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Case Report

Cervical Cystic Hygroma in a 6-year old child – A surgical case report

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ABSTRACT

Background: Cystic hygroma is an uncustomary benign neck swelling observed in the paediatric population aged less than two years.Generally, presents as a birth defect but it can also develop later in life. It is a congenital primary lymphatic malformation commonly located along the major lymphatic channels in the neck and axilla caused due to sequestration of the jugular lymph sac. It can lead to obstetric complications like obstructed labour and later may cause respiratory distress or septicaemia. In this paper, we discuss the case of a 6 year old child coming with right side lateral swelling of neck since 2 years.

Keywords : cystic hygroma ,lymphangioma , congenital , excision.

INTRODUCTION

Cystic hygroma, also known as cavernous lymphangioma or hygromacysticum coli is a congenital lymphatic malformation. 'Cyst' means a fluid-filled sac lined by single layer of epithelium. 'Hygroma' is derived from Greek words 'hygros' meaning moist/watery and 'oma' meaning tumor[1].

It is a benign birth defect with origins which trace back to the embryonic life. The lymphatic malformation causes sequestration of lymph near the major lymphatic channels henceforth most cases present with painless swelling in posterior triangle of the neck in supraclavicular region(75%) or axilla(20%). Other sites include cheek, tongue, groin, mediastinum[2].

Mostly idiopathic but at times it can be inherited as an autosomal recessive disorder. Other causes can be maternal viral infections or maternal substance abuse during the course of pregnancy.

90% of the cases present within 2 years of age and are quite rare in adults often hypothesised to occur following proliferation of lymph vessels in response to trauma or infections involving head and neck[3]. If it appears within 30 weeks of gestation it is associated with Turner syndrome, Noonan syndrome, trisomy, non-immune foetal hydrops, or cardiac anomalies. These children have a poor prognosis.

CASE REPOR

A 6 year old child came with painless swelling in the right side of the neck since 2 years . The swelling was initially small in size of the size of a pea and then progressed gradually over 2 years to the size of a lemon.

On examination swelling was soft , cystic in consistency , compressible present on the right lateral side of neck below the mandible. The swelling showed partial increase in size on coughing.

IMAGE 1 – SHOWING SWELLING IN RIGHT LATERAL SIDE NECK

FNAC and CT scan of the neck was done. CT scan neck was done which demonstrated a well circumscribed , homogenously hypoechoic cystic lesion , below the right mandibular angle anterior to the carotid bifurcation measuring 2 cm *2 cm*3 cm in size.

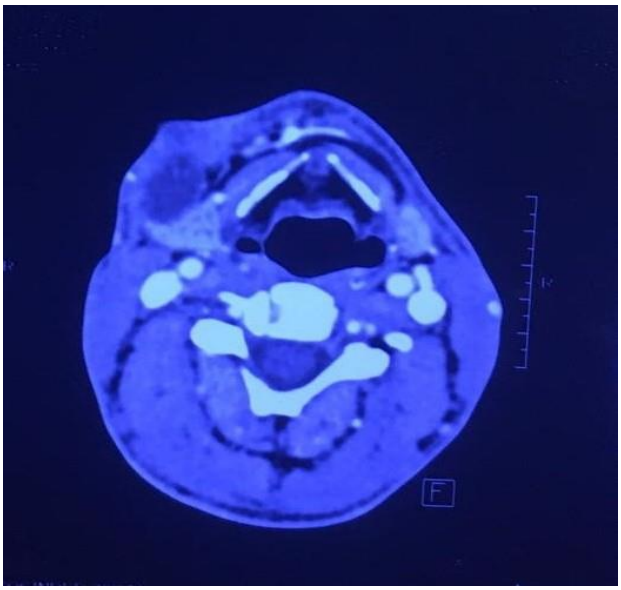

IMAGE 2 – WELL CIRCUMSCRIBED SOFT TISSUE LESION below the right mandible

Excision of the cystic lesion was done and the specimen was sent for histopathology which confirmed the presence of Cystic Hygroma.

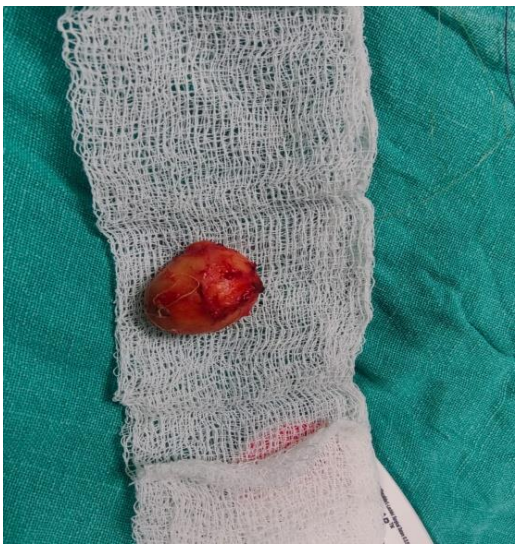
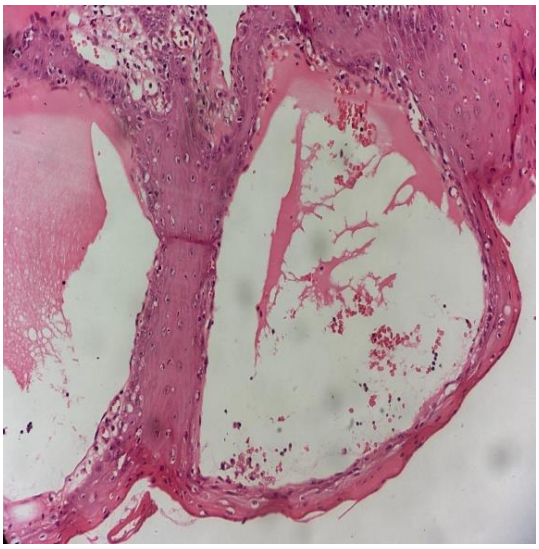

IMAGE 3 – EXCISED SPECIMEN IMAGE 4

IMAGE 4 - On Histopathology ,(**H&E stain)**showed stratified squamous epithelium with dilated lymphatic channels confirming it as Cystic Hygroma**(IMAGE 4)** .

DISCUSSION

The lymphatic system is an intricate, efficient, and vital system of our body. Our body has two main transit systems for transport of fluid, cells, nutrients, and waste: the blood system and the lymphatic system. The blood system carries 85% of these constituents back to the heart. The rest 15% are picked up by the lymphatic system which supports the blood system, immune system, and digestive system. Lymph is made up of proteins, fats, immune cells, and extra fluid which leaks out of the blood system.

Lymphatic malformation means that the lymphatic system does not form correctly. In cystic lymphatic malformations the lymph accumulates forming cysts. They can be microcystic, macrocystic or mixed. Microcystic appear very small and sponge-like clusters. Macrocystic are >2cm looking like small, smooth, translucent bubbles under the skin which may give it a blue tint. They can turn painful and cause pressure symptoms.

Histologically, lymphangiomas can be classified into two groups based on depth and size of abnormal lymph vessels. The superficial (<5cm) are called lymphangioma simplex orcircumscriptum having thin-walled lymphatic vessels[4]. The deep-seated are called cavernous lymphangioma or cystic hygroma distinguished by dilated lymphatic vessels.

Cystic hygroma is hence a benign congenital lymphatic malformation[5]. It is a cystic swelling caused due to sequestration of lymph in the jugular lymph sac during intrauterine life.90% cases are diagnosed before 2 years of age.

McGill and Mulliken classified cystic hygromas on the basis of anatomical location, histology, and CT findings[6]. This classification serves as a clinically relevant way to categorize cystic hygromas into Type I malformations which are located below the mylohyoid muscle, are macrocystic involving the anterior and posterior triangles of the neck. Type II malformations are located above the mylohyoid, are microcystic and invasive.

Child presents with swelling most commonly in posterior triangle of neck in supraclavicular region or axilla. The swelling is soft, smooth, multilocular, fluctuant, brilliantly transilluminates and partially compressible.The swelling increases in size on crying or coughing. It may or may not be painful.Disfigurement of the face of the child is a more worrisome factor for the parents. It can be emotionally distressing and socially isolating for the child.

The swelling is an aggregation of interconnected cysts which appear like soap bubbles. It shows a mosaic appearance with larger cysts near the surface and smaller cysts which are deep-seated. Each cyst is a sac of clear lymph lined by single layer of flattened epithelium. The fluid does not coagulate.

Due to its infiltrative nature within the soft tissues, cystic hygromas can extend between the compartments of the neck. It can also cross the midline and extend to the cheek, mediastinum, or axilla. It may also involve laryngeal and pharyngeal structures leading to complications.

When present since birth it may cause obstructed labour. Involvement of structures in larynx and pharynx can cause stridor, feeding problems, dyspnoea. Swelling may become large and compress surrounding vital structures like the sympathetic chain, carotid sheath, branches of hypoglossal, lingual, and facial nerves. Respiratory obstruction may require emergency tracheostomy. It may become infected forming an abscess which may lead to septicaemia.

The central conducting lymphatics transport chyle from the intestines. If these are malformed the body starts forming new routes for transportation. These routes however do not reach the heart causing the lymph to pool. It may leak into the chest causing a chylothorax or into the abdomen causing chylous ascites.

Diagnosis for cystic hygroma is made by corelating the findings of USG, CT or MRI with findings on clinical examination. Genetic testing can be done if chromosomal anomalies are suspected.

Conservative management involves intralesional injections of sclerosing agents like OKA-432, PICIBANIL. OKA-432 is inactive strain of group A Streptococcus pyogenes. Other sclerosing agents like ETHANOL, BLEOMYCIN, RIFAMPICIN, CORTICOSTEROIDS can also be used. Bleomycin is an antineoplastic drug which causes fibrosis of the cyst wall. Other modalities include radiofrequency ablation or surgical excision.Surgical excision of the swelling while preserving surrounding neural and vascular structures remains the most common treatment modality.Care should be taken to perform meticulous dissection across all planes to clear entire cyst wall.If there is abscess formation, then first under proper antibiotic coverage the abscess should be drained and later the sac should be excised. Macroscopic removal of the tumour leaves only 5% rate of recurrence. Resection is difficult if swelling is present on the lip, tongue or oral cavity.

Differential diagnosis of cystic hygroma arebranchial cleft cyst, inclusion cyst of the submandibular gland, lymphadenitis, laryngocoele, hemangioma, lymphoma, congenital vascular deformities.

CONCLUSION

Cystic hygroma is a benign congenital lymphatic malformation. It is a cystic swelling caused due to sequestration of lymph in the jugular lymph sac during intrauterine life.90% cases are diagnosed before 2 years of age.However it can manifest in later stages of life. Branchialcyst ,Laryngoceleand Lymphoma should be kept as differential diagnosis. Excision is the main line of management.

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