

Ectopic cervical thymus: case report and review of pathogenesis and management

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Abstract

Objective: Ectopic cervical thymus is a rare anomaly, and 50 per cent of cases occur in children. The aim of this report was to remind clinicians of this rare condition, which is part of the differential diagnosis of a paediatric neck lump.

Method: Case report together with literature review of the pathogenesis and management of ectopic cervical thymus.

Results: The ectopic cervical thymus is a rare cause of a benign neck mass, and is thus usually misdiagnosed as lymphadenopathy or neoplasia. We discuss its embryology, aetiology, presentation, histopathology, radiology and management, with reference to the reported case.

Conclusion: Ectopic cervical thymus should always be considered in the differential diagnosis of a paediatric neck lump. The diagnosis can often be confirmed by cytology and radiology prior to surgical excision.

Key words: Thymus Gland; Neck; Child

Introduction

Ectopic cervical thymus is a rare condition, and 50 per cent of cases occur in children.¹ Ectopic thymic tissue can occur anywhere between the angle of the mandible and the upper mediastinum, along the line of embryological descent. The ectopic cervical thymus is a rare cause for a neck mass, and is thus usually misdiagnosed as lymphadenopathy or neoplasia.² The correct diagnosis is usually made after surgical excision of the mass.

Case report

A seven-month-old boy presented to our department with a right-sided upper neck mass. The mass had first been noticed when the child was six weeks old, and there had been no change in size or shape since then. The neck mass was otherwise asymptomatic, and did not cause any breathing or feeding difficulties. The child was developing normally.

Clinical examination revealed a soft, ill defined mass in the right upper neck, measuring approximately 5 cm in maximal diameter. Intra-oral examination was normal.

An ultrasound of the child's neck demonstrated a well defined, non-cystic swelling. Magnetic resonance imaging (MRI) demonstrated a solid, homogeneous mass in the right upper neck, extending deeply into the carotid space and with some extension into the parapharyngeal space. Radiologically, the mass was separate from the submandibular and parotid glands and the sternomastoid muscle (Figures 1 and 2).

Fine needle aspiration cytology (FNAC) was performed; results suggested a benign lipoblastoma. (The important differential diagnosis for lipoblastoma is liposarcoma,

a malignant tumour which is fortunately exceedingly rare. Lipoblastomas slowly expand with time, and the treatment of choice is surgical excision.)

The mass was surgically excised. Intra-operatively, a soft, tan-coloured, lobulated tumour was identified deep to the sternomastoid muscle, which extended medially around the structures of the carotid sheath, as well as superiorly, deep to the posterior belly of the digastric muscle, around the hypoglossal and accessory nerves. The mass was removed completely, and measured approximately 6 × 2 cm.

The post-operative course was uneventful, and the patient was discharged home after 48 hours.

Histologically, the excised mass had the appearance of an ectopic thymus (Figures 3 and 4), rather than the lipoblastoma suggested on FNAC. Once the correct diagnosis had been established, the MRI scans were reviewed, and they confirmed the presence of a normal thymus in the anterior mediastinum.

The child was reviewed after six weeks and was doing well. The neck wound had healed well. The patient was discharged back to the care of his general practitioner.

Ectopic cervical thymus

Embryology and aetiology

The thymus gland starts to develop during the fourth to fifth week of gestation.³ It develops mainly from the ventral wing of the third pharyngeal pouch on either side. Bilateral thymic primordia appear at the beginning of the sixth week of gestation, and by the end of this week both thymic primordia separate from their pharyngeal pouches and migrate caudally and medially. At eight weeks of gestation, the bilateral thymic primordia fuse in the midline

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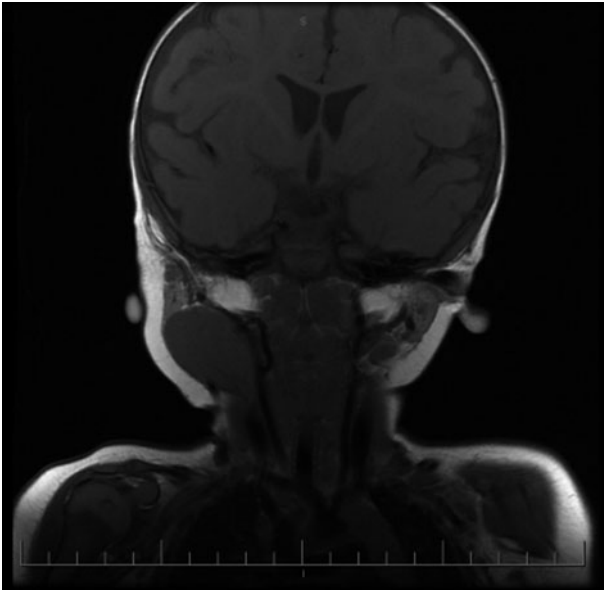


FIG. 1

Coronal, T1-weighted magnetic resonance imaging scan demonstrating a homogeneous lesion in the right carotid and parapharyngeal space.

and start to descend down under the sternum into the superior mediastinum. At the end of this descent, the upper end of the thymus is resorbed. The final position of the thymus is usually in the anterior mediastinum between the sternum, the parietal pericardium and the thoracic inlet.⁴

The aetiology and pathogenesis of ectopic cervical thymus include the following factors: (1) implantation of thymic tissue in the neck via a process of sequestration from the gland during descent; (2) persistence of a small remnant of thymic tissue left behind after descent, as the upper ends of the gland involute within the thymopharyngeal duct; and (3) incomplete or failed descent, with the thymus glands remaining in the cervical region.⁴⁻⁶

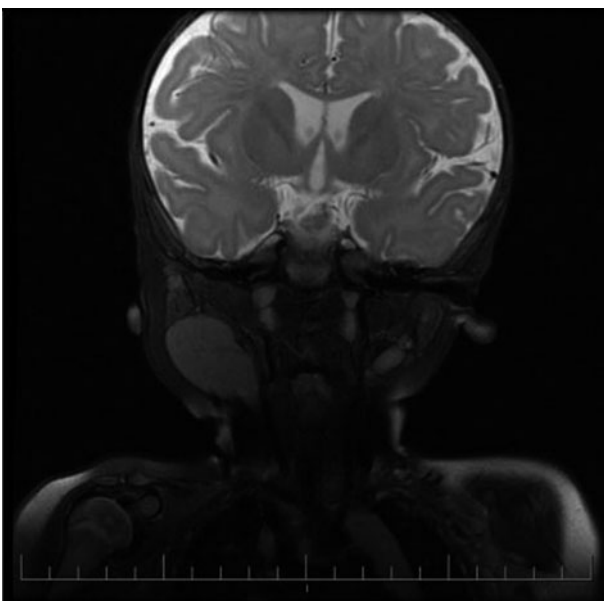


FIG. 2

MRI-Coronal T2 weighted image.

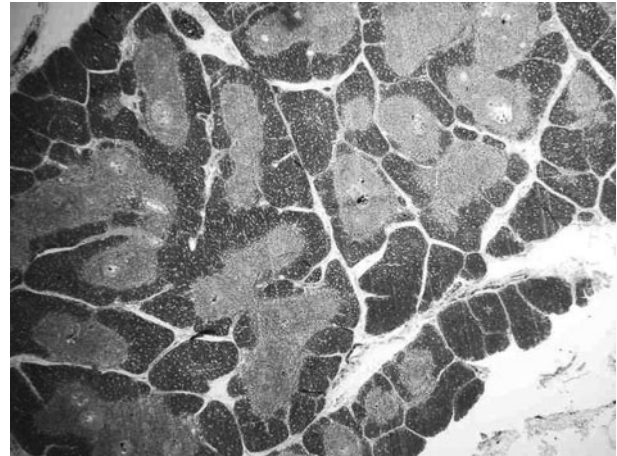


FIG. 3

Photomicrograph showing lobulated thymic tissue, with a distinct, dense outer cortex and pale-staining medulla (H&E; $\times 20$).

Presentation

Ectopic cervical thymic masses are congenital lesions which usually present as asymptomatic neck swellings.⁷ They can be either solid or cystic in nature. Large ectopic cervical thymic masses can occasionally present with hoarseness, stridor or dysphagia due to compression and displacement of neighbouring structures.⁸ Ectopic cervical thymic swellings are commonly reported on the left side; they are usually unilateral and are commoner in males.^{9,10} The differential diagnosis includes branchial cyst, cystic hygroma, cystic teratoma, thyroglossal duct cyst, thyroid or parathyroid lesions, lymphoproliferative disorders, reactive adenopathy, vascular tumours, and lipoblastoma.⁷

Histopathology

Ectopic cervical thymic cysts can be diagnosed by FNAC performed by an experienced pathologist. Tunkel *et al.* have shown that ectopic cervical thymic tissue can be reliably identified by FNAC, using morphological and flow cytometric evaluation of cytological material.⁷ Papanicolaou and Romanowsky stained smears of fine needle aspirate and demonstrated a dominant population of

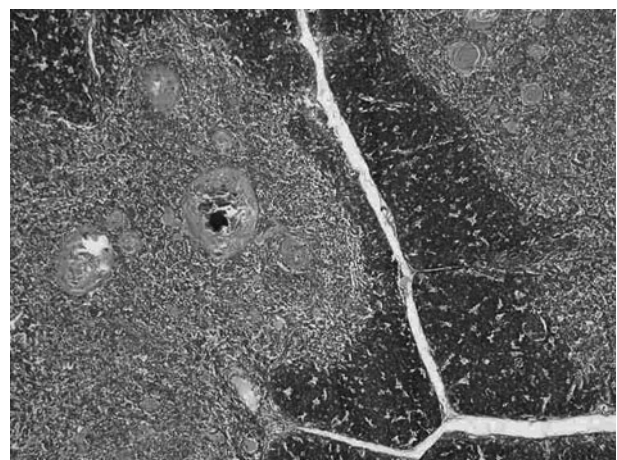


FIG. 4

Photomicrograph showing several concentrically keratinised Hassall's corpuscles upon a background of small lymphocytes (H&E; $\times 100$).

small, round, nonactivated lymphocytes with variable numbers of epithelial cells scattered in the background. The lack of overt malignant features, combined with an immature T-cell phenotype, favours thymic tissue on flow cytometry. The presence of fibrous septa, Hassall corpuscles and other characteristics of normal thymic architecture confirm the diagnosis of ectopic thymus.⁷

Histological examination of ectopic thymic tissue reveals concentrated lymphocytes at the cortex surrounded by sparser modularly lymphocyte. High power examination reveals monocytes surrounding eosinophilic breakdown products of lympholysis, forming the concentric thymic corpuscles of Hassall.^{11,12} Hassall corpuscles are diagnostic for thymic tissue.

Radiology

The radiological investigations used in the management of ectopic cervical thymus include ultrasonography, computed tomography (CT) and MRI scanning.

Advanced, high-resolution ultrasonography is capable of demonstrating the intrathymic anatomy, and it is thus possible to diagnose ectopic thymic tissue using this imaging modality.¹³ The ultrasonic appearance of the thymus is unique: septae are seen as echogenic, linear structures, the cortex appears hypoechoic and the medulla echogenic. The vessels between the septae are recognised as discrete, high level echoes.^{14,15}

- **The thymus plays an important role in the development of the immune system, especially in infants**
- **Ectopic thymus is a rare condition**
- **Ectopic cervical thymus should always be considered in the differential diagnosis of a paediatric neck lump**
- **Diagnosis can often be confirmed pre-operatively**
- **The presence or absence of a normal mediastinal thymus should be confirmed before surgical resection of an ectopic thymus**

Computed tomography and MRI are also excellent imaging modalities for the diagnosis of ectopic cervical thymus. The ectopic thymus appears homogeneous, iso- or slightly hyperintense compared with muscle on T1-weighted images, and hyperintense on T2-weighted images. Magnetic resonance imaging can also be used to visualise the tract between the ectopic and mediastinal thymus; identification of this tract makes the diagnosis of an ectopic thymus more likely.^{16–18}

Management

The natural history of the ectopic cervical thymus is unknown. Almost all cases reported in the literature have been surgically removed. The probable reason for this is difficulty in diagnosis and the small risk of malignancy.^{19,20} It is very important to identify the presence of the normal mediastinal thymus prior to removal of an ectopic cervical thymus, especially in children younger than one year of age, as in this age group removal of a sole thymus results in decreased numbers of T cell subsets.^{21–23} Therefore, normal thymic tissue should be preserved if possible when treating patients with thymic anomalies. In cases in which no normal thymus is identified pre-operatively, intra-operative frozen section analysis should be performed to confirm the diagnosis and to rule out

malignancy, with the aim of thymus preservation if at all possible (to prevent immunodeficiency).

Conclusion

An ectopic cervical thymus should always be considered in the differential diagnosis of a paediatric neck mass. The diagnosis could be confirmed pre-operatively with FNAC, with or without ultrasound, CT or MRI scans. The presence or absence of a normal mediastinal thymus should be confirmed prior to surgical resection, especially in infants, as it plays an important role in the development of the immune system.

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