



Various Presentation of Lymphangioma

Authors

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Abstract

Introduction: Cystic tumors of neck and axila are rare in adults. Some of them include metastatic nodes, branchial cysts, thyroglossal cysts and lymphangiomas, among others. Clinically speaking, lymphangiomas are slow-growing soft masses located in different spaces of the neck and axila.

Case Report: This paper reports the case of a 38-year-old woman presenting with cyst in left submandibular region, who consulted due to a cyst in the left side of for evolution without associated systemic symptoms. Although relapse is frequent, the patient was successfully treated with surgery.

This paper reports the case of a 65-year-old woman presenting with lymph-angioma, who consulted due to a right lateral mass in the right lateral wall of breast for 2 years of evolution without associated systemic symptoms. Although relapse is frequent, the patient was successfully treated with surgery, without evidence of recurrence at 12 months of follow-up.

Discussion: When cystic tumors of the neck occur in children, surgical urgencies may arise due to obstruction of the airway. However, lymphangioma in adults only produce contour deformity and rarely require urgent intervention, which allows for conservative management such as observation, repeated drainage or sclerotherapy that can be done using OK-432 (Picibanil). Nevertheless, surgery remains a good treatment option, but some complications may occur.

Conclusion: Different treatment options were reviewed, which led to conclude that surgical resection of lymphangiomas continues to be a good treatment for this complex neck lesion and axila.

Keywords: Adult; Lymphangioma, Cystic; Sclerotherapy; Picibanil.

Introduction

Cystic lesions of the neck are rare and difficult to interpret for clinicians, since they can be benign or malignant pathologies. The most common location is the posterior triangle, where inflammatory, metastatic adenopathies or lymphoproliferative diseases can occur. In addition, congenital malformations such as branchial cysts, hemangiomas and lymphangiomas may be observed in this area. Branchial cleft cysts and hemangiomas are easily diagnosed in children less than 2 years of

age due to their clinical characteristics. These lesions are observed as a mass that usually does not involve inflammatory changes and presents a slow and painless growth or lymphangiomas, which are rare after the third decade of life.

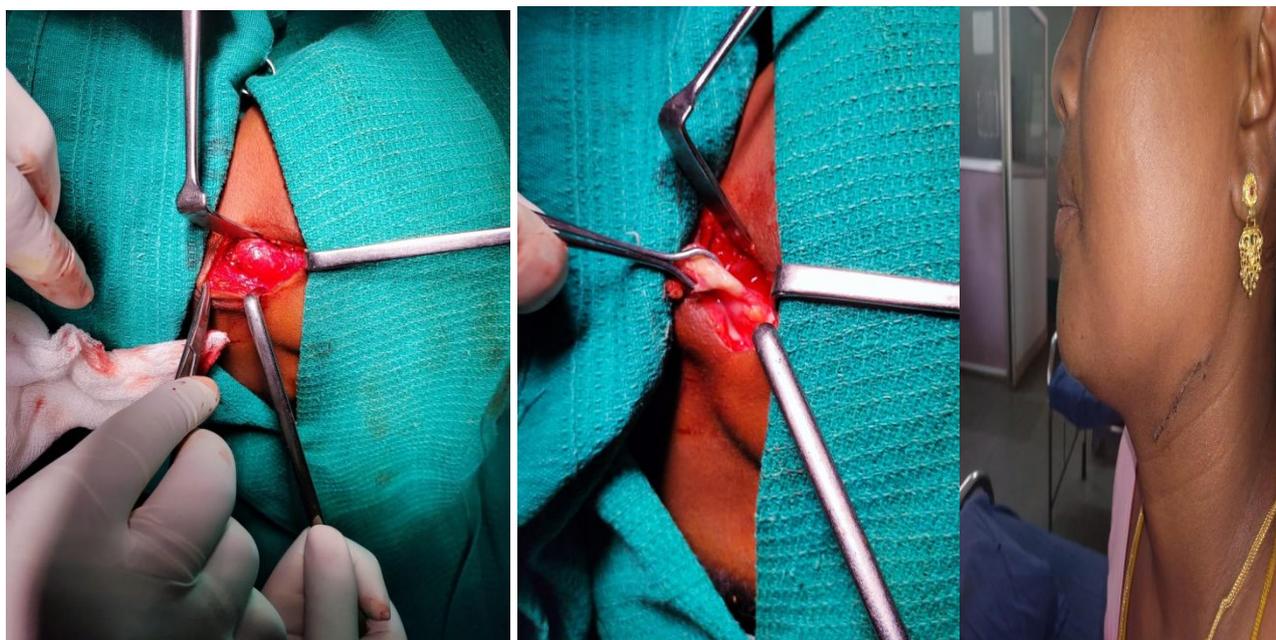
The current trend to treat lymphangiomas is different; conservative interventions such as sclerotherapy, now performed with new substances, have relegated surgery to the background due to the high rate of relapse or persistence. Considering that lymphangiomas are rare in adults and that the patient treated was a 36-

year-old woman who underwent a complete surgical resection, a review of the subject is presented with a description of the different treatment options.

Case Presentation

Case 1

Patient came with complaints of swelling in left side of neck and painful and developed over a period of 1 week and on examination, 3*2 cm swelling noted over left submandibular region. No warmth, non -Tender. Mouth opening adequate.



USG Report Neck- multiloculated cystic swelling left side of neck posterior to left submandibular gland 2.4*1.5 cm mildly abutting the submandibular gland. Possibility of infected cyst.

CT Neck Report- Enlarged level 1b and level 2 noted in left side of neck with areas of necrosis. Major salivary glands were normal. oropharynx, nasopharynx, oesophagus, trachea were normal. FNAC of swelling-shows cystic lesion in submandibular region.

Case 2

Patient came with complaints of swelling over Right axilla for past 2 years and No H/O pain/difficulty in lifting arm /nipple discharge/loss of appetite/body weight loss,

On examination- lump of size 15*10 cm noted over Right axillary region, No visible skin discoloration/No engorged veins. Swelling was freely mobile and firm in consistency and not fixed to skin/underlying structures. B/L Breast normal. No nipple retraction/dimpling/palpable lump/venous engorgement.

Mammogram Report- B/L Breast parenchyma normal. Well defined anechoic {cystic} lesion

noted in Right Anterior axillary line possibility of lymphangioma to be considered.

MRI Right Axilla Report- Well defined T2 and T1hyperintense cystic lesion with few septations noted in right axilla measuring 10.7*7.3 cm. The lesion causes mild displacement of pectoralis major muscle near its upper border and abutment of lateral chest wall

Surrounding sub cutaneous tissue appears normal
No significant enlarged lymph nodes

Visualized portions of both breast appears normal.
Neuro vascular structure appears normal.

Impression-cystic lesion in right axilla -possibility of lymphangioma.

traumatic, infectious and chronic inflammatory factors, as well as alterations in embryological development. The latter includes sequestration of lymphatic tissue, defects in the fusion of the venous system, and lymphatic obstruction that causes its expansion and obstruction. In adults, late proliferation of cellular nests of the lymphatic system may appear due to stimulation caused by trauma or infection. Lymphangiomas correspond to 25% of all vascular tumors in children and adolescents; 70% of cases are found in the head and the neck. In 2006, Thompson proposed classifying them into cystic (cystic hygroma)(2), capillary or cavernous, but there are different histological classifications:

Colbert *et al.* classified them into *macrocysts*, formerly known as cystic hygromas, and in *microcysts*, which may occur simultaneously in a lesion.

Bhayya *et al.* classified them in *lymphangioma simplex*, composed of small thin-walled lymphatics; *cavernous lymphangioma*, comprised of dilated lymphatic vessels with surrounding adventitia, *cystic lymphangioma*, consisting of a large lesion, with macroscopic lymphatic spaces surrounded by fibrovascular tissue and smooth muscle, and *benign lymphangio endothelioma*, lymphatic channels that seem to be dissected through dense collagen bundles.

In 1995, de Serres *et al.* proposed a classification according to anatomical location: *Stage I*, unilateral infrahyoid lesion; *Stage II*, unilateral suprahyoid lesion; *Stage III*, unilateral suprahyoid and infrahyoid lesion; *Stage IV*, bilateral suprahyoid lesion, and *Stage V*, bilateral suprahyoid and infrahyoid lesion.

Based on the last classification system, the case presented here corresponds to stage I (right lateral extension from a horizontal plane to the hyoid bone and to the superior clavicular ridge). This made dissection and complete resection easier, without evidence of residual tissue on the surgical bed.

The diagnostic studies used to determine cystic tumors of the neck include high resolution

ultrasound, which can prove the presence of a cystic lesion with multiple septa and without internal vascular flow that allows differentiating them from mixed vascular lesions. However, CT and magnetic resonance imaging (MRI) determine more precisely the relationship of the mass with the adjacent structures. The T1 and T2 sequences of the MRI facilitate the visualization of cleavage planes with the muscles and can define whether it is a vascular malformation.⁽³⁾

Fageeh *et al.* proposed observation, percutaneous drainage⁽⁴⁾, carbon dioxide laser (CO₂) and Nd-YAG laser, diathermy and surgical resection as management options. Recently, Miceli & Stewart recommended sclerotherapy with doxycycline or radiotherapy as treatment options. Observation may be indicated for patients younger than 3 years who do not have obstruction of the airway and have lesions <4cm, since they could present spontaneous regression.

Surgical resection has traditionally been advocated as the best treatment option, but in cases where the lesions extend into deep neck spaces, as the floor of the mouth or parapharyngeal space, complete removal may be difficult to achieve without damaging the nervous and vascular structures. This situation has led to seek alternatives such as drainage and sclerotherapy with different substances such as tetracycline, bleomycin and triamcinolone. Radiofrequency ablation is another therapeutic option to treat this entity. Complications and associated morbidity can include infections, tissue necrosis, cranial nerve lesions, vascular thrombosis and even endocrine disorders. Ogita *et al.* described the use of a streptococcal preparation OK-432 (Picibanil) as a safe alternative to perform sclerosis in cystic lesions of the neck, considering that this substance produces fibrosis secondary to inflammatory and cicatricial changes with the consequent contraction of the lymphangioma. OK-432 is a lyophilisate made of group A *Streptococcus pyogenes* incubated with penicillin, which is initially used as

immunotherapy for cancer in cases of pleural and peritoneal carcinomatosis.

These authors used this preparation in 83 patients with benign cystic lesions of the neck; 12 of them had lymphangiomas, achieving complete reduction in 67% and almost complete in 33% of the cases. Only some patients developed minor complications such as fever between 37.5°C and 38°C for a few days after the injection, which was controlled with antipyretics and without antibiotics.

Conclusions

Radiological images such as CT, MRI and neck ultrasound with FNA are of great help for diagnosing and planning the management of lymphangiomas of the .

Clinical suspicion of lymphangioma helps to establish the treatment for a patient with soft mass and radiological images suggestive of cysts. However, the definitive diagnosis is only obtained with the final pathology.

In adult patients, conservative observation treatment or sclerotherapy⁽⁵⁾ with different preparations such as picibanil are management options, especially when dealing with a complex lesion extended to the parapharyngeal space, submandibular space, pterygoid fossa or floor of the mouth.⁽⁶⁾

References

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