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Caso Clínico

Benign Thyroidal Lesion: Intrathyroidal Ectopic Thymus

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A B S T R A C T

Thyroid nodules are uncommon in pediatric patients, much less prevalent than adults. Despite having a higher probability of malignancy, most thyroid nodules are benign. Intrathyroidal ectopic thymus (IET) is a rare entity due to abnormal thymic migration during embryogenesis. The diagnosis is usually incidental and patients tend to be asymptomatic. Ultrasound is the recommend diagnostic method, since IET has a typical and unique appearance on ultrasonography. It is essential to recognize and differentiate IET from malignant thyroid nodules in order to avoid unnecessary surgeries and/or invasive diagnostic procedures, such as fine needle aspiration biopsies. The risk of malignant transformation is extremely low, therefore IET can be safely managed with serial ultrasound, without any treatment.

Palavras-chave:
Criança;
Doenças da Tiroide;
Timo.

Lesão Tiroideia Benigna: Timo Ectópico Intratiroideu

R E S U M O

Os nódulos tiroideus são pouco frequentes em idade pediátrica, com uma prevalência muito inferior à dos adultos. Apesar da probabilidade de malignidade ser maior, a maioria deles é benigna. O timo ectópico intratiroideu (IET) é uma entidade rara, resultante da migração aberrante do timo durante a embriogénese. O seu diagnóstico geralmente é acidental e os doentes assintomáticos. O melhor método diagnóstico é a ecografia, pelas características ultrassonográficas únicas e típicas do IET. É essencial fazer o diagnóstico diferencial com nódulos malignos da tiroide, evitando assim cirurgias e/ou procedimentos diagnósticos invasivos desnecessários, como biópsias por aspiração com agulha fina. Uma vez que o risco de transformação maligna é extremamente baixo, está recomendado apenas controlo ecográfico seriado, sem qualquer tipo de terapêutica dirigida.

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Introduction

Thyroid nodules are rare in children, with a prevalence of 0.2% to 2%, much lower than adults.[1] Despite having a higher probability of malignancy (20%-26%), most thyroid nodules are benign.[2]

Intrathyroidal ectopic thymus (IET) is a benign differential diagnosis of thyroid lesions in pediatric population, first described in 1937 by Gilmour,[3] and is due to an aberrant migration of the thymus during embryogenesis. Patients tend to be asymptomatic and IET is usually discovered incidentally.[4] It has been described as a rare entity although imaging advances have increased its diagnosis in the last decade. Recent studies have showed that 1% to 13% of children who undergone neck ultrasound (US) have lesions demonstrating radiographic features of IET.[5]

Case Report

A previously healthy seven-year-old boy was observed in a Pediatric Surgery consultation for a palpable left cervical mass. A neck ultrasound was requested and revealed a lesion with characteristics of a branchial remnant and incidentally, a nodular heterogeneous lesion with 24 mm of diameter, located posterior and laterally to the left thyroid lobe that extended until the upper portion of the thymus. This lesion had similar echotexture within the thymus and the most probable diagnosis was cervical and intrathyroidal ectopic thymus.

The patient underwent surgical treatment for excision of the branchial remnant that was later confirmed by a pathological study, and referred to a Pediatric consultation. Personal and family histories were irrelevant and the patient had no clinical complaints. Upon physical examination, no alterations were found and no cervical mass was palpable.

A year later, a control US was performed and it described a hypertrophied thymic left lobe that was in contact with the inferior portion of the left thyroid lobe, and a well demarcated solitary thyroid nodule in the right lobe, with 6x4 mm. (Fig. 1) Serum

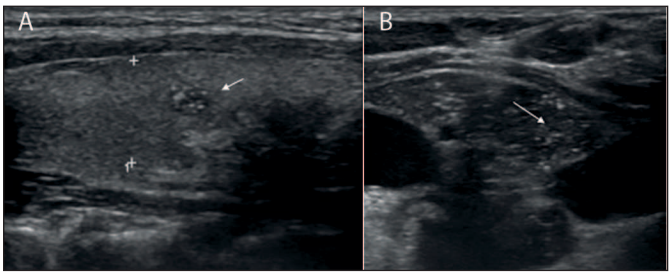

Figure 1. Thyroid and mediastinum ultrasound.

Longitudinal ultrasound showing a 6 mm, well defined, hypoechoic nodule in the right thyroid lobe (arrow in A). The nodule has a heterogeneous echotexture, characterized by punctate and granular bright internal echoes; this was identical to the echotexture of the orthotopic thymus (arrow in B). These imaging findings, compatible with an intrathyroidal ectopic thymus, were better characterized by MRI (Figure 2).

TSH concentrations, FT3 and FT4, serum parathyroid hormone and calcitonin concentrations were normal. Antiperoxidase and antithyroglobulin antibodies were negative. A cervical magnetic resonance imaging (MRI) confirmed the presence of thymic hyperplasia, that projected to the cervical region laterally to the trachea. A nodule with 7 mm in the right thyroid lobe with similar echotexture to the thymus was detected, confirming the diagnosis of intrathyroidal ectopic thymus and cervical thymus. (Fig. 2)

Two years after the first US, another one was performed and

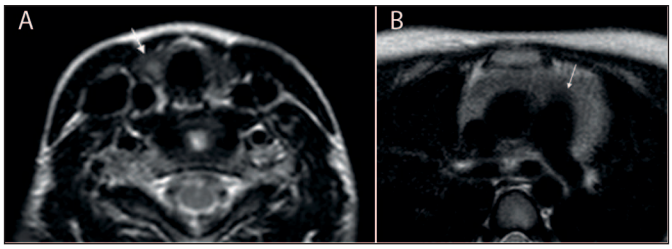

Figure 2. Thyroid and mediastinum MRI.

Axial T2W TSE images of the thyroid and mediastinum showing a 6 mm well defined nodule in the right thyroid lobe (arrow in A) with high T2W signal intensity, identical to that of the mediastinal thymus (arrow in B), findings in keeping up with an intrathyroidal ectopic thymus.

it presented the same heterogeneous nodule with punctate echogenic foci with 5 mm of diameter, with well-defined margins, maintaining the same echotexture within the thymus. The cervical thymus had involved, as expected to the age group. The annual ultrasounds have demonstrated a decrease in size and margins less defined of the intrathyroidal ectopic thymus, as well as a complete involution of the cervical thymus, and the annual blood analysis showed normal thyroid and parathyroid functions. The patient is now ten years-old and remains asymptomatic without any palpable cervical mass.

Discussion

The thymus is the central lymphoid organ of infancy and is responsible for the formation of mature T lymphocites.[6] The development of the thymus starts by the sixth week of gestation from ventral sacculation of the third pharyngeal pouch and minor portions of the fourth pharyngeal pouch. Around the eighth week of gestation, the bilateral primordial thymus fuses at the midline to form the bilobed thymus and later descend into the superior mediastinum.[1] It is possible to find ectopic thymic tissue along the normal pathway of descent to the thorax, but other locations have been described (e. g. pharynx, trachea, esophagus or mediastinum).[7]

IET has a slight male preponderance.[1,5,8] The majority of cases described in the literature correspond to incidental findings in neck US that was requested for other reasons, like torticollis, palpable swelling in the neck, goiter or congenital hypothyroidism.[8]

Once a thyroid nodule is found, the differential diagnosis includes nodular goiter, follicular adenoma, colloid cyst, branchial remnants, thyroglossal duct cyst, intrathyroidal ectopic thymus, lymphatic or vascular malformations and thyroid malignancies.4 The correct diagnosis of IET avoids unnecessary invasive procedures as surgery and/or fine-needle aspiration (FNA) biopsies.[1,9]

IET rarely gives rise to symptoms,[7] and the same happens regarding the thyroid and parathyroid hormones. Most cases of IET are euthyroid and abnormal thyroid levels are caused by concurrent pathologies, such as thyroiditis.[1]

This entity has very typical US features. All articles describe IET as a hypoechoic lesion, with punctate and linear internal echogenic foci.[4,6,8,10] The most common shape is fusiform (up to 90%) with well-defined but slightly irregular borders. Fusiform shape and a maximum diameter of 9 mm are the most selective criteria to predict IET diagnosis, according to Aydin *et al.*[1] The most typical location is not consensual, as some authors describe the lower posterior portion of the thyroid as the most common,[4,6] and others say it is more frequently located in the mid portion of the thyroid.[10] Hypoechogenic lesions, with irregular margins, and increased intranodular blood flow, and the presence of microcalcifications and/or abnormal cervical lymph nodes in US increase

the suspicion of malignancy, justifying the need of FNA biopsy.[2]

Segni et al reported a decrease in size of the lesions and margins less defined with time, probably reflecting the normal thymus involution occurring with increasing age.[7] Purcell *at al* also found a significant decrease in volume of IET over time.[5] The thymus tends to increase in size during childhood before involving during adolescence.[5] It is expected that IET follows the same growth trend of the thymus, since it is normal thymic tissue in an abnormal location.[6] The same happened to our patient, reinforcing the idea that intrathyroidal thymic nodules can be monitored annually by US, without the need of surgery or fine-needle aspiration biopsies. In case of doubt, magnetic resonance imaging (MRI) may be considered, especially for larger lesions, to compare the texture of the nodule with that of thymic tissue within the mediastinum.[5] Cases of malignant transformation were reported in the literature. Intrathyroid thymic carcinoma is an extremely rare entity that has been estimated to account for 0.08%–0.15% of primary thyroid malignant tumors, with a much more favorable prognosis than a primary thyroid squamous cell carcinoma or a poorly differentiated thyroid carcinoma.[11]

The widespread of thyroid and neck US in current days may lead to an increase detection of IET and other thyroidal nodules. Intrathyroidal ectopic thymus must always be kept in mind, in order to avoid fine-needle aspiration biopsies and/or thyroid surgery. IET has unique ultrasound features and neck US performed by an experienced radiologist can prevent unnecessary invasive diagnostic procedures. This entity can easily and safely be monitored by serial ultrasounds.

Responsabilidades Éticas

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