, blood vessels. In this condition, unroofing, partial cystectomy and drainage of the cystic content can be performed. In this kind of treatment, recurrence rate of 10-15 % was reported.[11] To avoid injuring adjacent neurovascular structures, surgical planning is crucial before resection. Some surgeons prefer to use MRI to facilitate resection because it allows improved anatomical demarcation. Complications of resection include infection, bleeding, hematoma, and postoperative seromas. Injury to facial, hypoglossal, glossopharyngeal, recurrent laryngeal, and lingual nerves has been reported.[12]

Complete excision of a cystic hygroma has been shown to have an 81% cure rate. When only part of the lymphatic malformation is excised, there is an 88% recurrence rate.[4] Several studies have shown an increase in rates of recurrence, morbidity, and complications for lymphangiomas located in the suprahyoid versus infrahyoid region, whereas one study found no difference

in recurrence rates when comparing these locations.[11] Charabi and associates conducted a 35-year follow-up on 44 adult patients who were treated surgically for macrocystic lymphatic malformations of the head and neck.[12]Fifty percent of these patients had residual or recurrent disease, and 40% had complications involving speech, food intake, breathing, or swallowing. A direct correlation was observed between the extent of the lesion and the number of operations performed, rate of recurrence, and residual disease. Charabi and associates state that nonencapsulated lesions recur more frequently because of their tendency to infiltrate major neurovascular structures.[12] Baer and Davis reported an 89-year old with lymphangioma, the oldest such patient reported in the literature.[13] Thirty-two patients with cervical lymphangioma were treated at the Mayo Clinic; this is the largest series of the literature.[14]

Although neck and axilla are the most common sites, different places have been also reported. Ates et al reported on right adrenal gland, Shaffer et al reported thoracic lymphangioma, Nakazato et al reported a case in mediastinum.[15-17] Chung et al. reported a case in the breast and Solomou et al. reported a splenic lymphangioma.[18-19] Aneeshkumar et al. suggested that trauma could trigger formation of lymphangioma.[3] Sclerosing agents and radiation therapy have not been shown to play a role in the primary treatment of cystic hygromas. They may be indicated in lymphatic malformations that are macrocystic and not amenable to surgical resection.[14] Sclerotherapy with OK-432, a lyophilized, low-virulence SU strain of group A *Streptococcus pyogenes*, has been suggested as a possible therapy for macrocystic lesions. To date, however, the efficacy of OK-432 has not been proven in prospective, controlled trials.[14-16]

Conclusion

Although a rare, 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. Observation is recommended for small, non-expanding swelling. Complete Surgical excision remains the treatment of choice for large, persist or recurrent lymphangiomas in adult. This will help in lowering the rates of recurrence.

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