

Cervical ectopic thymus in an infant

Prasad T R S, Chui C H, Ong C L, Meenakshi A

ABSTRACT

Cervical ectopic thymus, a common embryological anomaly detected incidentally at autopsy, is rarely described in clinical patients. About 100 cases have been described in the literature, ten percent of which occurred in neonates. We report a case of solid cervical ectopic thymus in a three-month-old male infant presenting as a neck lump and snoring at sleep. The embryopathogenesis, clinical features, diagnostic modalities and management options are discussed, together with a review of the literature.

Keywords: aberrant thymus, ectopic thymus, heterotopic thymus, infant, neck mass

Singapore Med J 2006; 47(1):68-70

INTRODUCTION

Cervical ectopic thymus (CET) presenting as a neck lump is rarely considered in the differential diagnosis of neck swellings. Although it is a common anomaly detected incidentally at autopsy, clinical presentation in the form of a neck swelling either asymptomatic or with compressive symptoms on the surrounding structures has rarely been reported in the literature. Most of the cases of CET are not diagnosed preoperatively, as they are not usually considered due to its rarity. Preoperative diagnosis of the lesion with confirmation of normal mediastinal thymus is essential to avoid inadvertent total thymectomy and subsequent immunodeficiency, especially in infants and children.

CASE REPORT

A three-month-old male infant presented with history of right-sided neck swelling and snoring at sleep for a duration of three weeks. On examination, there was a 4x3cm firm, globular and non-transilluminant swelling in the right submandibular region anterior to the sternocleidomastoid muscle (SCM) and deep to the platysma. With a provisional diagnosis of a lymphangioma or a branchial cleft cyst, a computed

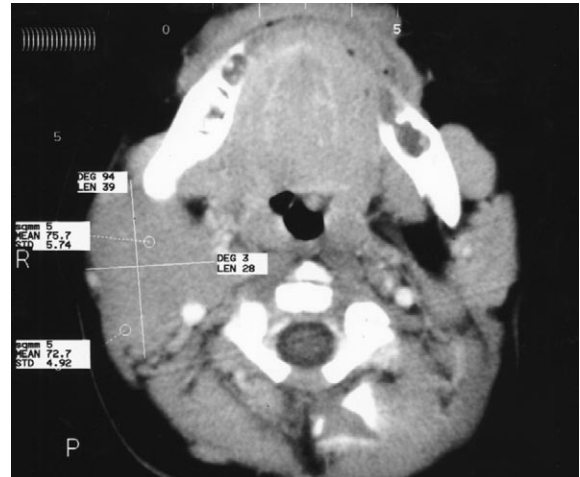


Fig. 1 Axial CT image of the neck shows an ectopic thymus in the right submandibular region.

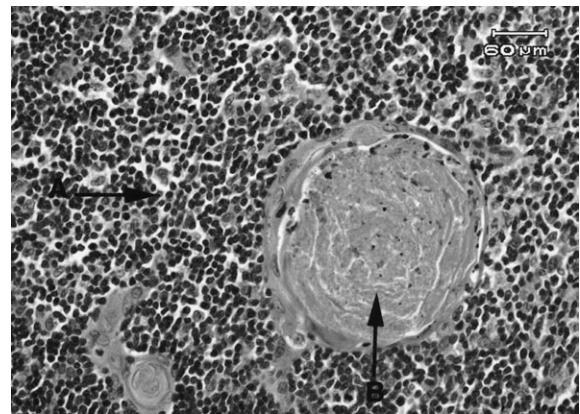


Fig. 2 Photomicrograph shows thymic lymphoid tissue (A) with Hassall's corpuscle (B) (Haematoxylin & eosin x40).

tomography (CT) of the neck was done. CT revealed a 3.9 x 2.8 x 2.4cm lobulated mass inferior to the right parotid gland and anterior to the right SCM with a homogenous attenuation suggestive of a soft tissue mass or cystic swelling with proteinaceous fluid (Fig. 1). A diagnosis of a benign soft tissue mass, cystic hygroma or a branchial cleft cyst was considered.

At surgical exploration, a firm lipomatoid mass of the above-mentioned dimensions was found closely

Department of
Paediatric Surgery
KK Women's and
Children's Hospital
100 Bukit Timah Road
Singapore 229899

Prasad T R S, MBBS,
MRCS, MCh
Senior Registrar

Chui C H, MBBS,
FRCS, FAMS
Consultant and Head

Department of
Diagnostic Imaging
KK Women's and
Children's Hospital

Ong C L, MBBS,
FRCR, FAMS
Senior Consultant

Department of
Histopathology
KK Women's and
Children's Hospital

Meenakshi A,
MBBS, MD
Clinical Associate

Correspondence to:
Dr Sai Prasad TR
Tel: (65) 6394 1113
Fax: (65) 6291 0161
Email: tsai@
rediffmail.com

adherent to the right carotid sheath and infiltrating towards the retropharyngeal space. The mass was excised easily by blunt dissection. The diagnosis of solid ectopic thymus was made on histopathological examination (Fig. 2). The child had an uneventful postoperative recovery with relief from snoring and is doing well two years after the surgery.

DISCUSSION

CET is an uncommon cause of a neck lump and is usually described in sporadic case reports. Tovi and Mares reviewed 68 reported cases of ectopic thymus (ET) in 1978⁽¹⁾. Ten years later, in a collective review of 91 cases by Nowak et al, 76 presented as neck masses while the rest were mediastinal in location⁽²⁾. Slightly more than 100 cases of CET have been reported in the world literature^(3,4,8). The majority of cases are seen between two and 15 years of age, with a male preponderance^(3,4). Only 10% of the patients are infants⁽⁴⁾.

Defective pathways of embryological descent of thymic primordia lead to a clinical spectrum of thymic anomalies. The thymus is a paired organ developed from the ventral sacculles of the third and occasionally fourth pharyngeal pouches during the sixth week of foetal life. The thymopharyngeal tract elongates and descends into the superior mediastinum. The bilobed thymus develops by the third embryonic month⁽⁵⁾. The human thymus exhibit age-related variation with respect to the size and weight. The thymus attains its largest relative size at the age of three years, and continues to grow to attain its largest weight of 30-40 grammes by puberty. Later, it involutes to approximately 15 grammes in the adult^(1,6). The thymus, including ET, exhibits hyperplasia during the first decade of life or following infection or vaccination⁽⁴⁾. The thymus has a significant physiological role, especially in infants and children, in the immune mechanisms of the body and prevention of autoimmune diseases.

Ectopic thymic masses are located along the pathway of descent of the thymus. Hence, it could be sited anywhere from the angle of mouth or base of the skull to the superior mediastinum. These thymic vestiges are relatively common anomalies but since they are asymptomatic, they are detected occasionally. An incidence of 21%-30% is noted at autopsy⁽⁷⁾. Few authors believe that the ET is so common that it should be considered as a normal variation.

CET is often asymptomatic, with only 10% of patients being symptomatic in the form of pain, upper respiratory tract infection and pressure symptoms like stridor, dyspnoea, dysphagia

and hoarseness of voice^(8,9). Snoring during sleep in association with CET has not been previously described. We postulate that in our patient, compression over the oropharynx caused snoring and was relieved following excision. Acute presentation as a result of life-threatening tracheal compression due to intralesional bleed or infection is rare.

CET presents as aberrant solid thymic tissue, thymic cyst or cervical thymoma⁽³⁾. Solid CET constitutes about 10% of all the ectopic thymic masses. The pathogenesis of solid cervical thymic tissue is a result of arrest in its normal descent, failure of involution or sequestration of thymic tissue during descent⁽⁸⁾. Cervical thymic cysts result due to degeneration of Hassall's corpuscles or cystic change within the remnants of the thymopharyngeal duct. Thymic parenchyma, lymphoid tissue of thymic origin and Hassall's corpuscles found within the cyst wall are considered pathognomonic findings.

Although CET was described more than a century ago, they were rarely mentioned in the differential diagnosis of neck swellings. Lymph node swelling, branchial cleft cyst, cystic hygroma and lymphoma are the usual diagnoses entertained in children. Other causes of lateral neck swelling include sternomastoid tumour, lipoma, plunging ranula, dermoid cyst, thyroid and parathyroid cysts, eccentric thyroglossal cyst, cervical bronchogenic cyst, teratoma, neuroblastoma, and ET. We did not suspect CET due to its rarity in infants. Preoperative diagnosis of CET is rare and difficult, as no study has determined the optimal means of evaluating neck masses in children. Ultrasonography aids in differentiating solid and cystic neck lesions. Newer ultrasound techniques like Doppler and pulsed wave ultrasonography give a detailed description of the morphology and vasculature and are noninvasive and cost effective⁽¹⁰⁾. Advanced high-resolution ultrasonography demonstrates intrathymic anatomy, including connective-tissue septa and blood vessels within the septa⁽¹¹⁾.

CT can demonstrate thymic cysts and the CET is seen as a homogeneous mass with non-specific attenuation value of soft tissue. In our case, the mass had a solid appearance on the CT due to high Hounsfield units obtained. Since a similar appearance could be seen in complicated cysts with haemorrhagic fluid or proteinaceous fluid, the diagnosis of a benign soft tissue mass, cystic hygroma or a branchial cleft cyst was considered. On magnetic resonance (MR) imaging, the thymus is slightly more intense than muscle on T1-weighted images and isointense relative to fat on T2-weighted images. Cystic CET is

usually hypointense on T1- and hyperintense on T2-weighted MR images⁽¹⁰⁾. The imaging findings described are usually recognised retrospectively and hence a high index of suspicion is the key to the diagnosis. Fine-needle aspiration cytology (FNAC) usually clinches the diagnosis and should be considered in evaluating neck masses⁽⁸⁾. Other imaging modalities define the extent of the lesion including mediastinal extension, relation to surrounding structures and the presence of normal mediastinal thymus.

CET, although benign, should be excised as they may undergo malignant degeneration. Malignant degeneration of solid ET, squamous cell carcinoma in a thymic cyst, and thymoma in CET have been reported as anecdotal case reports^(3,4,8). Preoperative confirmation of normal thymus is considered essential before excision of CET as it may render the patient athymic in the absence of normal mediastinal thymus^(4,12). Although the long-term clinical consequences of thymectomy in early life are unclear, evidence of impairment of parameters of immunity has been found later in children who had routine thymectomy during cardiac surgery in early life⁽¹³⁾. At surgery, despite closely adherent to major vessels and nerves in the neck, the thymic tissue can be excised in toto with blunt dissection. Some authors believe that since the lesion is benign, a more conservative approach could be adopted without surgery. If there is any change in the characteristics of the lesion or there is questionable patient compliance, excision is advocated. In cases with life-threatening presentation due to tracheal compression with absence of mediastinal thymus, partial excision to relieve the obstruction is recommended so as to maintain normal immune function⁽⁸⁾. On the contrary, cervical thymic cysts can be excised without untoward effects as no active thymic tissue is found in thymic cysts⁽³⁾.

Although CET is a common embryological anomaly at autopsy, there are only few more than 100 clinical case reports of the same. Solid ET constitutes about of 10% of ET lesions. CET in infants is rare and constitutes about 10% of the total cases. High index of suspicion, pre-operative ultrasonography and FNAC usually aid diagnosis. Confirmation of normal mediastinal thymus is essential prior to surgical resection to prevent inadvertent total thymectomy.

REFERENCES

1. Tovi F, Mares AJ. The aberrant cervical thymus. Embryology, pathology, and clinical implications. *Am J Surg* 1978; 136:631-7.
2. Nowak PA, Zarbo RJ, Jacobs JR. Aberrant solid cervical thymus. *Ear Nose Throat J* 1988; 67:670-7.
3. Ozturk H, Karnak I, Deveci S, et al. Multilocular cervical thymic cyst: an unusual neck mass in children. *Int J Pediatr Otorhinolaryngol* 2001; 61:249-52.
4. Kacker A, April M, Markentel CB, Breuer F. Ectopic thymus presenting as a solid submandibular neck mass in an infant: case report and review of literature. *Int J Pediatr Otorhinolaryngol* 1999; 49:241-5.
5. Spigland N, Bensoussan AL, Blanchard H, Russo P. Aberrant cervical thymus in children: three case reports and review of the literature. *J Pediatr Surg* 1990; 25:1196-9.
6. Boyd J, Templer J, Havey A, et al. Persistent thymopharyngeal duct cyst. *Otolaryngol Head Neck Surg* 1993; 109:135-9.
7. Wagner CW, Vinocur CD, Weintraub WH, Golladay ES. Respiratory complications in cervical thymic cysts. *J Pediatr Surg* 1988; 23:657-60.
8. Millman B, Pransky S, Castillo J 3rd, Zipfel TE. Cervical thymic anomalies. *Int J Pediatr Otorhinolaryngol* 1999; 47:29-39.
9. Shah SS, Lai SY, Ruchelli E, et al. Retropharyngeal aberrant thymus. *Pediatrics* 2001; 108:E94.
10. Cacciaguerra S, Rizzo L, Tranchina MG, et al. Ultrasound features of ectopic cervical thymus in a child. *Pediatr Surg Int* 1998; 13:597-9.
11. Han BK, Yoon HK, Suh YL. Thymic ultrasound. II. Diagnosis of aberrant cervical thymus. *Pediatr Radiol* 2001; 31:480-7.
12. Saggese D, Ceroni Compadretti G, Cartaroni C. Cervical ectopic thymus: a case report and review of the literature. *Int J Pediatr Otorhinolaryngol* 2002; 66:77-80.
13. Brearley S, Gentle TA, Baynham MI, et al. Immunodeficiency following neonatal thymectomy in man. *Clin Exp Immunol* 1987; 70:322-7.