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Case-Report Open Access

Cystic Hygroma- A Case Report and its Embryological Basis

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Abstract

Cystic hygroma is an uncommon swelling in the neck among paediatric population. It is a fluid filled sac resulting from blockage in the lymphatic system and is commonly located in the cervical region and axilla. It usually presents as a birth defect or can develop at any time. A case report of a 2 year old developmentally normal female child presenting with cystic hygroma in the right side of the neck triggered by respiratory tract infection has been described. The objective of this report is to discuss the embryological basis of the cystic hygroma

Keywords: Cystic hygroma; Lymphatic system; Embryological basis

Introduction

Cystic hygroma or hygroma cysticum coli are also called as cystic lymphatic malformations. It is a benign congenital malformation of the lymphatic system. It usually presents within 3 years of life. Approximately 50 - 60% of these malformations appear before one year and 80-90% before the end of second year of life [1]. When cystic hygroma appears before 30 weeks of gestation it is associated with Turner syndrome, Noonan syndrome, trisomies, fetal hydrops and cardiac anomalies. The prognosis is poor for these types of hygromas [2-7]. Occasionally cystic hygroma is inherited as an autosomal recessive disorder. Its most common cause is idiopathic. The other causes being maternal viral infection such as parvo virus and maternal substance abuse like alcohol. In most cases of cystic hygromas, 70-80% appears in the neck and lower part of the face due to its rich lymphatic drainage. Other sites are axilla, superior mediastinum, retroperitoneum, mesentry, pelvis and lower limbs [3].

Case Report

A 2 year old developmentally normal female child presented with complaints of swelling in the right side of the neck for 20 days. 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 right upper part of the neck. Margins were not well defined. Swelling was mobile, cystic, fluctuant, non-tender, non-pulsatile (Figure 1). Transillumination test was positive. USG was performed. The report described an irregular, multiloculated, cystic swelling of 4.3x2 cm size in the right upper anterior triangle of neck extending to the posterior triangle of neck and was diagnosed as cystic hygroma (Figure 2).

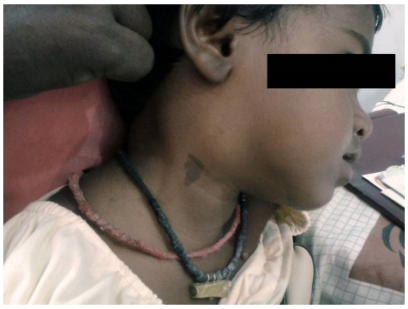

Figure 1: Right lateral view of neck showing the swelling over the upper part of the neck.

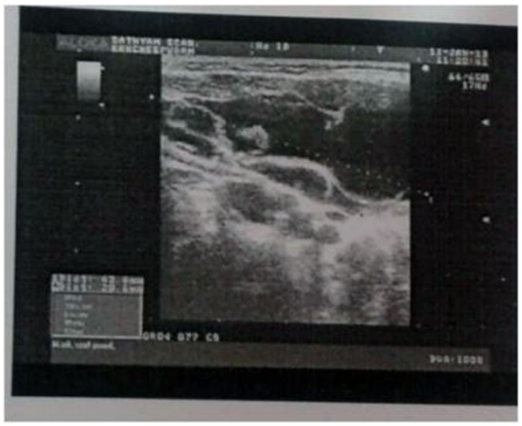

Figure 2: USG of the neck showing cystic hygroma.

Discussion

Lymphangiomas are divided histologically into two major groups based on the depth and the size of abnormal lymph vessels. The superficial ones are called lymphangioma circumscriptum. The more deep seated ones are cavernous lymphangioma or cystic hygroma [4]. Another classification is to divide them into three types as capillary, cavernous and cystic [5]. McGill and Mulliken proposed a classification of cystic hygromas based on location, histology and CT findings [6]. Type I malformations develop below the mylohyoid muscle and are macrocystic involving the anterior and posterior triangles of the neck. Type II malformations are found in the neck above the level of the mylohyoid and are invasive and microcystic. Resection will be difficult if it presents on the lip, tongue or oral cavity. 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].

Cystic hygroma 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, the sympathetic chain, contents of the carotid sheath and branches of hypoglossal, lingual, and the facial nerves [8].

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 [9,10].

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 [11].

We shall now discuss the embryological basis of cystic hygromas.

Embryological Basis

There are two mechanisms suggested for the origin and pathophysiology of cystic hygroma. Sabin described a theory of a venous origin with a centrifugal spread [12]. Huntington and McClure proposed a mesenchymal origin with a centripetal spread. Van der Jagt and Kutsuna supported a theory of combined venous and mesenchymal origin [13].

The lymphatic system begins to develop in the end of the 6th week approximately 2 weeks after the development of the recognizable primitive cardiovascular system. In the 8th of gestation of gestation, six lymphatic sacs are identified in the developing embryo, two are located lateral to jugular vein, two are iliac, one is retroperitoneal and the last one is the cysterna chylli. Finally the lymphatic sacs communicate with the venous system. During 9th week of gestation the lymphatic sacs are invaded by connective tissue to form lymph nodes [14].

Embryologically, lymphangiomas are thought to originate from lymphatic tissue sequestration from the primitive lymphatic sacs, during development of lymphatico-venous sacs. These sequestered tissues fail to communicate with remainder of the lymphatic or venous system. Following which, dilatation of the sequestered lymphatic tissues occurs, resulting in the cystic morphology of these lesions [15].

A genetic basis of cystic hygroma has also been proposed. Cystic hygroma commonly occurs in 0.5% cases of spontaneous abortions and up to 1 in 700 cases of low risk pregnancies but is seen rarely in the neonate. It is also suggested that 20% of fetuses with a cystic hygroma will have a normal karyotype [16]. Garabedian and Wallerstien et al in their study establishes the occurrence of cystic hygroma due to a deletion of 16q24.1 with haploinsufficiency of FOXF1 and FOXC2 genes. They recommend prenatal diagnosis of FOX gene clusters and mutation for the diagnosis of fetal hydrops and cystic hygromas [17].

Conclusion

The embryology of cystic hygroma is now better understood. The incidence of diagnosis of these lesions has increased due to routine antenatal ultrasound screening. These cases may rarely require genetic counseling following prenatal diagnosis and for successive pregnancies.

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