Ectopic intrathyroidal thymus in children: Two case reports and review of the literature

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ABSTRACT

Thymic vestiges remaining at certain locations during embryologic thymic descent may result in ectopic thymus formation. Ectopic thymus most commonly occurs in the cervical area or mediastinum, whereas ectopic intrathyroidal thymic tissue is a rare occurrence. This report describes a 26-month-old male patient and a 52-month-old female patient, both of whom presented with an asymptomatic left intrathyroidal mass. The imaging findings were nonspecific and preoperative fine-needle aspiration (FNA) biopsies were lack, so preoperative diagnosis was difficult. After surgical resection, biopsy showed ectopic intrathyroidal thymic tissue. Differential diagnosis of thyroid nodules or neck masses should include ectopic thymus. Correct diagnosis of ectopic thymus must rely on the histopathologic results, and FNA biopsy may be a feasible way.

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1. Case reports

1.1. Case 1

A 26-month-old male patient was admitted to our hospital in November 2008 because of a left intrathyroidal nodule found during examination of the anterior cervical lymph nodes one month previously. The patient had no obvious symptoms such as fever or pain. Past history, including history of allergies, was unremarkable. The patient appeared to be in good general health, and no unusual findings regarding the head, chest, abdomen, or extremities were made. Two to three lymph nodes, approximately 0.3 × 0.5 cm in size, could be palpated on both sides of the neck but no obvious mass was found. Findings of the following thyroid function tests were normal: TT3 (total triiodothyronine), TT4 (total tetraiodothyronine), FT3 (free triiodothyronine), FT4 (free tetraiodothyronine) and TSH (thyroid-stimulating hormone). All tests for thyroid antibodies were negative. Chest X-ray was normal. An ultrasound of the neck revealed a hypoechoic nodule within the left thyroid. Neck CT revealed a space-occupying lesion within the left thyroid (Fig. 1A and B). Radionuclide scans identified a “cool nodule” in the middle and upper regions of the left thyroid lobe (Fig. 2A and B). According to the clinical manifestations and laboratory findings, preoperative diagnosis of a left thyroid adenoma was made. Because preoperative fine-needle aspiration biopsy was rejected by the parents, resection of the left thyroid nodule was then performed. During the operation, a soft nodule 1.0 × 0.8 × 0.6 cm in size was observed at the upper pole of the left thyroid; the capsule was complete and the boundary was clear. Postoperative biopsy showed thymic tissue in the left thyroid. Neck CT revealed a space-occupying lesion within the left thyroid (Fig. 3). Preoperative chest CT was relived postoperatively and showed no nodule tissue in the mediastinum. Immune system function (CD series) and thyroid function tests were both normal postoperatively, and no nodule recurrence was found at follow-ups throughout 4.5 years.

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1.2. Case 2

A 52-month-old female patient was admitted to our hospital in January 2011 because of the presence of a neck mass of 2 days duration. The patient exhibited no obvious symptoms such as fever, cough, pain, fatigue, or heat intolerance. Past history, including history of allergies, was unremarkable. The general health of the patient was good, and no unusual findings concerning the head, chest, abdomen, or extremities were made. A medium-textured mass could be palpated in the middle region of the left thyroid. The nodule size was approximately 2.0/1.2 cm, the shape was irregular, and the boundary was unclear. No tenderness was present, and nodule movement was observed during swallowing. Another medium-textured mass could be palpated in the lower region of the left thyroid above the sternal fossa; the size was approximately 1.0/0.6 cm and the boundary was unclear. No tenderness was present, and movement of this nodule was also observed with deglutition. Thyroid function tests included TT3, TT4, FT3, FT4, and TSH; all findings were normal. All tests for thyroid antibodies were negative. Chest X-ray was normal. Neck ultrasound revealed two hypoechoic nodules, respectively within and at the inferior pole of the left thyroid. Neck CT disclosed a space-occupying lesion inside the left thyroid, and some lesions extended to the thoracic inlet (Fig. 4A and B). Radionuclide scans revealed a “cold nodule” in the left thyroid lobe (Fig. 5). Considering all clinical manifestations and laboratory findings, the possibility of benign tumors in the left thyroid and its inferior area was considered preoperatively. Because preoperative fine-needle aspiration biopsy was failed, neck exploration and tumor resection were then performed. During the operation, a suspected “lymph node” was found superior to the sternum; the size was approximately 1.2 × 0.6 × 0.5 cm, and no adhesion between the lesion and the thyroid tissue was observed. A soft nodule was palpated at the middle and lower pole of the thyroid. The superior part of this lesion was connected with the upper pole of the thyroid by a fine cord (Fig. 6); the lesion was approximately 2.5 × 1.2 × 1.0 cm in size, the capsule was complete, and the boundary was clear. The “lymph node” and thyroid nodules were removed and cut open. Grossly, the tumors were fish-meat like appearance. Postoperative biopsies revealed that the “lymph node” inferior to the thyroid and all of the thyroid nodules were comprised of thymic tissue (Fig. 7). Preoperative chest CT was relived postoperatively and showed no thymus tissue in the mediastinum. Postoperative immune system function (CD series) and thyroid function tests were normal, and no recurrence of lesions was found at a 2.5-year follow-up.

Fig. 1. A and B. In case 1, neck CT scan showing left intrathyroidal mass (red arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article).

Fig. 2. A and B. In case 1, radionuclide scan showing even distribution of radioactivity in the right thyroid lobe, but sparse distribution in the upper and middle regions of the lobe which is consistent with the presence of a “cool nodule” (red arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article).
2. Discussion

Normal thymus tissue, which is located in the superior and anterior compartments of the mediastinum, develops from the endoderm of the 3rd and 4th pharyngeal pouches. At the 6th week of the embryonic period, the endodermal epithelium on the ventral side of the 3rd and 4th pharyngeal pouches and the ectodermal epithelium of the brachial groove on the opposite side proliferate to produce two cell cords. These cords extend toward the end of the embryo, with their ends reaching the thoracic cavity; the cords then proliferate, enlarge and merge to form a thymus primordium. Coordinately with epithelial proliferation and the downward movement of the thyroid primordium to the dorsal side of the thymic primordium, the thyroid primordium divides into two pairs of parathyroid glands followed by degradation of the cell cords. If degradation is incomplete, residual thymic cells can remain in the neck, thyroid, or thoracic cavity to form ectopic thymus tissues.

Ectopic thymus is most commonly found in the cervical area or the mediastinum but is rarely found in hilar tissue, pericardium, chest wall, or axilla. Ectopic intrathyroidal thymic tissue is a rare

Fig. 3. In case 1, histopathologic results showed intrathyroidal thymic tissue (HE staining, 10 × 10).

Fig. 4. A and B. In case 2, neck CT scan showing two masses, respectively within and inferior to the left thyroid (red arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article).

Fig. 5. In case 2, radionuclide scans showing even distribution of radioactivity in the right thyroid lobe, but uneven distribution is in the left lobe. A round radioactive defect with an unclear boundary is observable in the middle and lateral areas of the left thyroid lobe. The findings in the left thyroid lobe are consistent with the presence of a "cold nodule" (red arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article).

Fig. 6. In case 2, nodular mass within the left thyroid can be seen, and the superior part of the nodule is connected with the upper pole of the thyroid by a fine cord (blue arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article).
Surgical resection of the ectopic intrathyroidal thymus was performed for the two patients described in the present study. Active surgical treatment was performed in part because of being unaware of ectopic thymus, but mainly to avoid the possibility of malignancy. Although thyroid masses or nodules are rarely seen in children, the probability that the mass or nodule is malignant is higher in children than in adults; the incidence of cancerous childhood thyroid nodules approaches 20% [18]. Nonetheless, surgeons currently face considerable challenge in distinguishing between benign and malignant thyroid lesions. The clinical examination alone cannot provide a definite diagnosis, and imaging examinations cannot definitively distinguish between benign and malignant tumors. At present, fine-needle aspiration (FNA) biopsy is applied widely in the early diagnosis of adult thyroid nodules. However, the value of this procedure in the diagnosis of childhood thyroid nodules remains to be controversy. Skinner [19] reported the following three reasons. The first relates to the small size of thyroid nodules in children, who are less willing to accept this procedure. Accurate and safe aspiration biopsy is achieved only after repeated sedation of the subject. The second relates to insufficient experience of the pediatric pathologists in interpreting thyroid cytology. The third relates to the known incidence of cancerous thyroid nodules, which is higher in children than in adults. FNA biopsies have resulted in false negative rates of 1–6% such that treatment of malignant thyroid tumors may be delayed. In view of the above reasons, many pediatric surgeons forgo the FNA before the operation, but choose directly surgical exploration. Operative procedure is determined according to the frozen pathological results during the operation. To our experience and lesson, FNA should strongly be considered for thyroidal lesions, surgical resection may be obviated if FNA shows thymic tissue, but if FNA is inconclusive, surgical resection is probably best for thyroid lesions because of CA risks.

3. Conclusion

In conclusion, the presence of a thyroid mass or nodule in a child represents a considerable challenge to the pediatric surgeon. Because accurate evaluation is difficult, beware of delayed treatment of a malignant tumor and excessive treatment of benign tissues (such as ectopic thymus). Differential diagnosis of a thyroid nodule or neck mass should include ectopic thymus. Because the ectopic thymus location is not sure, and standard laboratory tests and imaging studies provide non-specific findings, the preoperative definitive diagnosis is difficult. Correct diagnosis of ectopic thymus must be made only after appropriate histopathologic assessment of the surgical specimen, and FNA biopsy may be a feasible way. Surgical resection is probably best for thyroid lesions because of CA risks if FNA is inconclusive.

Conflict of interest statement

The authors declare that there are no conflicts of interest about the paper “Ectopic intrathyroidal thymus in children: two case reports and review of the literature”.

References


