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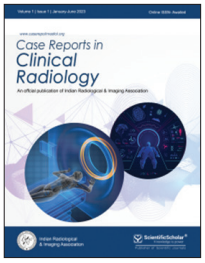

Case Report

A "no-touch" neck "mass" in a child

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ABSTRACT

Aberrant cervical thymus is a rare cause of pediatric neck masses related to the embryological development. In most of the cases, this condition is asymptomatic and essentially benign in nature. Here, we describe the sonological and magnetic resonance imaging (MRI) features of cervical thymus in a 2-month-old baby who presented with the left submandibular swelling. The unique imaging findings on ultrasound and MRI paralleling that of the normal thymus tissue were helpful in confidently establishing the diagnosis. Therefore, the radiologist must be aware of the condition and the typical normal appearance of thymus which can obviate the need for biopsy or surgical intervention.

Keywords: Aberrant cervical thymus, Child neck mass, Ultrasound, MRI neck

INTRODUCTION

Cervical thymus is a rare cause of neck mass in pediatric age group. It can occur anywhere along the embryological pathway of thymus descent from angle of the mandible to superior mediastinum.[1,2] Here, we present a case of 2-month-old baby boy whose mother perceived a submandibular swelling at 15 days of age. High resolution ultrasound and magnetic resonance imaging (MRI) were done to localize the lesion, study its size, extent, and composition which confirmed the diagnosis of aberrant cervical thymus and prevented further intervention and surgery.

CASE REPORT

A 2-month-old baby boy was taken to a pediatrician by his mother who perceived a bulge in the left submandibular region of the neck which was initially noticed at 15 days of age and apparently increased in size since then.

On clinical examination, there was a 3 × 2 cm sized non-tender soft palpable mass in the left submandibular region at the angle of the mandible. There were no signs of inflammation or any skin changes. No restriction of the neck movements was noticed.

High resolution ultrasound showed a well-defined oval shaped mass just inferolateral to the submandibular gland and anteromedial to the sternocleidomastoid muscle. It showed multiple echogenic linear structures and foci under hypoechoic background, which was reminiscent of the echo pattern of normal thymic tissue [Figure 1a]. The thymus in the superior

mediastinum was then imaged through the intercostal spaces which showed a similar echopattern as the lesion in the submandibular space [Figure 1b]. A provisional diagnosis of aberrant left cervical thymus was made. Other differentials include lymph nodes, ectopic thyroid, and fibromatosis colli.

MRI of the neck and upper thorax was performed to confirm the diagnosis and to see for any other ectopic thymus tissues in the neck. MRI revealed a well-defined homogeneous lesion in the left submandibular region and a small similar signal intensity lesion in left lower neck, medial to left common carotid artery along the thymopharyngeal tract with no obvious communication between the two. The signal intensity of both the lesions was similar to that of normal thymus in anterior mediastinum, which was isointense to muscle on axial T1W images, hyperintense to muscle on axial T2W images and coronal fat suppressed T2W images [Figure 2].

The diagnosis of cervical thymus was made based on the location and imaging characteristics of the mass.

As our patient was asymptomatic with typical imaging features consistent with cervical thymus, parents were reassured and explained that no specific intervention was required and the child was kept on clinical follow-up half yearly with high resolution ultrasound of neck.

DISCUSSION

Thymus is a specialized primary lymphoid organ of the immune system. Embryologically, it is developed from the ventral wings of third and fourth pharyngeal pouches on each side and it is pulled inferomedially forming thymopharyngeal duct.[1-3] During its descent, the tract obliterates and the thymus reaches its normal position in anterior mediastinum. However, in this process, the thymic remnant can occur anywhere along the tract from angle of the mandible to superior mediastinum.

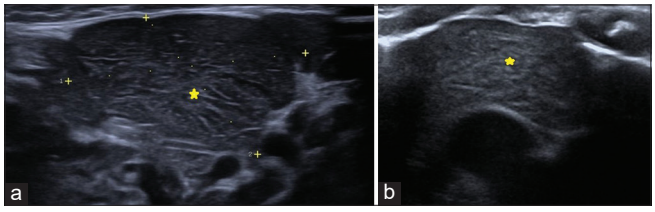

Figure 1: (a) High resolution ultrasound showing well defined hypoechoic lesion with multiple linear hyperechoic structures and foci in the left submandibular region, measuring 2.8 × 2.1 cm. (b) High resolution ultrasound image of the normal thymus gland in anterior mediastinum showing its unique echopattern with echogenic foci. Both the echopatterns were similar which is represented with a yellow *.

Aberrant cervical thymus is a rare cause of neck masses in children,[4] most of them being otherwise asymptomatic as in our case. Very small fraction of patients (6 in 100) can have symptoms related to compression of trachea or esophagus such as stridor, dyspnea, and/or dysphagia. It has a male preponderance and occurs relatively more commonly on the left side than on the right side. Small percentage of them, 5–7% are seen in midline or the pharynx. The mass could be because of the hyperplasia of the undescended thymus which is associated with absent thymus gland in its normal position or hyperplasia of sequestrated thymus remnants along with well-developed normal orthotopic thymus gland as in our case.

Aberrant thymus gland can be solid or cystic. Solid lesions represent early stage and cystic lesions represent late stage in thymic development. Other ectopic thymic masses include thymic cysts, thymopharyngeal tract or cyst, mediastinal thymus with cervical extension, and ectopic thymus out of the normal descent pathway of thymus. Very rarely, ectopic thymic tissue may undergo hyperplasia and malignant transformation such as thymic carcinoma or lymphoma with paucity of the literature.

Differential diagnosis of the neck masses in infants includes lymphadenopathy, venolymphatic malformations, dermoid, ectopic thyroid or thyroglossal cysts, branchial cysts, fibromatosis colli, and other metastatic lesions.[5,6] High resolution ultrasound is the first modality of choice particularly in children due to the lack of ionizing radiation and need for sedation or contrast administration.[6,7] The location the neck mass with typical thymus echopattern on ultrasound will confidently establish the diagnosis. According to a study by Han *et al*.[3] on ultrasound and histopathological thymus anatomy, multiple echogenic linear structures in thymus represent connective tissue septa and accompanying blood vessels. They have concluded that f are unique to make the diagnosis and biopsy may not be required. Zielke *et al*.[8] also concluded that the neck mass can be confirmed by ultrasound as aberrant thymus based on the unique imaging features. MRI is adjuvant in diagnosis of the cervical thymus by demonstrating the same signal to that of the orthotopic mediastinal thymus in all the sequences.[9] It is also useful to rule out other ectopic or aberrant thymic tissues elsewhere in the neck.

In the past, the diagnosis was made on biopsy and surgery. With the introduction of high resolution ultrasound and MRI, the diagnostic algorithm has shifted from histopathology to radiological diagnosis alone.[8]

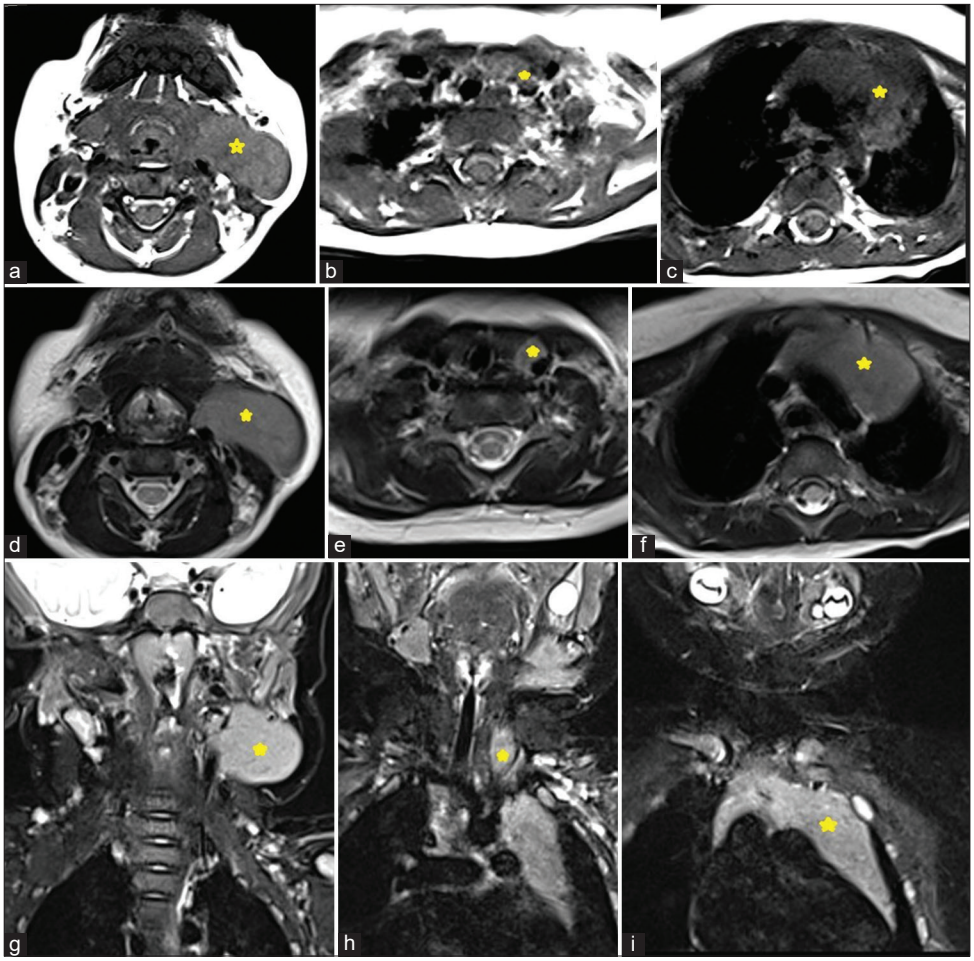

Figure 2: (a) Axial T1W image at the level of submandibular region showing a well-defined isointense lesion in the left submandibular region inferolateral to the left submandibular gland and anteromedial to the left sternocleidomastoid which is represented with a yellow *. (b) Axial T1W image at the level of the lower neck showing small isointense lesion in medial to the left carotid artery which is represented with a yellow *. (c) Axial T1W image at the level of superior mediastinum showing normal thymus which is isointense to muscle in anterior mediastinum. Signal intensity of both the lesions was same as that of normal thymus which is represented with a yellow *. (d) Axial image at the level of submandibular region showing well defined hyperintense lesion in the left side of neck, inferolateral to the left submandibular gland and anteromedial to the left sternocleidomastoid, which is represented with a yellow * (e) Axial T2W image at the level of the lower neck shows a well-defined hyperintense lesion medial to the left carotid artery, which is represented with a yellow *. (f) Axial T2W image at the level of superior mediastinum showing normal thymus which is hyperintense in anterior mediastinum. Signal intensity of both the lesions was same as that of normal thymus which is represented with a yellow *. (g) Coronal fat suppressed T2W images of neck at the level of the submandibular gland (g) and more anteriorly at the level of the thymus (h and i) showing well-defined hyperintense lesions, represented with yellow *, in the submandibular region at the angle of the mandible (g) and the left lower neck (h), whose signal intensity is identical to normal thymus gland in anterior mediastinum (i) which is represented with a yellow *.

CONCLUSION

Cervical thymus although a rare entity should be kept in the differential diagnosis of asymptomatic neck masses in children.[10] The unique imaging findings on ultrasound and MRI paralleling that of the normal thymus tissue should aid to confidently establish the diagnosis. The radiologist must be aware of the condition and the typical normal appearance of thymus which can obviate the need for biopsy or surgical intervention.

TEACHING POINTS

1. Aberrant cervical thymus should be included in the differential diagnosis of unilateral neck mass in children
2. High resolution ultrasound remains the first choice of imaging due to lack of ionizing radiation, ready availability, and non-sedative technique
3. The unique imaging features on ultrasound include well defined, hypoechoic lesion with multiple linear hyperechoic foci and septae, which parallels the echo-pattern of normal thymus
4. MRI shows the signal characteristics of the lesion similar to that of normal thymus in all the sequences and additionally helpful in identifying other lesions along the thymic tract
5. Radiologists should be aware of this condition to avoid unnecessary investigations and interventions.

MCQs

Q1. Ectopic thymus presents as
   a. Solid mass
   b. Cystic mass
   c. Either solid or cystic mass.

Answer Key: c

Q2. Normal embryological development of thymus
   a. Caudal and medial migration
   b. Caudal and lateral migration
   c. Caudal and posterior migration.

Answer Key: a

Q3. First investigation of choice in pediatric neck masses
   a. FNAC/biopsy
   b. Ultrasound
   c. MRI.

Answer Key: b

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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