Rare disease

Cervical ectopic thymic cyst: a rare preoperative diagnosis

Anupama Tandon,¹ Rajesh Tandon,² M Chandrashekhar,² Poonam Das,³ Bhavna Bansal,² Nalin Bhatia⁴¹Radiology and Imaging Department, UCMS & GTB Hospital, Delhi, India;²Histopathology Department, Max superspecialty hospital, New Delhi, India;³Lab Medicine Department, Max superspecialty Hospital, New Delhi, India;⁴Pathology Department, Max superspecialty Hospital, New Delhi, India**Correspondence to** Dr Rajesh Tandon, rajesh.tandon@hotmail.com**Summary**

We present a case of a 5-year-old child with a painless swelling on the neck. Imaging revealed a multilocular cystic lesion lying in close proximity to carotid vessels. Based on imaging a possibility of ectopic thymic cyst was considered, which was confirmed on histopathology. Ectopic cervical cyst is an infrequent cause of neck cyst in a child which is rarely diagnosed preoperatively.

BACKGROUND

Cervical ectopic thymic cyst is a rare entity with only about 100 reported cases¹. Due to its rarity it is usually not considered in the differential diagnosis of cystic neck mass and is often mistaken for branchial cleft cyst or cystic hygroma, which are much commoner. It is invariably diagnosed on histopathology and a preoperative diagnosis is seldom made.

However, making a preoperative diagnosis is vital as an inadvertent excision of thymic cyst can result in immune-deficiency if it is the only functioning thymic tissue with absent mediastinal thymus. Imaging aids in making a correct diagnosis and also in depicting the location and extent of the lesion².

We present the case of a 5-year-old boy with cervical ectopic thymic cyst where the diagnosis was suspected on (CT) and confirmed on histopathology.

CASE PRESENTATION

A 5-year-old boy presented to the paediatrics outpatient department with a painless left sided neck swelling of 6 months duration. The mass was asymptomatic and noticed by parents incidentally. The family and medical history was unremarkable. Physical examination revealed a non-tender, soft, mobile and poorly circumscribed neck mass of about 4x5 cms size. No overlying skin changes were seen. The child's neck position and mobility were unaffected by the mass. Clinical differentials of branchial cleft cyst, cystic hygroma and hemangioma were considered and a sonogram was requisitioned.

INVESTIGATIONS

Sonography revealed a multilocular cystic mass with absence of colour flow on doppler imaging. Contrast enhanced CT scan of the neck demonstrated a thin-walled fluid density lesion beneath the left sternocleidomastoid muscle lateral to the carotid sheath (figure 1). The cyst was seen to extend medially behind the carotid vessels up to the pyriform sinus and inferiorly below the thyroid cartilage as far as the upper mediastinum. The cyst was

essentially non-enhancing except for mild enhancement of the wall.

DIFFERENTIAL DIAGNOSIS

Branchial cleft cyst, cystic hygroma, dermoid/epidermoid and ectopic thymic cyst were considered in the differential diagnosis. The close proximity to the carotid sheath, medial extension up to pyriform sinus and inferior extension up to the upper mediastinum favoured the diagnosis of ectopic thymic cyst. The presence of the normal thymus in the anterior mediastinum was then ascertained (figure 2).

TREATMENT

Patient underwent surgical excision of the lesion. Intraoperatively a thin-walled cyst was found abutting the carotid sheath. In this case a blunt dissection technique was employed to conserve the integrity of external capsule of the cyst and the surrounding major vessels.

OUTCOME AND FOLLOW-UP

Histological sections revealed multiloculated cyst lined by bland squamoid epithelial lining. Ectopic thymic tissue with lobulated architecture and distinct cortex and medullary compartment was present in the cyst wall (figure 3). The cyst epithelial lining strongly expressed Pan cytokeratin (CK) and CK 5/6 immunopositivity. A final diagnosis of ectopic cervical thymus with multilocular cyst was made.

Patient had an uneventful postoperative period without neurological defects or injuries and he was apparently well at 6 month follow-up.

DISCUSSION

Defective pathways of embryological descent of thymic primordia lead to a clinical spectrum of thymic anomalies. Thymus is a paired organ that develops from ventral sacculus of third and fourth branchial pouches during sixth week of foetal life. Proliferation of endodermal cells gives rise to paired solid masses which later fuse in midline by seventh week and descend to its location in superior mediastinum by eight week. Cervical thymic cyst presents as a cystic

Figure 1 a) Axial CECT image shows the cyst (arrow) extending posterior to carotid artery and terminating at the pyriform sinus. b) Coronal CT image showing extension of the cyst below the thyroid gland up to upper mediastinum (black arrow).

Figure 2 CT section through the superior mediastinum showing the normal thymus (T).

mass on lateral aspect of neck, deep to sternocleidomastoid muscle. It can occur anywhere along the line of descent that is between pyriform recesses and the mediastinum.

Majority of them are seen in infants or young children less than 10 years of age³. A slight higher incidence on the left side has been seen.³ They vary in size from 1–17cm and most are multilocular with smooth inner lining. Some authors report an equal sex distribution while others detected a 2:1 male predominance. Majority of the lesions present as asymptomatic slowly growing lateral neck masses.

The clinical features are non-specific and indistinguishable from other causes of cystic neck masses such as thyroglossal duct cyst, branchial cleft cyst, lymphangioma, dermoid/epidermoid cysts and cystic nerve tumours. Necrotic lymphadenopathy and abscess may be difficult to distinguish from a cyst particularly if it has been previously infected.

Imaging contributes by defining the nature and extent of mass. Sonography shows an anechoic multilocular cyst with thin septations on lateral aspect of neck. A CT scan better defines the character and relations of the cyst.

Figure 3 Photomicrograph (10X) shows lobulated thymic tissue with distinct cortex and medulla in the cyst wall.

Cranially the ectopic thymic cyst passes posteromedial to the carotid vessels and ends near the pyriform sinus while caudally it often extends below the thyroid, sometimes as far as the mediastinum². On MRI, it is seen as T2 hyperintense lesion situated between carotid artery and internal jugular vein, MRI better delineates the thin band like extension upto the mediastinum. Additionally imaging helps to ascertain the presence/absence of the normal mediastinal thymus.

The differential diagnoses of cystic neck mass include second branchial cleft cyst, cystic hygroma, lymphangioma, thyroglossal cyst, ectopic thymic cyst, vallecular cyst, dermoid, epidermoid, necrotic lymphadenopathy and cystic nerve tumours and cystic neuroblastoma.

The necrotic adenopathy and cystic nerve tumour/neuroblastoma have thick walls on sonography. A thin-walled cystic mass as in the presented case can be due to branchial cleft cyst, lymphangioma, thyroglossal cyst and ectopic thymic cyst. While thyroglossal cyst is more in midline the other three are lateral as in our case. Imaging appearances of these three can be indistinguishable except for some subtle pointers. While branchial cleft cyst passes between the carotid fork to terminate at the base of tonsils, thymic cyst passes behind the carotid artery to end at the pyriform sinus. In addition, thymic cyst extends more caudally, sometimes up to the mediastinum which is never seen with branchial cyst². A preoperative cytological examination from the cyst wall can further facilitate a confident diagnosis.

Though most cases remain asymptomatic, possible complications include myasthenia gravis associated with

ectopic thymic tissue or rarely development a malignant thymoma. Surgical excision is the treatment of choice and presence of normal thymic tissue in the cyst wall on histopathology confirms the diagnosis.

Learning points

- Ectopic cervical thymic cyst is a rare cause of cystic neck swelling in a child
- Occasionally, this may be the only functioning thymic tissue with absent mediastinal thymus
- Imaging and cytology aids in making a preoperative diagnosis and in establishing the presence of normal mediastinal thymus
- Since, its inadvertent removal may result in immune-incompetence, it should be included in differentials of a cystic neck mass in a child despite its rarity.

Competing interests None.

Patient consent Obtained.

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