An Atlas of Lumps and Bumps: Part 16

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Cystic Hygroma

A cystic hygroma is a benign lymphatic malformation resulting either from an abnormality in the control of lymphatic growth, remnants of embryonic lymphatic tissue that retains the potential for proliferation, or from an arrest in the normal development of the primitive lymphatic channel whereby the peripheral lymphatic vessel becomes sequestrated and fails to communicate with the remaining lymphatic system.[1-5] The formation of a cystic structure containing the accumulated lymphatic fluid outside the lymphatic system is referred to as a cystic hygroma.[6] Histologically, the lesion consists of multiloculated cysts lined by endothelial cells with serous lymphatic fluid inside the cystic structure. Some cysts communicate with each other, whereas others are separated.

The incidence of cystic hygromas is approximately 1 in 12,000 live births.[5] The male-to-female ratio is approximately equal.[3,4] In the majority of cases, the cause is idiopathic.[7] Cystic hygroma might follow maternal exposure to alcohol, aminopterin, or trimethadione.[3,4] The condition is more common in patients with chromosomal abnormalities (such as Turner syndrome, Klinefelter syndrome, trisomy 13, trisomy 18, and Down syndrome) and in patients with Noonan syndrome, Proteus syndrome, multiple pterygium syndrome, Roberts syndrome, fetal alcohol syndrome, and Beckwith-Wiedemann syndrome.[8-10] The condition may also be inherited as an autosomal recessive and autosomal dominant trait.[7,11]

A cystic hygroma typically presents as a soft, nontender, multilocular cystic swelling that is compressible, fluctuant, and brilliantly translucent **(Figures 1 and 2)**. The fluid contained inside the cyst is serous, serosanguinous, or straw-colored.[2,7,9,12] The size varies from 1 to 30 cm

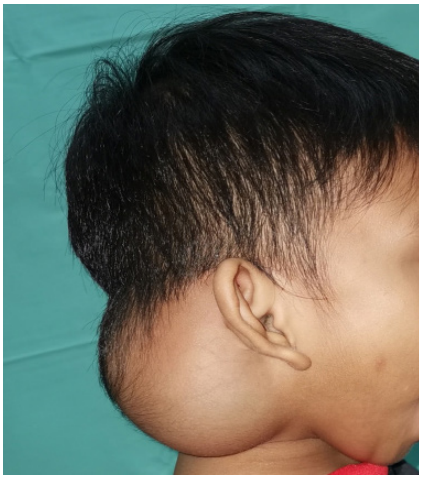

Figure 1. A cystic hygroma typically presents as a soft, nontender, multilocular cystic swelling that is compressible, and fluctuant.

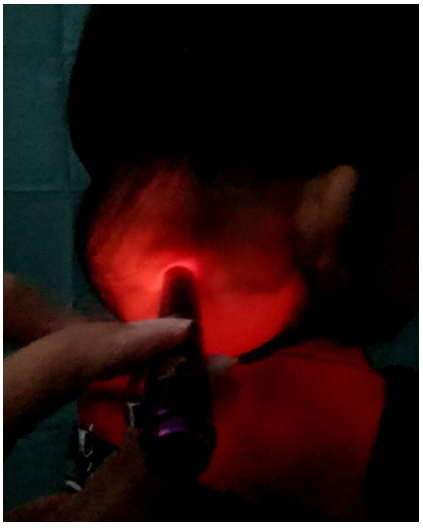

Figure 2. A cystic hygroma typically presents as a brilliantly translucent mass.

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in diameter.[1,6] Typically, the lesion is not attached to the skin but may be attached to underlying structures. The overlying skin is normal.[6] Approximately 50% to 60% of cases are identified before 1 year of age, and 80% to 90% of cases are diagnosed by 3 years of age.[1,7,13] Onset in adulthood is rare.[1,14] Approximately 75% of cystic hygromas occur in the neck, typically in the posterior cervical triangle, and lower part of the face.[1-4,6,7,12,13] Cystic hygromas occur twice as often on the left side of the neck than on the right.[5] They are usually unilateral but can be bilateral. Approximately 20% of the lesions occur in the axilla.[1,6] Other less common sites include the mediastinum, omentum, mesentery, retroperitoneum, trunk, inguinal area, extremities, and rarely, in the cheek, suprasternal area, and even in the epidural space and spinal soft tissue.[1-4,15] The growth of a cystic hygroma is usually, but not always, proportional to the growth of the child. The growth may increase during pregnancy. Cases of rapid enlargement of the lesion over a short period of time have been reported.[2] The lesion rarely regresses spontaneously.

Although cystic hygromas are benign lesions, they have the potential for extension or infiltration into the surrounding structures such as the soft tissue of the neck and mediastinum and may cross the midline.[1,2] Depending on the structures involved, airway obstruction, obstructive sleep apnea, dysphonia, dysphagia, feeding problems, and restriction of neck movement might result.[2,6,7,16] Rarely, huge cystic hygromas may cause neural encroachment.[1] Other potential complications include rupture, infections that may lead to abscess formation, and hemorrhage within the cystic hygroma.[2,6] Fetal cystic hygroma, especially in the presence of concomitant congenital anomalies and hydrops fetalis, is associated with adverse pregnancy outcomes.[10]

Cystic hygromas and their congenital aneuploidies can be first noted during prenatal ultrasonography scanning.[17] Increased nuchal thickness may be noted as early as the first trimester.[17] Postnatally, the diagnosis is mainly clinical based on the characteristic features of the lesion. Ultrasonography scanning may be used to define the size and extension of the lesion and to demonstrate the multicystic nature of the lesion with no flow on color Doppler study.[12]

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