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Right Ectopic Cervical Thymus-A Rare Case Report

Case Report

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Abstract

Cervical ectopic thymus is rare cause of solid neck masses and are the silent majority. Cervical ectopic thymus many times diagnosed incidentally and mistaken for neoplasm. Here we are reporting a case of 45 years old female presented with a history of slowly enlarging mass on the right side of the neck in paratracheal region. The patient complained of pain at the site of the enlarged mass. Thyroid function tests were within normal limits. On ultrasonography, right cervical ectopic thymus was revealed in the paratracheal region in relation to right side of thyroid gland.

Keywords: Neck mass; Ultrasonography; Ectopic Thymus; Neoplasm; Aberrant thymus; Thymus hyperplasia

Introduction

The thymus is first lymphoid organ, which grows substantially in infants [1,2]. Cervical ectopic thymus as neck lump is rarely considered in the differential diagnosis of neck swellings. It is, a prevailing anomaly identified incidentally at autopsy [3]. However, the presence of ectopic thymus tissue is acclaimed pathological essence along the embryological descending track in the neck [4]. Ectopic thymic tissue may come across anywhere along the path of descent of the thymopharyngeal ducts (e.g-cervical, retrocaval, posterior mediastinal) [5].

Ectopic thymic tissue adjoining to the thyroid gland is a rare entity and completely incidental finding either preoperatively or at autopsy [6]. Thymic remnants when found in the neck, they are unusual cause of patient presentation at the clinic and the diagnosis is difficult to accomplish in a pre-surgical walk up [7].

Here we report a case of adult female with ectopic cervical thymus and was an incidental finding which encountered in association with neck mass.

Case Report

A 45 years old female was referred for radiological scanning for the swelling in the neck.

Clinical Examination

A patient was afebrile with all parameters normal. She complained of the mass in the neck on the right side since last 4 months and was increased since last one month. On palpation of neck revealed soft, tender and fluctuating mass, which was approximately 3 to 4 cms in diameter; below the right angle of the mandible and anterior border of upper $1/3^{\rm rd}$ of right sternocleidomastoid muscle. It was difficult to portray exact outline of the neck mass. The overlying skin showed no discolouration and there was no intraoral swelling noted.

The patient was then scanned on ultrasonography, which revealed an enlarged lymph node on the right side. During scanning incidentally detected a mass (0.6x0.5x0.2cm) on right paratracheal region, which was situated between right submandibular gland and right side of thyroid gland (Figure 1).

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The mass showed homogeneous echo pattern similar to normal thymic tissue with normal vascularity (Figure 2) with no other abnormalities.

No other lymph nodes were palpable and were noticed on USG. The thyroid function test was within normal range. Hence, ectopic cervical thymus was identified. The patient was sent to the surgery department for further management of necrotic lymph node. Our case is unique in that ectopic cervical thymus was seen on the right paratracheal region. This has been rarely documented in the literature.

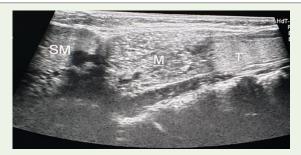


Figure 1: Ultrasonography of right side of neck showing the cervical ectopic thymus between the right submandibular gland and right side of thyroid gland..

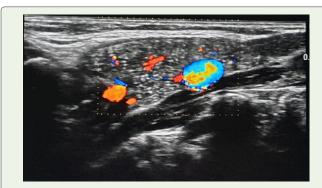


Figure 2: Ultrasonography of right side of neck showing normal echo pattern and vascularity in cervical ectopic thymus.

Discussion

Ectopic and accessory thymic remnants can be found anywhere along the course of migration of the thymo-pharyngeal duct from the $3^{\rm rd}$ and $4^{\rm th}$ branchial arches to the superior mediastinum. In case of ectopic thymus, the mediastinal thymus may be small orabsent[1]. In our case, it was absent.

Cervical ectopic thymus is an uncommon cause of neck lump and is usually described in sporadic case reports[3]. Tovi and Mares reviewed 68 reported cases of ectopic thymus in 1978 [3,8]. Ten years later in a collective review of 91 cases by Nowak et al, 76 presented as neck masses while the rest were mediastinal in location [3]. In our case, ectopic thymus was situated in neck on the right side of the thyroid gland. The majority of the cases are seen between 2 to 15 years of age, with a male preponderance [9]. In our case, it was incidentally detected in 41 years old female.

Ectopic thymic masses are located along the pathway of descent of the thymus. Hence it could be sited anywhere from the angle of the mouth or base of the skull to the superior mediastinum. These thymic vestiges are relatively common anomalies but since they are asymptomatic they are detected occasionally [10]. In our case, ectopic thymus was present on right paratracheal region, below the right angle of the mandible between right submandibular gland and right side of thyroid gland.

Cervical ectopic thymus is often asymptomatic with only 10% of patients being symptomatic in the form of pain or pressure symptoms like strider, dyspnoea, dysphagia and hoarseness of voice [11]. However, in our case the neck swelling was due to necrotic lymph node while the ectopic cervical thymus had normal echogenicity.

Most cervical thymus are unilateral and for unknown reasons are more commonly reported on left side and in male patients they have been known to occur as high as the mandibular angle and as low as the thoracic inlet and superior mediastinum. Thymic masses in the trachea, pharynx and at the base of skull havebeen reported [12]. In our case, also it was unilateral and presented on the right side in a female patient. It was high upto the right mandibular angle.

The characteristic appearance in an ultrasound scan is the finding of a remnant between the thyroid gland and the neck muscles with multiple linear structures and echogenic foci that give it a starry sky appearance [8]. Probably due to the presence of Hassall's corpuscles. This is however variable as it has been shown that the thymic parenchyma may sometimes be hypoechoic and less characteristics. In our study ectopic cervical thymus showed normal homogeneous echo pattern on USG with no any significant changes in the tissue; however at superior mediastinum thymic tissue was not traceable.

Embryological Basis

Knowledge of the embryologic development of the thymus is essential in understanding the pathogenesis of the aberrant / ectopic thymus [4]. The primordial thymus begins to appear early in the 6th week of fetal life from the ventral wing of the thirdpharyngeal pouch on each side of the most cephalad portion of the thymus are derived from the fourthpharyngeal pouch. The proliferation of endodermal calls within the outpouchings gives rise to paired solid structures. By the 7th week of generation, the thymic primordia lose their connections with the pharyngeal wall & join in the midline. A mesenchymal capsule surrounding the developing thymus and maintains the organ in close association with the partial pericardium. Together these structures descend to their final anatomic positions in the anterior mediastinum.

The medial caudal migration pathway forms the thymophalyngeal tract, which runs from the angle of mandible to the manubrium of the sternum bilaterally. Normally this tract involutes by the completion of development; however, thymic vestiges may persist anywhere along its course. Lymphocyte invasion occurs at 10 week of gestation, whereas subsequent endodermal regression from the Hassall'scorpuscle [12].

Cervical thymic anomalies may occur because of an arrest in medial caudal migration of thymic primordia and persistence of

INDIAN JOURNAL OF APPLIED RADIOLOGY

remnants of thymopharyngeal tract [8]. In case of an undescended thymus due to migrationarrest, only half of the normally bilobed thymus is present in the mediastinum. However, a normal chest radiography, which lacks the absence or diminution of thymic shadow, suggests cervical sequestration of thymic remnants. In addition, several reports have demonstrated the coexistence of the thymus and parathyroid glands in lateral cervical masses [8].

Conclusion

A cervical thymic remnant is a rare cause of neck masses; however, it must be considered in the differential diagnosis in neck masses. As they are common and rarely considered in the preoperative differential of neck mass. It is important to identify these thymic rests for what they are by their typical US appearance so that they will not be confused with other neck masses arising from the thyroid, para thyroid or lymphatic systems.

However, invasive surgical procedures may result in unhealthy problems and rise for developing immune dysregulation. If surgery is the choice in such patients, we recommend preoperative imaging studies to assess the presence of mediastinal thymic tissue.

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