Rare disease

A rare congenital neck lump

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Summary

We describe the case and present a radiological image of a neck lump identified antenatally with ultrasound imaging. Postnatally a left-sided asymptomatic neck lump was identified in the left posterior triangle of the neck. Repeat ultrasound and CT imaging were arranged confirming a cyst adjacent to the cervical oesophagus and displacing the carotid sheath anteriorly. Complete cyst excision was achieved with no complications. Histological analysis identified a 30×22×20 mm cyst with a smooth muscle layer within the cyst wall and a lining of respiratory epithelium. These findings were consistent with a diagnosis of cervical duplication cyst (CDC). Proximity to the carotid sheath and oesophagus can make CDC excision potentially dangerous hence preoperative CT scanning was useful to establish the anatomical relations of the cyst in this case.

BACKGROUND

Cervical duplication cysts (CDC) are a rare but potentially dangerous pathology due to airway compression and close relations to vital anatomical structures. Understanding the pathological nature of duplication cysts is important to avoid surgical complications. In the cervical region iatrogenic adjacent structure injury is an added concern.

CASE PRESENTATION

Baby S (male) was born at 37 weeks of gestation weighing 3077 g. Antenatal ultrasound scanning at 20 weeks gestation had suggested a mass in the cervical region and hence repeat ultrasound was arranged in the early postnatal period. On clinical assessment Baby S was asymptomatic with no respiratory compromise or torticollis. There was no significant family history. Examination revealed a 10×15 mm soft immobile oval lump palpable in the left posterior triangle of the neck deep to the skin.

INVESTIGATIONS

Postnatal ultrasound was performed at 6 weeks of age and confirmed the presence of a fluid-filled cyst located in the left neck with close association with the cervical oesophagus and carotid sheath. In view of the potential risks of iatrogenic injury to the related structures at surgical excision, a CT scan was requested to clarify these anatomical associations (figure 1). CT scan revealed a cyst measuring 10×16×35 mm directly posterior to the carotid sheath and displacing these vessels anteriorly. The cervical oesophagus abutted the medial aspect of the cyst. The inferior pole of the cyst extended to the level of the upper mediastinum and the superior pole was at the level of the hyoid bone.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses of lateral neck swellings in neonates include lymphadenopathy, lymphoma, branchial cyst, cystic hygroma, stenomastoid tumour, haemangioma, submandibular or parotid gland tumours, neoplasms and

duplication cysts. In this case, the duration of findings indicates a congenital pathology and the proximity of the lesion to the cervical oesophagus is suggestive of CDC.

TREATMENT

Surgical excision was performed at 7 weeks of age under general anaesthesia. A transverse incision was made over the lesion with respect to Langer's lines and a 30×22×20 mm cyst was completely excised from the posterior aspect of the carotid sheath. The medial aspect of the cyst was adherent to the cervical oesophagus but no fistula was demonstrated.

OUTCOME AND FOLLOW-UP

Postoperative recovery was routine and there were no complications. Histological analysis of the specimen confirmed a smooth muscle wall lining to the cyst and a lining of respiratory epithelium. Ultrasound and radiological imaging

Figure 1 Axial CT image showing a cervical duplication cyst in the left neck (blue arrow) adjacent to the oesophagus (green arrow) and displacing the carotid sheath anteriorly (yellow arrow).

confirmed that there was no associated urinary tract or vertebral abnormalities.

DISCUSSION

CDCs are a rare cause of congenital neck lumps. Diagnosis may be suspected antenatally as in this example, however, this is unusual. Respiratory compromise is the most commonly associated symptom and has occurred before a clinically identifiable mass is identified.¹ Feeding difficulties and torticollis have also been described as associated symptoms.

Associated anomalies can occur and consideration should be taken for further imaging to exclude vertebral and urinary tract abnormalities.²⁻³ The histological criteria to confirm the diagnosis of CDC are cyst attachment to the cervical oesophagus, a smooth muscle layer within the cyst wall and cyst contents of squamous epithelium or a lining of the embryonic oesophagus.

Previous reports have identified pus and positive bacterial cultures from the cyst contents in both the presence and absence of gastrointestinal communications. In view of this, cyst excision can be considered a clean-contaminated procedure in the absence of clinical infection hence preoperative antibiotics to decrease the risk of surgical site infection may be prudent.⁴

Two published reports of incomplete cyst excision and oesophageal breach were complicated with wound infection and persistent sinus.⁵⁻⁶ Complete cyst excision is likely to avoid wound-related problems though clearly iatrogenic injury to the oesophagus should be avoided. There is little evidence to support any specific operative management strategies of clinically identified oesophago-cystic fistula; however, two published reports employed simple ligation of fistulae no postoperative complications.⁷⁻⁸

To facilitate complete cyst excision several reports specifically highlight the advantages of placing a transoral bougie or dilator to better localise and identify the cervical oesophagus.³⁻⁴⁻⁹ In the presented case this was not necessary as the oesophagus was clearly identified and preserved.

Learning points

- Preoperative CT scan may be useful to clarify anatomical relations to the cyst and better define the full extent of the cyst and plan excision.
- The use of preoperative antibiotics should be considered as gastrointestinal fistulae may be present.
- Complete cyst excision may be associated with fewer complications.
- Placement of a transoral oesophageal dilator or bougie has been previously described to facilitate complete cyst excision and help identify the cervical oesophagus intraoperatively to avoid iatrogenic injury.

Competing interests None.

Patient consent Obtained.

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