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Case Study

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LYMPHATIC MALFORMATION - A SERIES OF THREE CASES WITH VARIABLE PRESENTATIONS IN A RURAL TERTIARY CARE CENTRE.

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ABSTRACT: Lymphatic malformations (LMs) are slow flow vascular malformations due to a localized abnormality affecting the development of the lymphatic system. LM is synonymous with cystic lymphangioma or cystic hygroma or hygroma cysticum coli, which is also known as a macrocystic lymphatic malformation. It was first described in 1828 by Redenbacker and the term cystic hygroma was coined by Werner in 1834. LMs usually affect head and neck (~75%), with a predilection for the left side. Within the neck, the posterior triangle tends to be the most frequently affected. Review of literature, clinical features, pathology and treatment of these lesions in 3 cases are discussed and reported in the present case series.

KEYWORDS: Lymphatic malformations; Cystic hygroma; Transillumination.

INTRODUCTION

Cystic hygroma or the lymphatic malformations are benign congenital malformation of the lymphatic system that occur due to lack of development of communication between the lymphatic and venous systems. The cyst may be unilocular or multilocular and could be of variable size but is characteristically brilliantly transilluminant. The incidence of cystic hygroma is approximately 1/6000 live births [1]. 70–80% of cystic hygromas occur in the neck, usually in the posterior cervical triangle.[2] The remainder 20–30% occurs in the axilla,superior mediastinum, chest wall, mesentery, retroperitoneal region, pelvis and lower limbs.[3] Cystic hygroma is known to present at birth in about 50% of the affected newborns and 90% present by the age of 2 years.[4] Here, the authors present a series of 3 such cases of cystic hygroma with variable presentation, reported in the department of Otorhinolaryngology at B.P.S Government medical college for women, Khanpur Kalan, a rural tertiary care centre.

Case 1

A 17-year-old female came to Department of E.N.T. with complaint of swelling on right side of neck since 2 years. Patient was asymptomatic 2 years back after which she noticed a swelling in the neck; which was initially small and had gradually increased. Patient did not give any history of trauma or upper respiratory tract infection. On examination, there was a large single swelling on the right side of neck measuring 10 cm x 5 cm. The swelling was soft, non-tender, nonfluctuant, brilliantly translucent extending into both anterior and posterior triangle of the neck. The patient underwent ultrasonography (USG) neck followed by Contrast Computed Tomography (CT) scan of neck and thorax which showed a large non enhancing hypodense multiloculated thin walled cystic lesion on the right side of the neck extending from submandibular to supraclavicular region involving anterior as well as posterior triangle with mass effect and was suggestive of a lymphatic malformation. A preoperative diagnosis of lymphatic malformation suspicious of cystic hygroma was made.

The patient underwent surgical excision of the mass and specimen was sent for histopathological examination which confirmed the diagnosis of Cystic Hygroma.

Case 2

A 13-year-old male came to Department of E.N.T. with complaint of swelling on left side of neck since childhood. Patient complained of pain and increase in the size of swelling after an attack of upper respiratory tract infection 2 years back. On examination, there was a single large swelling on the left side of neck measuring 8 cm x 5 cm. The swelling was soft, compressible, non pulsatile, non-tender, brilliantly translucent situated in the region of posterior triangle of the neck. M.R.I findings showed a multiloculated, hypodense cystic mass with fluid attenuation of size 8 x 5 x 4 cm, in the posterior cervical triangle. The lesion was hyperintense on T2-weighted images and hypointense on postcontrast T1-weighted images. A preoperative diagnosis of

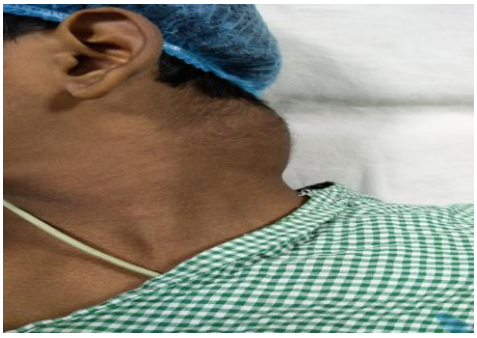
lymphatic malformation suspicious of cystic hygroma was made.

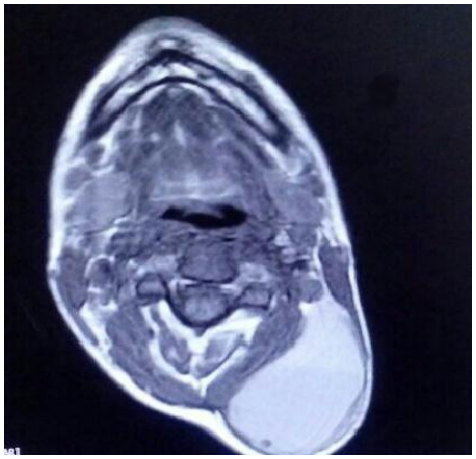
The patient underwent surgical excision of the mass and specimen was sent for histopathological examination which confirmed the diagnosis of Cystic Hygroma.

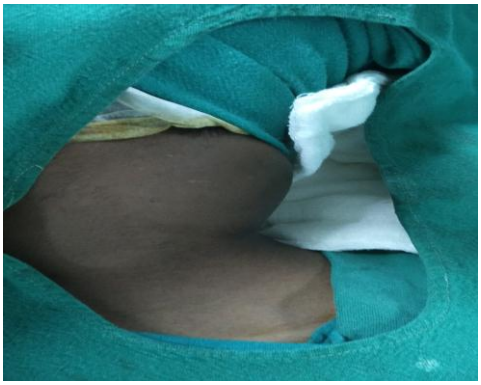
Case 3

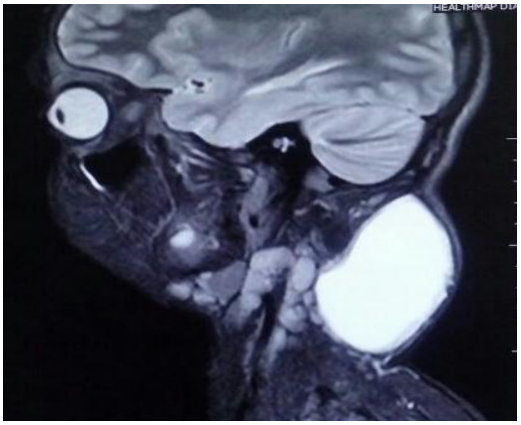
A 5 year old boy presented with complaints of swelling in the left side of the neck for 2 months. Previous history showed that the child was apparently normal except for a history of respiratory tract infection 2 weeks before the onset of swelling. Initially the swelling was small and gradually increased to attain the present size. On inspection, the swelling was approximately 3x2.5 cm, present in the left upper part of the neck.. Swelling was mobile, cystic, fluctuant, non-tender, non-pulsatile and margins were not well defined. Transillumination test was positive. USG was performed. The report described an irregular, multiloculated, cystic swelling of 4.3x2 cm size in the left upper anterior triangle of neck extending to the posterior triangle of neck and was diagnosed as cystic hygroma. Complete surgical excision was done and histopathology report confirmed the diagnosis of cystic hygroma.

Images









DISCUSSION

Cystic hygroma is an aberrant proliferation of lymphatic vessels resulting from abnormal development of the lymphatic system.[1,2] Within the literature the term cystic hygroma is used interchangeably with lymphangioma and lymphatic malformation. It is believed to arise from a congenital malformation of the lymphatic system in which a failure of communication between the lymphatic and venous pathways leads to lymph accumulation. Most cystic hygromas present in-utero or in infancy. They are quite rare in adults.[2]

It is usually multiloculated and contains clear lymph fluid; the wall of the sac is lined by a single layer of flattened epithelium. The cysts intercommunicate with each other and can insinuate between muscle planes, this gives it a sign of compressibility. Large lesions can compress many vital structures in the neck like, sympathetic chain, contents of the carotid sheath and branches of hypoglossal, lingual, and the facial nerves.[5,6]

Cystic hygromas extends between the compartments of the neck due to its infiltrative nature within the soft tissues. It may also cross the midline, reach into the cheek or extend into the mediastinum and axilla and can involve the recurrent laryngeal nerve and the brachial plexus. Symptoms can vary from a painless, enlarging mass to dysphagia, respiratory compromise, and difficulty in feeding with regurgitation[7,8]. It has been established that two third of the cases are asymptomatic

and literature suggests that cystic hygroma may be triggered by upper respiratory tract infections.[7]

Imaging studies are important to assess extent of the lesion. Although ultrasound scanning is sufficient to establish the diagnosis, computed tomography (CT) or MRI is useful to show adjacent tissue extension.[5,9]

The differential diagnosis of cervical masses include lymphadenitis (caused by mycobacteria tuberculosis, other bacterial and viral infections), inclusion cyst of submandibular gland, branchial cleft cyst, laryngocele, haemangioma, lymphoma and congenital vascular malformations [10,11]. Surgical excision is the treatment of choice. En bloc resection is difficult due to the adhesive characteristics of the tumors. Inadequate surgical intervention often leads to recurrent disease.

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