

ECTOPIC CERVICAL THYMUS: A CASE REPORT

Timo ectópico cervical: Presentación de un caso

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Summary

Introduction: Ectopic thymus is a rare disease characterized by a non-painful mass on the neck, which may be cystic or solid, resulting from an alteration in the process of migration of the thymus primordia during gestation. Proper interpretation, within the broad spectrum of differential diagnoses, is very important to avoid unnecessary invasive management.

Case presentation: We present the case of an 8 months-old boy, with no relevant history, with bulging in the right submandibular region, not painful, that in the exploration by ultrasound and magnetic resonance imaging characteristics were found identical to the thymus orthoticks, constituting a rare case of solid ectopic thymus, which was taken to surgery. The pathology corresponded to ectopic thymus.

Discussion: The thymus is an organ located in the anterosuperior mediastinum that plays an important role in cell-mediated immunity. It develops embryologically from the third and fourth brachial arches and migrates through the pharyngeal thymus conduit from the angle of the mandible to the mediastinal cervical junction. Ectopic thymic tissue can occur at any point along the pharyngeal thymus conduit. The incidence is not clearly known. They are more common in the left neck, cystic, in men, between 2 and 13 years and of unilateral presentation. The normal appearance of the thymus, and therefore of the solid ectopic thymus, is exactly the same in the different imaging modalities. Differential diagnoses include thyroglossal duct cysts, gill cysts, dermoid tumor, hemangioma, fibromatosis colli, teratoma, lymphoma and neuroblastoma, among others. **Conclusion:** The ectopic thymus is a rare cervical lesion that simulates pathology, so it is important to understand the radiological characteristics that allow an adequate diagnosis to prevent interventions and unnecessary surgeries.

Resumen

Introducción: El timo ectópico es una patología poco frecuente que se caracteriza por una masa en el cuello no dolorosa, que puede ser quística o sólida, resultado de una alteración en el proceso de migración de los primordios del timo durante la gestación. La adecuada interpretación, dentro del amplio espectro de diagnósticos diferenciales, es muy importante para evitar manejos invasivos innecesarios. **Presentación de caso:** Se trata de un niño de 8 meses, sin antecedentes relevantes, con abultamiento en la región submandibular derecha, no dolorosa, a quien en la exploración mediante ecografía y resonancia magnética se le encontraron características de imagen idénticas al timo ortotópico, lo que constituye un raro caso de timo ectópico sólido. Posterior a la cirugía, el resultado de patología correspondió a timo ectópico. **Discusión:** El timo es un órgano



Key words (MeSH)

Thymus gland
Head and neck neoplasms
Magnetic resonance imaging

Palabras clave (DeCS)

Timo
Neoplasias de cabeza y cuello
Imagen por resonancia magnética



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ubicado en el mediastino anterosuperior que tiene un papel importante en la inmunidad mediada por células. Proviene embriológicamente del tercer y cuarto arcos braquiales y migra a través del conducto timofaríngeo desde el ángulo de la mandíbula hasta la unión cérvico-mediastínica. El tejido tímico ectópico puede ocurrir en cualquier punto a lo largo del conducto timofaríngeo. La incidencia no es claramente conocida. Es más común en el cuello izquierdo, quístico, en hombres, entre los 2 y 13 años y unilateral. La apariencia normal del timo, y por ende de los timos ectópicos sólidos, es exactamente igual en las diferentes modalidades de imagen. Los diagnósticos diferenciales incluyen quistes del conducto tiroglosa, quistes branquiales, tumor dermoide, hemangioma, *fibromatosis colli*, teratoma, linfoma y neuroblastoma, entre otros. **Conclusión:** El timo ectópico es una lesión cervical rara que simula patología, por lo cual es importante entender las características radiológicas que permitan un adecuado diagnóstico para prevenir intervenciones y cirugías innecesarias.

Introduction

The thymus is a mediastinal organ involved in cellular immunity, with a characteristic embryonic origin that can lead to ectopic tissue along its migration route.

Ectopic thymus is a rare pathology, characterized by a painless neck mass, which may be cystic or solid, and its proper interpretation within the broad spectrum of differential diagnoses is very important to avoid unnecessary invasive management.

In this article we present the case of an 8-month-old boy in the study of a submandibular asymmetry that was diagnosed by images as an ectopic thymus, had surgery and the pathology confirmed the diagnosis of ectopic thymus.

Presentation of the case

This is an 8-month-old child, with no relevant history, whose mother consults because she perceives a bulge in the right submandibular region, not painful, approximately from the age of 2 months and which has apparently increased in size.

On physical examination, slight asymmetry of the soft tissues of the neck was observed, with no pain on palpation and no evidence of a defined mass. The ultrasound scan identified a well-defined mass, hypo-echoic, with small septa inside, linear, thin and echogenic, with flow to the scan with color Doppler (Figure 1). When exploring the anterior mediastinum intercostally, the thymus was found with the same characteristics of ecogenicity of the structure described in the submandibular region (Figure 2).

A magnetic resonance imaging (MRI) of the neck with contrast medium was performed, in which a mass was identified posterior to the right submandibular gland, anterior and lateral to the carotid space and inferior to the parotid gland (Figures 3 and 5b), homogeneous, with well-defined contours, intermediate signal with T1 information (Figure 2) and discreetly high in T2 and STIR (Figure 4), with slight homogeneous enhancement after injection of contrast medium (Figure 5). The average mass was approximately $31 \times 21 \times 26$ mm and had the same signal characteristics as the mediastinal thymic tissue in all sequences.

Discussion

The thymus is an organ made up of two lobes, located in the anterosuperior mediastinum, behind the sternum, which plays an important

role in cell-mediated immunity as it is the primary site of T lymphocyte maturation. It is characterized histologically by structures called Hassall corpuscles, which give the histological diagnosis of thymic tissue (1-3).

The thymus comes embryologically from the third and fourth gill arches bilaterally at approximately six weeks of gestation and contains elements derived from the three germ layers (1, 4). Approximately in the seventh week of gestation there is an elongation and cylindrical formation that gives rise to the thymopharyngeal duct, which will be the structure through which the caudal and medial migration of the thymic primordium occurs, from the angle of the mandible to the cervix-mediastinal junction, ventral to the carotid space. Finally, the migrating bilateral primordians fuse in the anterior and superior mediastinum at approximately week 8 (1,4,5).

Ectopic thymic tissue can occur at any point along the thymopharyngeal canal as a result of descent failure, sequestration, or remnant tissue involution failure (2,5).

The ectopic thymus is rare, its incidence is not clearly known because the frequency of subclinical ectopic thymus has not been estimated (6). It should be clarified that the definition of ectopic is different from aberrant, when there is aberrant tissue is located in regions different from those explained by the migration process, such as retropharyngeal or dermis (5).

The most common locations of ectopic thymus found in some case series indicate that it is in the cervix-mediastinal junction or lower neck (1); however, there are other case series that report the most frequent location in the submandibular region, just anterolateral to the carotid space, posterior to the submandibular gland and anterior to the sternocleidomastoid muscle (6). They are most common in the left neck (70%) (1), cystic in nature, in men, between 2 and 13 years of age and unilateral presentation (1,3).

The ectopic thymus is characterised by its fusiform shape, with the rostral portion extending towards the angle of the mandible and the caudal aspect, with a tail, extending towards the upper mediastinum. Approximately 50% (1) can connect with the anterior mediastinum by a cord or direct extension (1,3).

The clinical presentation varies depending on the age of the patient, the size of the lesion and whether it is cystic or solid. 90% usually manifests as a painless cervical mass (3) that can vary in size by isolated processes of respiratory infection or vaccination and can be misinterpreted as a pathological process. Stridor, dyspnea and dysphagia caused by mechanical compression are rare (1,5).

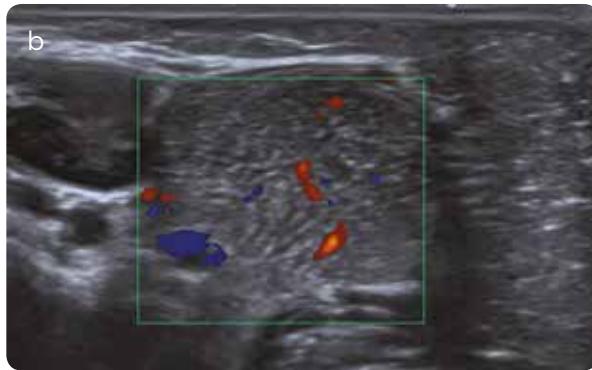
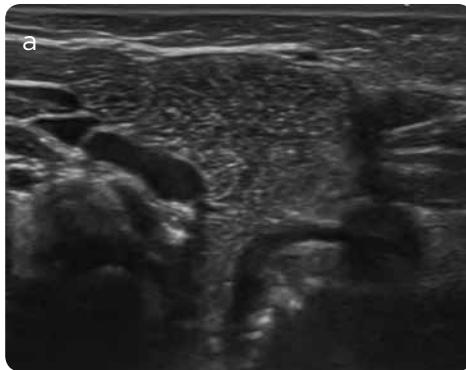


Figure 1. a) Ultrasound mode B: a well-defined, hypo-echoic mass is identified, with small septa in its interior, linear, thin and echogenic. b) Mass with flow after color Doppler exploration.

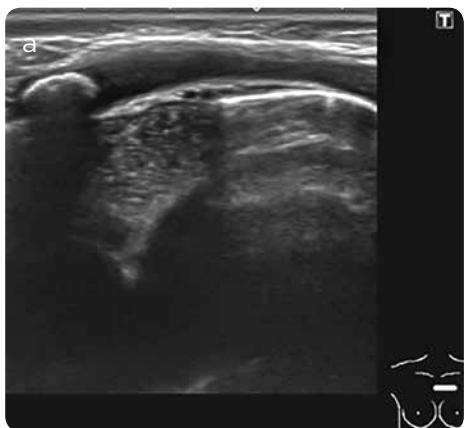


Figure 2. a) Intercostal exploration of the anterior mediastinum: the mediastinal thymus is observed, with the same characteristics of ecogenicity of the mass in the submandibular region (b).

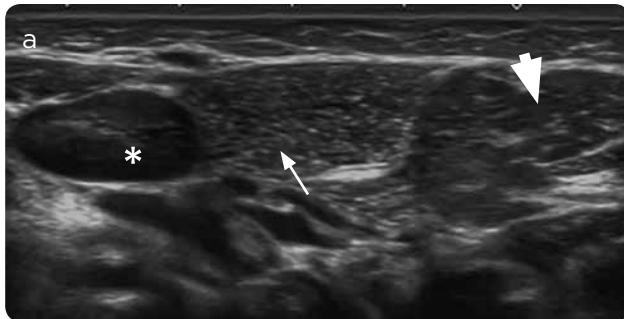


Figure 3. a) Ultrasound and b) MR with axial T1 information: mass (short arrow) posterior to the right (arrowhead) and anterior submandibular gland and lateral to the carotid space (long arrow). The jugulodigastric ganglion (asterisk) can also be observed on ultrasound.

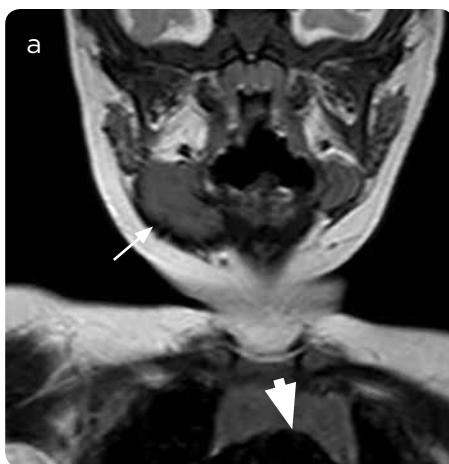


Figure 4. a and b) MR with T2 and STIR information: slightly high signal strength is identified for both the submandibular mass (arrow) and the mediastinal thymus (arrowhead).

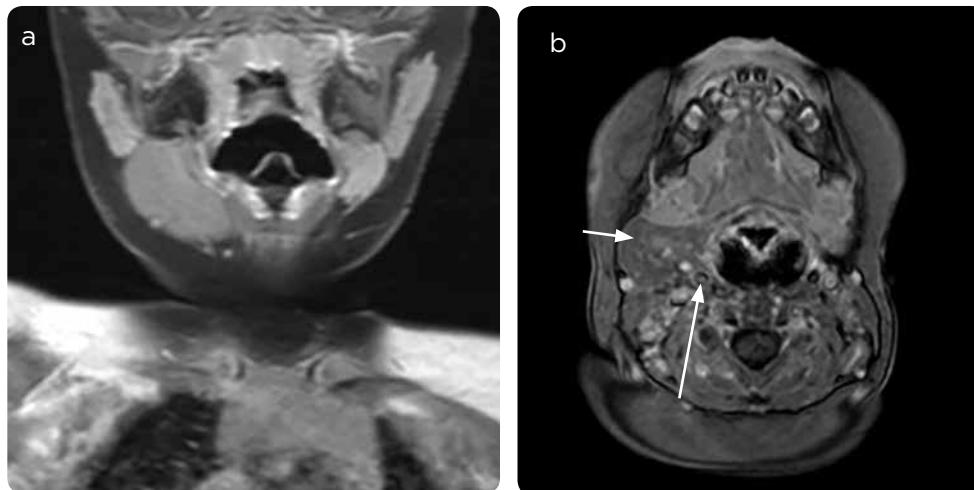


Figure 5. a) MR with gadolinium: slight and homogeneous enhancement of both the submandibular mass and the mediastinal thymus. b) Anterior and lateral relationship of the mass (short arrow) with the carotid space (long arrow).

The findings in images depend on the cystic or solid nature of the ectopic tissue, of which the most frequent is the cystic type (1,3). The cystic nature can be explained by the degeneration of Hassall's corpuscles within the ectopic thymus (1,6).

Ultrasound is the best imaging modality for the initial study. In cystic ectopic thymus, a hypo- or anechoic, unilocular or internal septa mass is identified (1).

The normal appearance of the thymus, and thus of the solid ectopic thymus, is small linear echoes representing septa of connective tissue and vessels within the septa.

In contrast media scans they are observed as a non-specific low signal mass with peripheral ring enhancement that requires further study, so it is not the study of choice (1,6).

In MRI, the ectopic thymus shows greater signal intensity than cerebrospinal fluid with T1 information and high signal in T2. It has no restriction on diffusion and after administration of contrast shows thin and peripheral enhancement of its wall. For solid ectopic thymus the same signal strength can be seen in all sequences to the normal mediastinal thymus, if present (1,3).

Differential diagnoses include benign solid and cystic masses and less likely malignant tumors. These include thyroglossal duct cysts, gill cysts, dermoid tumor, hemangioma, fibromatosis colli, teratoma, lymphoma, and neuroblastoma. There is no evidence that ectopic tissue has an increased risk of malignancy, although a small number of cases of transformation to malignant thymoma have been described (6).

Thanks to the introduction of ultrasound and MRI, there was a change in the diagnosis and management of patients with ectopic thymus, since now, in most cases, imaging is considered sufficient without the need for surgical interventions and with conservative management (6,7). If the images are clear in the diagnosis and the patient is asymptomatic, conservative management can be made, reserving the surgery for the moment when it becomes symptomatic or has complications (3,6,7).

It is important to bear in mind that before surgical management, the anterior mediastinum must be examined to determine if there is thymic tissue in its normal location and not to perform total thymectomy, because the patient may develop immunodeficiency. This is because the absence of orthotopic thymus with ectopic or aberrant tissue has been found in up to 50% of cases (3,6,7).

Conclusions

The ectopic thymus is a rare cervical condition that simulates pathology, so it is important to understand the radiological characteristics that allow a proper diagnosis in order to prevent unnecessary interventions and surgeries. Differential diagnosis should always consider this diagnosis for all cystic masses in the pediatric neck and remember that when a solid mass is found it should always be compared with the orthotopic thymus.

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Received for assessment: August 14, 2017

Accepted for publication: June 6, 2018