

A newborn female with a smooth mass in the region of the base of the tongue

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A newborn female born to an Indian mother by lower segment cesarean section at full term was transferred to the neonatal ICU at Salmaniya Medical Complex, Bahrain, after developing feeding difficulty. During feeding, the attending nurse noted that the baby had a localized smooth mass of about 2 centimeters in the region of the base of the tongue on the left side. Her physical examination revealed no remarkable findings in the neck. The newborn was immediately referred to us for an ultrasound examination of her neck (Figure 1), followed by a CT scan of the neck as a part of further work-up (Figure 2).

- What are the ultrasound and CT scan findings?
- What would be the likely differential diagnosis based on ultrasound and CT scan findings?
- How is the diagnosis confirmed?

(see answer on page 180)

Figure 1. High resolution ultrasound demonstrating a well-defined cystic lesion in the hypopharyngeal region, mostly on the left side.



Figure 2. CT scan of the neck showing a cystic structure abutting the left side of the base of the tongue, contiguous with the vallecula and ipsilateral tonsillar fossa.

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Diagnosis: Vallecular Cyst

Radiological findings

Radiologic studies (Figures 1 and 2) showed a well-defined cystic structure measuring 1.47 x 1.64 centimeters in size in the supraglottic region on the left side abutting the base of the tongue close to the vallecula or ipsilateral fossa. Differentials based on ultrasound and CT scan findings include vallecular cyst, thyroglossal duct cyst, dermoid cyst, haemangioma, lymphangioma, cystic hygroma, ectopic thyroid tissue, and teratoma.

Direct laryngoscopy is regarded as the gold standard for confirming the diagnosis.

Discussion

Vallecular cyst is rare in newborn and infants. The earliest report of this condition dates back to 1881 by Abercrombie.¹ Infants present with sudden airway obstruction, which is observed immediately after birth or during the first week of life. The mode of presentation depends upon the position and size

of the cyst. Inspiratory stridor is the most frequent clinical presentation.² Other presentations include coughing, feeding difficulty, periodic spells of apnea, and cyanosis.³ Early recognition of this condition and differentiation from other causes of airway obstruction is an important step in management to prevent death from this cause.

Vallecular cysts have been reported under different names in the literature, e.g., mucus retention cyst, epiglottic cyst, base-of-the tongue cyst and recently, ductal cyst. The latter came from DeSanto et al's classification of laryngeal cyst, in which vallecular cyst is considered a subtype of laryngeal cyst. According to their review, there are two major categories of laryngeal cyst: saccular and ductal.⁴ Saccular cysts are supraglottic lesions and occur in the saccule of the ventricle. They are thought to result from cystic distention of the saccule and hence are more likely to cause airway obstruction. Alternatively, ductal cysts are mostly situated in the

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Figure 2. CT scan of the neck showing a cystic structure abutting the left side of the tongue, contiguous with the vallecula and ipsilateral tonsillar fossa.

vallecula and cause feeding problems before obstructing the airways. They are also known as mucus retention cysts and are thought to originate from obstructed submucosal glands.^{4,5}

Diagnosis of vallecular cyst requires a high index of clinical suspicion. As a part of the initial work-up, the diagnosis can be suggested by a lateral neck radiograph.^{4,6,7} Ultrasound helps in distinguishing the cystic nature of the lesion while CT scan is helpful in delineating the location and its extent in relation to the base of the tongue, vallecula and thyroid gland.⁸ Preoperative visualization of the thyroid gland is important to avoid inadvertent excision of the gland and to prevent hypothyroidism. The CT scan, however, has limited potential in narrowing the differential diagnosis. Definite diagnosis therefore, rests only on flexible direct laryngoscopy (as in our case). Based on ultrasound and CT characteristics, a list of possible differential diagnoses was suggested, which included vallecular cyst, thyroglossal duct cyst, dermoid cyst, haemangioma, lymphangioma, cystic hygroma, lingual thyroid tissue and teratoma. All show a similar low-density appearance on CT while lingual (ectopic) thyroid tissue and teratoma

show a high attenuation value and are the only entities distinguishable by CT.^{8,9} MRI could have helped in narrowing the differential diagnosis, but it could not be performed in our case due to the worsening condition of the infant. The infant, therefore, underwent direct laryngoscopy under general anaesthesia as a preoperative assessment of the cyst followed by therapeutic marsupialization. Direct laryngoscopy is, therefore, regarded as the gold standard examination in diagnosis of vallecular cyst.^{5,9}

Treatment of vallecular cyst is by transoral endoscopic surgical removal where the cyst can be marsupialized, excised or de-roofed. Endoscopic deroofing is technically simple and has an effective cure without recurrence.^{6,7,9} Incision and drainage alone is considered inadequate and has a high likelihood of recurrence, while wide cyst marsupialization is safe and ensures a low recurrence in particular when performed by CO₂ laser. Carbon dioxide laser is reportedly useful as it can vaporize the epithelial lining to prevent recurrence.^{2,10,11} Cyst removal by open surgical resection with external laryngofissure via lateral cervical approach has also been described in literature.¹¹

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