

Ectopic thymic cyst in the neck

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Abstract

Ectopic thymic tissue in the neck is rarely reported in medical literature. This paper presents the case of a young female, who presented with a soft, fluctuating mass in the left side of her neck. Surgical excision revealed an ectopic thymic cyst. Ectopic thymic tissue may be an infrequent finding, but it should be included in the differential diagnosis of neck masses, especially in children. This case report is accompanied by a short review of the relative literature.

Key words: Thymus gland; Ectopic tissue

Introduction

Fewer than 90 cases of ectopic, aberrant or accessory thymus gland¹ have been reported in the medical literature according to the most recent review by Nguyen *et al.*,² thus it was thought that the following case was worth reporting.

Case report

A 23-year-old woman presented with a soft, mobile, non-tender mass in the left side of her neck, located at the anterior triangle. The mass had been present for about one year before she referred to us and it was asymptomatic, except that its size fluctuated with time. No other signs or symptoms were associated with the mass.

Her physical examination revealed no other pathological findings and her complete laboratory workup was within normal limits.

A neck computed tomography (CT) scan showed a mass in contact with the left carotid sheath, extending from the level of the hyoid bone to the entrance of the thoracic cavity (Figure 1).

Operation: Interestingly, on the day of the operation the mass had reached its minimum size, not being palpable. An incision at the level of the upper edge of the thyroid cartilage was made, parallel to the normal lines of the skin, over the sternocleidomastoid muscle. A mass was found anterior to the sternocleidomastoid, at the bifurcation of the common carotid artery. The mass was cystic in nature and consisted of tissue folded like cloth. It was elongated, with an extension to the entrance of the thoracic cavity. A second incision was made lower in the neck, to search for the end of the extension, but this continued into the thorax. Surgical removal was complete after opening the carotid sheath and the mass was ligated at the entrance of the mediastinum. It was not considered necessary to follow it there. The post-operative course was uneventful.

Macroscopically, two specimens, $5 \times 1.5 \times 1$ cm and $3.5 \times 1 \times 0.5$ cm were assessed. The sections of the largest one showed a cyst, with a thin fibrous wall and mean diameter of 2.5 cm, filled with a soft, whitish necrotic material.

Microscopically, the inner surface of the cyst had been destroyed and the squamous, non-keratinized epithelium found was replaced by an inflammatory infiltrate containing numerous cholesterol clefts (Figure 2, one arrow). The presence of thymic tissue in the walls of the cyst (Figure 2, two arrows), along with the pathognomonic presence of Hassall's corpuscles (Figure 2, arrowhead) identified the thymic origin of the cyst. In conclusion, the diagnosis was of a broken thymic cyst, that was completely excised.

Discussion

Thymus gland derives its origin from the third and, in some instances, the fourth pharyngeal pouch; it develops bilaterally early in foetal life and descends down the



FIG. 1

CT scan showing a mass in front of the left common carotid artery and the jugular vein, medially to the sternocleidomastoid muscle, at the level of the thyroid gland (arrowhead).

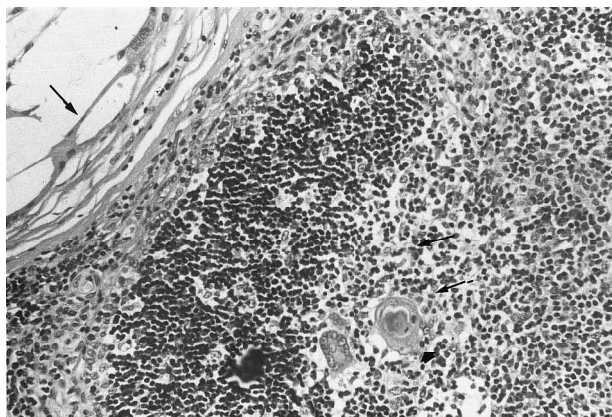


FIG. 2

The epithelium has been destroyed and replaced by an inflammatory infiltrate containing numerous cholesterol clefts (one arrow). Thymic tissue in the wall of the cyst (two arrows) and a Hassall's body (arrowhead) (H & E; $\times 200$).

neck during the sixth to eighth week of gestational life, where the two primordia fuse to form the gland, which then reaches its definitive position in the mediastinum, behind the sternum. The presence of thymic tissue in sites other than normal is characterized as ectopic or aberrant thymus.

It seems that the persistence of aberrant solid thymic rests in the neck is not such a rare condition, if we consider results of autopsies,³ but the absence of clinical manifestations is responsible for its not being diagnosed. On the other hand, cystic thymic tissue is more likely to be diagnosed, because of its clinical presentation. That was the case in the present report.

Clinically, in most cases, ectopic thymic tissue presents as a unilateral, asymptomatic neck mass, commonly in the left side of the neck.⁴⁻⁷ However, there are some reports connecting ectopic thymic tissue to respiratory distress^{8,9} and tracheal obstruction,⁹ to dysphagia,³ to infection and sudden enlargement due to upper respiratory tract infection,^{9,10} to laryngeal displacement, to myasthenia gravis¹¹ and to malignancy.^{12,13} The mass can be solid or cystic. The interesting thing in the present case is the fluctuating character of the lesion, which could not be associated to any other condition.

Moreover, the majority of reports concern infants and children, as this gland presents its relative maximum size at the age of two and its absolute maximum size at puberty. It seems that almost two thirds of cases concern patients in their first decade of life.¹⁴ Adult cases share just a small part of the literature. In Lewis⁹ review, only five out of 34 cases concerned adults older than 18 years and according to Guba,³ 75 per cent of patients with histologically proven thymic cyst were less than 20 years old at presentation. This was one reason we considered important to add the present case to the literature.

The pathogenesis of the ectopic thymus has not been fully clarified yet. Several theories have been proposed, including:

(a) Complete or partial failure of the unilateral gland to migrate to its final position. The presence of parathyroid gland in the mass supports the idea of non-descending tissue.^{8,10} Also, the fact that aberrant thymus is usually found in the normal pathway that the gland follows for its descent to the thorax is in favour of this theory.

(b) Sequestration of accessory cervical foci along the normal pathway of descent. In that case, there would be normal thymic tissue in the mediastinum, as most of it would have migrated.¹⁵

(c) Ectopic thymic gland, in the form of masses located in the pharynx, the trachea or the base of the skull.⁹

(d) Failure, after descent of the majority of the gland, of rostral fragments to involute, leading to separate accessory lobes or cords.

In our report the operative findings suggest that some fragments of the gland failed to descend in the mediastinum, while other fragments did and the tissue extension in the thorax stands as a proof for this.

As for the pathogenesis of the cyst formation, there is still controversy. According to Speer¹⁶ it may be due to embryological remnants in the neck, sequestration products in pathological involution, neoplastic process, degeneration of Hassall's bodies, mesenchymal elements.

Today there are two mainstreams in the theories about the pathogenesis of a thymic cyst. The first one relates it to acquired progressive cystic degeneration of Hassall's corpuscles, of unknown aetiology and the second to cystic changes in persistent unincorporated remnants of the thymopharyngeal duct.

Differential diagnosis includes all possible causes of neck masses according to age, starting from congenital cysts (bronchiogenic or of the thyroglossal duct), cystic hygroma, lymphadenopathy, thyroid and parathyroid gland lesions, lymphoma or other tumours of the area. However, it is interesting that the diagnosis of ectopic thymus has rarely been made pre-operatively.¹⁵

Possible complications include myasthenia gravis associated with ectopic thymic foci^{9,11} as well as the development of malignant thymoma in the ectopic tissue.^{12,13}

Surgical removal is indicated in symptomatic cases, in order to make a differential diagnosis from all the other causes of neck swelling. Also, the possibility of the prementioned complications presents sufficient reason to proceed to the excision of the mass. However, in infants and children it is of great importance to establish the presence of normal thymic tissue in the mediastinum before proceeding to the operation. This can be easily done with a chest X-ray.

Surgical removal in our case proved not to be difficult, as suggested by literature, and the patient will be followed to establish the absence of recurrence of any other related events.

Acknowledgement

We would like to thank E. Geropoulo for her help with the pathology for this paper.

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Dr S. Vlachou takes responsibility for the integrity of the content of the paper.
Competing interests: None declared.
