

## Case Report

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# Ectopic cervical thymus: A case report of uncommon neonatal neck mass

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**Keywords:** ectopic thymus; congenital anomalies; neck masses.

**Abbreviations:** CT: computed tomography; MRI: Magnetic resonance imaging; US: ultrasound; WI: weighted image.

### Abstract

**Introduction:** Ectopic thymus is an uncommon abnormality in which there would be a mass in the neck which may be solid or cystic and usually the patient experiences no pain. Consideration of this mass entity in the differential diagnosis is necessary to avoid proceeding to any non-indicated invasive intervention.

**Case presentation:** A case of two-months baby, with no relevant history, whose parents asked for sought for medical advice because they noticed a bulge in the left submandibular region which appears to be non-painful. The sonographic examination revealed a well-defined hypo-echoic rather oval mass that shows echoic, with multiple thin linear hyperechoic septa as well as homogeneous discretely distributed foci that shows hyperechogenic patten giving rise to an appearance that can be called "speckled" or "starry sky". On Color Doppler study some flow was detected.

A contrast enhanced magnetic resonance imaging (MRI) of the neck was done. It revealed a well-defined mass lesion that located at anterior aspect of common carotid artery, jugular vein & sternocleidomastoid muscle, posterior to the left mandible. It extends caudally to lower cervical region. It displayed homogenous signal intensity, iso-intense to the muscles at T1WI and intermediate signal at T2 WI. Slight post contrast homogenous enhancement was detected. The signal of the mass was similar to that of the normal mediastinal thymic tissue. A tail component was noted extending from the lesion to deep cervical layer. The average mass was approximately 34 X 27 X 17 mm.

**Conclusions:** The ectopic thymus is an uncommon lesion that may be mistaken with other pathologies that can happen in that anatomical area, so it should be included in the differential diagnosis for all pediatric neck masses. Proper diagnosis can be achieved by understanding the radiological characteristics to avoid unnecessary interventional procedures. Confirmation of presence or absence of orthotopic thymus is crucial in reporting.

## Introduction

The thymus is an organ that has a role in the process of cellular immunity, it is normally located in the mediastinum. From embryological view the thymus has a unique origin which may result in presence of its tissue in ectopic place along the course of its migration [1-5].

Ectopic thymus is an uncommon abnormality in which there would be a mass in the neck which may be solid or cystic and usually the patient experiences no pain [6,7].

Consideration of this mass entity in the differential diagnosis is necessary to avoid proceeding to any non-indicated invasive intervention [8-10].

In our current work, we present a case of a two-months baby with left submandibular neck swelling extending to upper cervical region that was diagnosed by images as an ectopic thymus.

## Case presentation

A case of two-months baby, with no relevant history, whose parents asked for sought for medical advice because they noticed a bulge in the left submandibular region which appears to be non-painful.

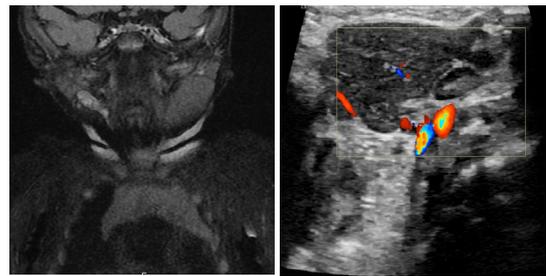
On clinical examination, asymmetry of the neck soft tissues was noticed with left submandibular & upper cervical swelling that showed no pain on palpation.

The sonographic examination revealed a well-defined hypo-echoic rather oval mass that shows echoic, with multiple thin linear hyperechoic septa as well as homogeneous discretely distributed foci that shows hyperechogenic pattern giving rise to an appearance that can be called "speckled" or "starry sky". On Color Doppler study some flow was detected (Figure 1).

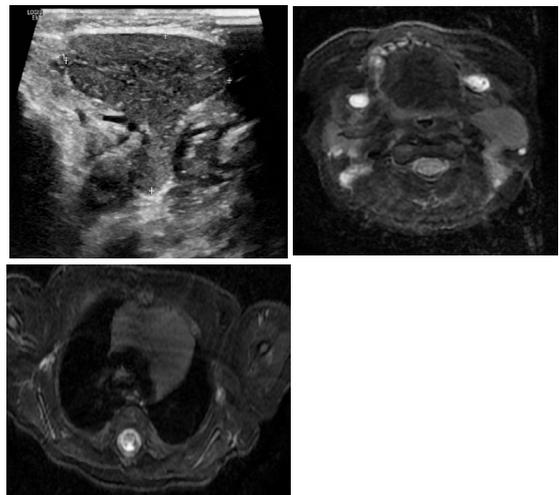
After informed parent's consent, a contrast enhanced magnetic resonance imaging (MRI) of the neck was done. It revealed a well-defined mass lesion that located at anterior aspect of common carotid artery, jugular vein & sternocleidomastoid muscle, posterior to the left mandible. It extends caudally to lower cervical region. It displayed homogenous signal intensity, iso-intense to the muscles at T1WI and intermediate signal at T2 WI. Slight post contrast homogenous enhancement was detected. The signal of the mass was similar to that of the normal mediastinal thymic tissue. A tail component was noted extending from the lesion to deep cervical layer. The average mass was approximately 34 X 27 X 17 mm (Figure 2). Biopsy showed normal thymic tissue (Figure 3).

## Discussion

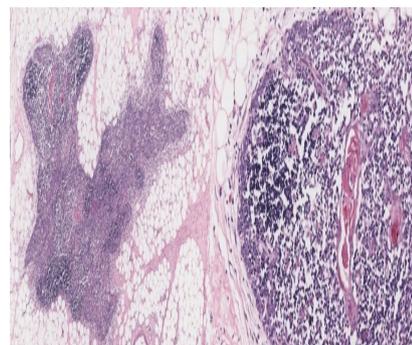
The thymus lies behind the sternum (in the antero-superior mediastinum) and it has two lobes. The thymus has an important participation in the immunity (cell-mediated) as it is considered the primary location of the T-lymphocyte maturation [1-3].



**Figure 1:** Ultrasound (1a) shows a well-defined, hypo-echoic mass with small septa in its interior, linear, thin and echogenic and shows Doppler signal at Doppler study (1b).



**Figure 2:** MRI T1 WI (2a) and T2 WI (b) : the mass displays homogenous signal intensity, iso-intense to the muscles at T1WI with tail component was noted extending from the lesion to deep cervical layer, it displays intermediate signal at T2 WI, its signal is similar to that of the normal mediastinal thymic tissue (2c).



**Figure 3:** Histopathological examination showing normal thymus tissue.

Histologically, the thymus is characterized Hassall corpuscles structures giving its tissue the histological characterization [3-6].

Embryologically, the thymus starts to be developed in the 6th gestational week from the 3rd and 4th gill arches bilaterally and containing the three germ layers [7-9]. In the 7th gestational week, the an elongation and cylindrical formation takes place to form the thymopharyngeal duct.

The caudal and medial migration of the thymic primordium occurs through the thymopharyngeal duct from the mandibular angle to the cervix-mediastinal junction, ventral to the carotid space. Lastly, in the 8th gestational week the two sides primordians should fuse in the superior part of anterior mediastinum. So, as a result of failed descent, sequestration, or failed involution of tissue remnant ectopic thymus will occur. This can take place at any stage of this migration along the thymuspharyngeal canal [1,3-6].

While aberrant thymus means there is aberrant tissue present away from migration tract such as such as retropharyngeal or dermis [5].

The ectopic thymus is uncommon; however, its actual incidence is not well estimated because of missing of subclinical cases [8].

The literature showed different sites reported to be the most common for the ectopic thymus; in some series the cervix-mediastinal junction or lower neck was reported at the most common, in another study the highest incidence was reported to be at the submandibular region (anterolateral to the carotid space, posterior to the submandibular gland and anterior to the sternomastoid muscle. They are most frequent was in the left side of the neck (70%) [2-8].

Characteristically, the ectopic thymus has a fusiform shape, with its cephalic part extending towards the mandibular angle while caudally, its tail extends towards the upper part of mediastinum. In half of cases, it can be connected through a cord or directly with the anterior mediastinum [4,5].

Clinically, the ectopic thymus has variable presentations depending on the patient's age, the lesion size and its consistency (solid or cystic). The majority of cases as presented as a non-painful cervical mass [5]. Rarely it can cause in mechanical compression resulting in dyspnea, stridor and dysphagia [2,5].

Cystic type is more common than solid type in the ectopic tissue this can be attributed to the degeneration which takes place in the Hassall's corpuscles presented in the ectopic thymus [1,6].

Initial imaging can be done by US (ultrasound), the appearance of cystic ectopic thymus can be hypo- or anechoic echopattern, it can be unilocular or having internal septa [1-6].

The solid type shows small linear echoes which corresponds to connective tissue septa and its contained vessels Computed tomography (CT) can show a homogenous density lesion that has intimate relation to the carotid sheath. Usually, the mass has no mass effect on airway or vessels in infancy [3].

In MRI (magnetic resonance imaging) studies, the ectopic thymus has higher signal intensity than cerebrospinal fluid at T1WI with high T2WI signal. It has diffusion restriction. In post

contrast study the lesion shows peripheral wall enhancement while solid type will have similar signal as that appreciated from mediastinal thymus. MRI can be useful also in detection of the connection between the ectopic cervical mass and the mediastinal thymus [1-4].

The differential diagnoses can include broad spectrum of benign solid and cystic masses with less likely and to include malignant masses. The spectrum can include branchial cysts, thyroglossal duct, lymphadenitis, cystic hygromas, dermoid tumors, teratoma, epidermoid cysts, hemangioma, fibromatosis colli, lymphoma, and neuroblastoma.

While little numbers of cases had been reported to show malignant transformation, yet the increased risk of malignancy in ectopic tissue is not confirmed [1-3,10].

Imaging features of ectopic thymus in ultrasound and MRI are reported to be sufficient in most of cases to confirm the diagnosis and to consider conservative management without the need of invasive maneuvers [2-4,6].

So, if the imaging feature are diagnostic for ectopic thymus in asymptomatic patient the conservative management is considered keeping the surgery as a substituting solution in case of complicated and symptomatic situations [5-8].

In about half of cases of ectopic or aberrant thymus, it was noted that orthotopic thymus is absent so it is very crucial to determine the presence of normal site thymus before total thymectomy to avoid subsequent immunodeficiency [5-7,9].

Surgical excision if decided can be done with great care as the mass can be adherent related structures especially the carotid artery, jugular vein and Vagus, hypoglossal, phrenic and recurrent laryngeal nerves. Recurrence after surgical excision is not expected and the prognosis on long-term is reported to be excellent [1-3].

## Conclusions

The ectopic thymus is an uncommon lesion that may be mistaken with other pathologies that can happen in that anatomical area, so it should be included in the differential diagnosis for all pediatric neck masses. Proper diagnosis can be achieved by understanding the radiological characteristics to avoid unnecessary interventional procedures. Confirmation of presence or absence of orthotopic thymus is crucial in reporting.

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