Malaysian J Pathol 2010; 32(1) : 65 – 68

CASE REPORT

Cervical ectopic thymus masquerading as metastatic thyroid papillary carcinoma

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Abstract

Cervical ectopic thymus (CET), a common embryological anomaly detected incidentally at autopsy, is rarely described in clinical patients. Furthermore, aberrant cervical thymic tissue is an infrequently reported cause of paediatric neck masses. We report a 12-year-old female presenting with multinodular thyroid swelling since seven years of age. FNAC revealed adenomatous goitre with suspicious cystic papillary neoplastic foci, for which she underwent total thyroidectomy along with excision of a nodular swelling near the lower pole of the right thyroid lobe which was per-operatively suspected to be a lymph node. Histopathological examination revealed a follicular variant papillary carcinoma of the thyroid with background thyroiditis, and a nodule of ectopic thymic tissue. Though the presence of CET is rare, one should be aware of this entity, especially in children because it may be confused with lymph node metastasis which may lead to morbid radical neck dissection.

Keywords: ectopic thymus, neck mass, thyroid, children

INTRODUCTION

Ectopic cervical thymic tissue is an uncommon cause of neck masses in children.[1] Although it is a common anomaly detected incidentally at autopsy, clinical presentation in the form of a neck swelling either asymptomatic or with compression symptoms on the surrounding structures has rarely been reported in the literature. Most cases of cervical ectopic thymus (CET) are not diagnosed preoperatively, as they are usually not considered due to its rarity.[2] Sporadic cases have been reported of ectopic thymic tissue formed along the path of embryologic descent from the mandibular region to the mediastinum, usually manifesting as an asymptomatic mass.[3] We report an instructive case which illustrates how CET may be mistaken for a lymph node metastasis in the setting of thyroid carcinoma.

CASE REPORT

A 12-year-old female presented with multinodular midline neck swelling since seven years of age. She was euthyroid. Examination revealed a multinodular thyroid enlargement, more prominent at the right side, which was moving with deglutition. No cervical swelling or lymph node could be palpated. Ultrasonogram revealed a multinodular goitre with cystic degeneration without any lymphadenopathy or neck mass apart from the thyroid swelling. Fine needle aspiration cytology revealed features of adenomatous goitre along with the presence of a few pseudopapillae and follicles of neoplastic cells displaying nuclear features of papillary carcinoma without the presence of true papillary fronds. The patient underwent total thyroidectomy along with excision of an identifiable nodule 1.5 × 1 cm adjacent to the right lower lobe, noted during surgery.

Pathology

Histopathological examination of the thyroid revealed a follicular variant of papillary carcinoma in the right lobe, evidenced by the presence of foci of round to oval mildly acidophilic to clear cells with pleomorphic optically clear orphan Annie vesicular nuclei (figure 1) without the presence of classical papillary fronds. There was background thyroiditis and a focal granulomatous lesion

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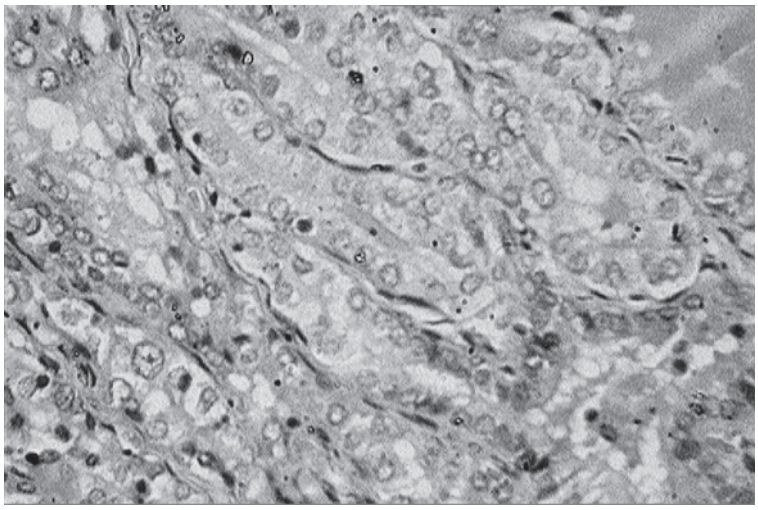

FIG. 1: Follicular variant of papillary carcinoma of thyroid showing optically clear nuclei with grooves. H&E X 400.

around the colloidal component simulating de Quervain's thyroiditis. An adherent parathyroid gland was also present.

The nodular swelling adjacent to the lower pole of the right thyroid lobe, thought to be a lymph node per-operatively, revealed lobular lymphoepithelial components (figure 2) containing Hassall's corpuscles (figure 3). No histological features of lymph nodal tissue was present. The epithelial elements and Hassall's corpuscles showed strong immunohistochemical staining for epithelial membrane antigen (EMA) and cytokeratin (CK) (figure 4). The dense population of lymphocytes displayed strong positivity for CD45RO (clone UCHL1) suggesting they were T-lymphocytes. These findings were in support of ectopic thymus rather than lymph node.

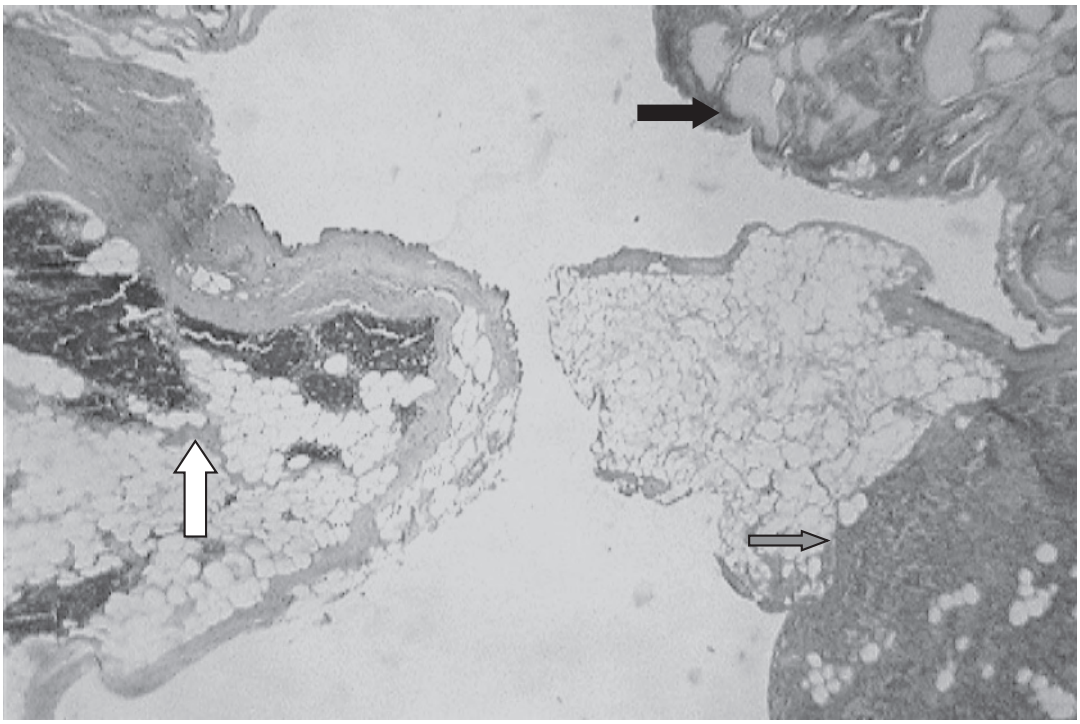

FIG. 2: Low power view shows closely associated thyroid (black arrow), parathyroid (gray arrow) and thymic tissue (white arrow) H&E X 100.

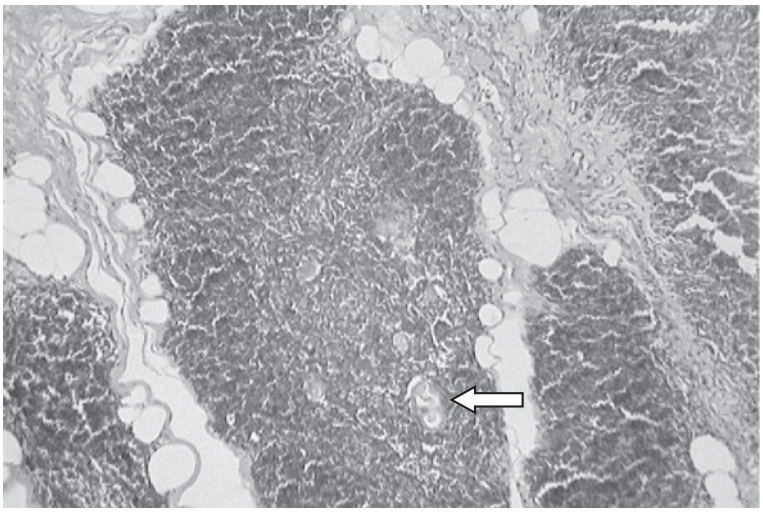

FIG. 3: Ectopic thymic tissue comprising lobular lymphoepithelial components with Hassall's corpuscles (white arrow) and closely associated adipocytes. H&E X 200

DISCUSSION

Cervical ectopic thymus (CET) presenting as a neck lump is rarely considered in the differential diagnosis of neck swellings. It is important to be aware of this entity to prevent anxiety on the part of the patient and inappropriate investigations and/or intervention on the part of the endocrine surgeon.[4]

The thymus is a paired organ developed from the ventral saccules 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 exhibits age-related variation with respect to 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 gm by puberty. Later, it involutes to approximately 15 gm in the adult.[6,7] Defective pathways of embryological descent of the primordial thymus can lead to a spectrum of thymic anomalies.

Ectopic thymic masses are located along the pathway of descent of the thymus. Hence, it could be sited anywhere from the angle of mandible or base of the skull to the superior mediastinum.[8] Among 3236 paediatric necropsies studied over 23 years by Bale and colleagues,[9] abnormal positioning of thymic tissue was recorded in 34 cases. The maldescended and ectopic tissue were localized near the thyroid gland or lower in the neck and at more cranial locations, including seven cases found medial to the submandibular salivary gland and one at the base of the skull.

CET presents either as aberrant solid thymic

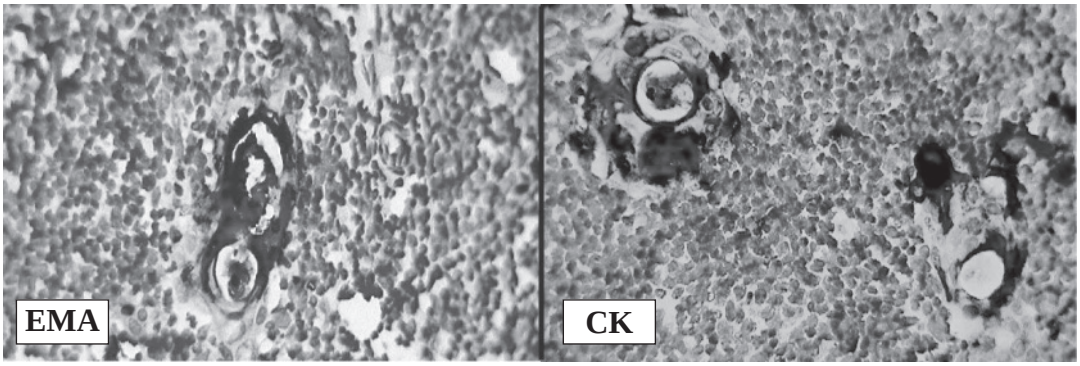

FIG. 4: Hassall's corpuscles and epithelial elements show strong immunohistochemical positivity for EMA and CK. IHC X 400.

tissue, thymic cyst or cervical thymoma.[10] Solid CET constitutes about 10% of all the ectopic thymic masses. The pathogenesis of solid cervical thymic tissue include, arrest of the thymus in its normal descent, failure of involution or sequestration of thymic tissue during descent.[11] Our patient presented with aberrant solid thymic tissue.

Some ectopic thymuses were cystic and classified as thymopharyngeal cysts or pouches. The size of an ectopic thymus varies and may even mimic a tumour, prompting surgical intervention.[9] Cervical thymic cysts result from 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.[2]

The present case report has been made because of the presence of ectopic solid thymus in the neck, an interesting rarity, which per-operatively masqueraded as a lymph node metastasis. Although rare, clinicians, radiologists and pathologists should be aware of CET while evaluating neck masses, especially in children, so that a misdiagnosis, and consequential morbid radical neck dissection, can be avoided.

ACKNOWLEDGEMENT

We wish to thank senior technicians Mrs. Ushanandini and Mr. Ramana for performance of the immunohistochemical stains.

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