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Cystic Hygroma Mimicking Thyroid Lesion, Rare Presentation at Age of 15 Years

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ABSTRACT

Cystic hygromas are congenital lymphatic malformations of pediatric age group commonly in head and neck region. These arise from developmental defects in the lymphatic system. In this article we report a case of 15 years old male patient who presented with right supraclavicular swelling. Initial physical examination was followed by Ultrasonography which reported differential diagnosis as lymphatic malformation, cystic thyroid lesion. The histopathological examination of the surgically excised specimen confirmed it to be Cystic hygroma. Hence, histopathology plays an important role in confirming the diagnosis required for optimal management and better prognosis.

Keywords: Cystic Hygroma, Lymphatic malformation

INTRODUCTION

Lymphangiomas are the most common benign, painless tumors of lymphatic vessel. [1] Wernher classified the lymphatic malformation into 3 types: Cystic, Capillary and Cavernous. [2] Cystic Hygroma (CH) is a type of lymphangioma also known as cystic lymphangioma or Macrocystic lymphatic malformation. [2] It is a fluid filled sac, single or multiple, usually affecting head and neck region but can also affect any part of body. In neck it mostly lies in posterior triangle. [1]

It is commonly detected during pregnancy in female on ultrasound or in infants or children upto 2 years of age. It is rarely seen in adults. [1]

CH can occur due to genetic or environmental factor. Genetically, it can be due to failure to establish lymphatic connection to venous system or due to the abnormal budding of lymphatic tissue. It can be associated with Turner syndrome, Trisomy13, Trisomy18 or Trisomy 21. [3] They develop due to the failure of connection between the primary lymph sac and the venous system during their course of development. [4]

Environmental factor includes maternal viral infection with Parvovirus of Fifth disease, maternal substance abuse like alcohol. Diagnosis of CH can only be confirmed on histopathological examination. Complete surgical removal is the treatment of choice.

CASE PRESENTATION

A 15 years old male presented with gradual onset, painless swelling in right supra clavicular region of neck. The swelling was initially small which gradually increased. Patient did not give past history of any trauma or upper respiratory tract infection. No history of swallowing or breathing difficulty was noted. On Physical examination, a globular swelling which was insidious in onset, gradually progressive, approximately 3 cm in diameter was noted in right supraclavicular region of the neck. The swelling was mobile with soft doughy consistency and skin over the swelling was normal. The swelling was immobile on deglutination. No palpable cervical lymph nodes were identified around the swelling.

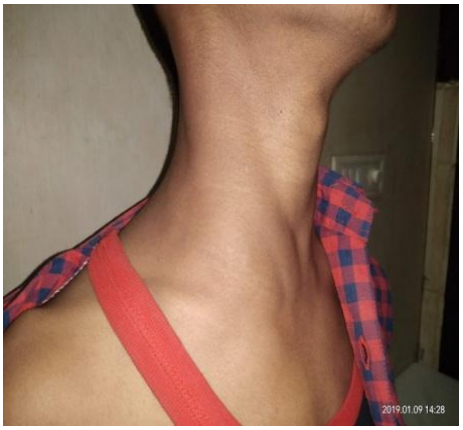
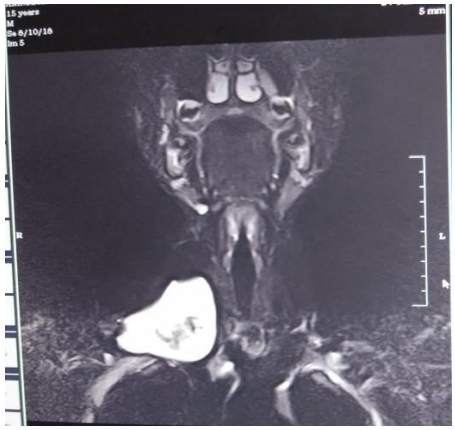

Figure 1. Photograph showing Swelling in Right supraclavicular region of neck
Figure 2. MRI showing swelling in Right supraclavicular region of neck

Patient underwent ultrasound examination which showed well defined anechoic collection measuring 7.6 x 7 x 2.7 cm in intramuscular plane over right side of lower neck crossing the midline and extending to left side suggestive of lymphatic malformation or cystic lesion of thyroid. On further radiological examination with CT scan neck and MRI cervical spine it was reported as a possible case of cystic hygroma and lymphatic malformation respectively.

On fine needle aspiration yielded 3 cc hemorrhagic fluid predominantly comprising of small lymphocytes admixed with macrophages in hemorrhagic background. No evidence of epithelioid granuloma or malignancy was noted. Cytomorphological features were suggestive of cystic lesion.

We received surgically resected cystic mass measuring 7x5x1.5 cms. External surface was greyish white, bosselated with smooth capsule. Cystic mass on cut section showed multiloculation and drained 2 ml of straw coloured fluid.

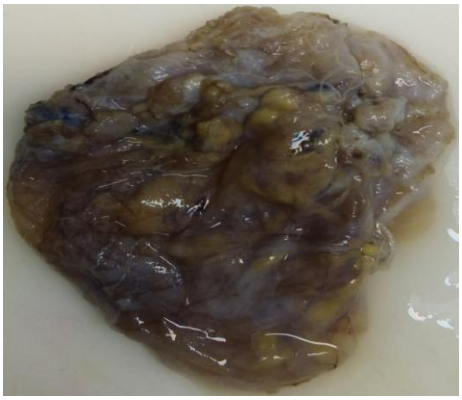

Fig.3 Gross appearance of cystic mass

Microscopic examination demonstrated multiple large cystic spaces. Cyst wall was lined by flattened endothelium and wall comprised of lymphoid population.

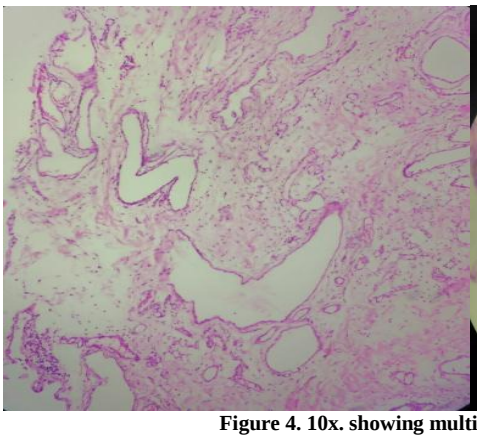
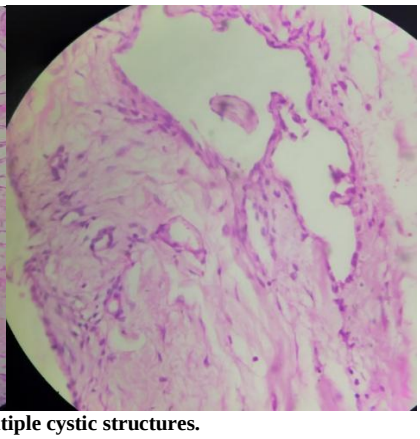

Figure 4. 10x. showing multiple cystic structures.
Figure 5. 40x. showing cyst lined by flattened endothelial cells.

DISCUSSION

Cystic hygroma or lymphangiomas are congenital lymphatic malformations majority are found under the age of 2 years. These lymphatic malformations are mostly seen in head and neck region[2]. Due to infiltrative nature of these lesions these may extend anteroposterior, cross midline or may reach upto mediastinum or axilla. [1] Various classification systems have been documented. Smith et al [5] described lymphatic malformations in three categories viz. microcystic, macrocystic and mixed. Kennedy [6] divided these lesions in to 4 types such as superficial cutaneous, cavernous, cystic hygroma and diffuse systemic. The staging system proposed by de Serres [7] classified them according to location and extent of lesion: Stage 1- Unilateral infrahyoid, Stage 2 - Unilateral Suprahyoid, Stage 3- Unilateral Suprahyoid and infrahyoid, Stage 4 – Bilateral infrahyoid; Stage 5 Bilateral infrahyoid and suprahyoid

Differential diagnosis of neck swelling includes thyroid swelling, Thyroglossal cyst, Brachial cleft cyst, tubercular lymphadenitis. Other d/d includes soft tissue sarcoma, abscess, and hematoma. Therefore, there is high possibility of misdiagnosis. [8]

Surgical resection is the treatment of choice for the benign lesions. Surgical complications include infections, injury to major neck vessels, sepsis, fistula formation, nerve injury, facial palsy, vocal cord palsy, injury to brachial plexus. [9]

Partial removal can lead to recurrence in 10 – 15% cases. [9] Fliegelman et al. [08] reported recurrence in relation to histological encapsulation of lesion. Delay in surgery can lead to complications like cyst rupture, spontaneous bacterial infection, and adhesion. Thus long term follow up is advised.

There are three subtypes of cystic hygromas.: capillary or simplex lymphangioma which has characteristic small and thin-walled channels, cavernous lymphangioma contains larger, thin-walled channels and third subtype is cystic lymphangioma (hygroma) which contains flattened endothelium along with large cystic spaces. [12]

In Summary, we report a case of cystic hygroma in 15 years old male who presented with right supraclavicular swelling. With this unusual presentation of lesion, it could have lead to misdiagnosis by other radiological techniques, but the diagnosis was confirmed by histopathology. Therefore, clinical, radiological and pathological correlation is most important for the management of such lesions.

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