

Teaching Round

Congenital Massive Neck Swelling in a Neonate

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A four day old neonate presented with a massive swelling on the left side of neck since birth (Figure 1). The swelling was cystic, partially compressible having lobulated surface and margins were not well defined. It was brilliantly translucent. Regional lymph nodes were not involved and there was no evidence of swelling in any other part of body. The swelling increased in size when the child cried. Initially it occupied the lower part of neck and gradually extended upwards towards the ear. There were no signs of inflammation over the swelling and there was no respiratory compromise. It was fullterm vaginal delivery carried out at home, had cried immediately after birth and there was no history of birth trauma. The neonate was on exclusive breast feeds.

What is the clinical diagnosis?

A clinical diagnosis of cystic hygroma was entertained in this case because of the characteristic nature of the swelling- congenital

nature, typical location, cystic nature, partially compressible, ill-defined margins, increase in size on crying and the brilliantly translucent nature. A cystic hygroma is a congenital multiloculated lymphatic lesion that can arise anywhere, but is classically found in the left posterior triangle of the neck [1]. It was first described by Wernher in 1843. Cystic hygroma is most frequently encountered in the head and neck (75%) with a left-sided predilection, while approximately 20% occur in the axilla [2]. It is a lymphatic malformation which manifests itself during early infancy, and 90% of them are detected before the end of second year of life.

Enumerate other causes of congenital neck swelling in a neonate?

Congenital neck swellings in a neonate could have several causes apart from cystic hygroma [3]. These include teratoma, lipoma, haemangiomas, branchial cleft cyst, ranula, thyroglossal duct cyst. Lymphadenopathy, thyroid gland swelling, neurofibroma, perichondritis of the thyroid cartilage or subhyoid bursitis can also result in neck swelling. Recently, a congenital laterocervical cystic mass in a neonate was diagnosed as choristoma. Choristoma is a tissue or mass with a normal histology at an abnormal location [4].

Thyroglossal duct cysts are almost always midline in the neck and inferior to the hyoid bone. They usually move upward with tongue protrusion or swallowing. Most branchial cleft cysts occur anterior to the middle one third of the sternocleidomastoid muscle. Thyroglossal duct cysts and branchial cleft cysts may also present for the first time as infected neck masses. Hemangiomas are usually not present at birth but appear in early infancy and may enlarge rapidly. In most cases, they recede spontaneously by 5 to 6 years of age. They are usually much smaller

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than cystic hygromas, do not transilluminate, and may be recognized by their reddish colour (capillary or strawberry hemangioma) or by a bluish hue of the overlying skin (cavernous hemangioma).

Although not pertinent to the present case, half a century back a “rule of 7” was proposed, in which the average duration of symptoms for cervical masses caused by infections was 7 days, for neoplasms 7 months and an interval of 7 years was characteristic of developmental anomalies. This rule of 7 was quoted in a very interesting study carried over a 10 year period published three decades back [5].

How can one confirm the diagnosis?

The clinical features are virtually diagnostic in a case of cystic hygroma. However, sonography of the lesion or alternatively CT scan or MRI can delineate the structure very nicely, though it may not be cost-effective. In the index case, the diagnosis was confirmed by CT scan and histopathology. On ultrasound the lesions appear as multilocular cystic masses, containing septa of variable thickness; the fluid inside can appear completely anechoic or hypo to hyperechoic because of infections, haemorrhage or high lipid content [6]. On CT scan, cystic hygroma appears as a multiloculated cystic lesion composed of hypodense formations separated by septa with an increased density.

What are the modalities to diagnose this entity during the intrauterine period?

Cystic hygroma diagnosed during the intrauterine period, especially during the first trimester carries poor prognosis. Sonographic evaluation of fetal nuchal translucency thickness in the first trimester is crucial [7]. Sonographic evaluation of the foetus along with MR imaging can be useful not only for the diagnosis but also for planning management of cystic hygroma before or at the time of delivery. In fact, MRI can improve the delineation of the neck mass and can reveal compression of the airways [8].

Are there any modalities existing for intrauterine management?

EXIT procedure (EX utero Intrapartum treatment) encompasses a multidisciplinary approach in situations where airway obstruction is anticipated and has been used in foetuses with large neck

masses [9]. The prenatal/intrauterine use of single intralesional injection of OK-432, a sclerosing agent, has shown good results- a decrease in volume and no feeding or respiratory complications in the neonates [10].

What are the management options for this neonate?

Surgical excision can be performed and is usually the treatment of choice; occasionally instillation of a sclerosing agent can be a viable alternative. Successful excision of the cyst was performed in this case.

Key Points

- Cystic hygroma is a congenital multiloculated lymphatic swelling, partially compressible, brilliantly transilluminant and commonest location is in the posterior triangle of the neck followed by axilla.
- Other causes of neck swelling include teratoma, haemangioma, lipoma, branchial cleft cyst, thyroglossal cyst, ranula, thyroid gland enlargement, lymphadenopathy and infections.
- Sonographic evaluation aided by MRI can be a handy tool antenatally. CT scan can also be used for purposes of diagnosis and planning management after birth.
- Management in the neonatal period is surgical excision, although intralesional injection of sclerosing agents can also be effective.

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