

PRESENCE OF THYMIC TISSUE IN THE ANTERIOR MEDIASTINAL FATTY TISSUE AND ITS SIGNIFICANCE IN THYMECTOMY FOR MYASTHENIA GRAVIS PATIENT: A CASE REPORT

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ABSTRACT

In 1970s, presence of thymic tissue in anterior mediastinal adipose tissue around the thymus was found. Here we report a case of ectopic thymic tissue in the mediastinum and the possible relevance of this distribution of thymic tissue outside thymus to the therapeutic yield of thymectomies in myasthenia gravis. A 30-year lady with myasthenia gravis (nonthymomatous) presented with difficulty in swallowing and breathing for the last 1 years. She was under medical treatment but with little improvement. She underwent extended thymectomy, after which an en bloc resection of the anterior mediastinal fat tissues from pericardium and pleura, including the thymus, was performed. Grossly the soft tissue specimen taken from near left lateral area of heart was fibrofatty tissue. Microscopically isolated thymic tissues were seen interspersed among the fatty tissues composed of mature lymphocytes, epithelial cells and few Hassall's corpuscles were observed. Thymus specimen was within normal histological limits. To ensure complete removal, the adipose tissue at the anterior mediastinum as well as the gross thymus should be removed. Thymic tissue incidence in individual locations was as follows: Retrothyroid, 3(6%); peritracheal, 5 (10%); retrotracheal, 1 (2%); right phrenic nerve, 2 (4%); left phrenic nerve, 14 (28%); right recurrent laryngeal nerve, 2 (4%); left recurrent laryngeal nerve, 2 (4%) and periaortic, 0. Trans-sternal thymectomy was found to be beneficial to all patients of mild-to-moderate myasthenia gravis with 70.2% patients showing improvement postoperatively.

KEYWORDS

Ectopic thymic tissue, Thymectomy, Mediastinum.

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INTRODUCTION: In 1970s, presence of thymic tissue in anterior mediastinal adipose tissue around the thymus was found, and centres advocated extended thymectomy, meaning an en bloc resection of the anterior mediastinal adipose tissue including the thymus for complete thymectomy.

Here we report a case of ectopic thymic tissue in the mediastinum and the possible relevance of this distribution of thymic tissue outside thymus to the therapeutic yield of thymectomies in myasthenia gravis. Klimek and colleagues reported the distribution of thymic tissue in the individual as; retrothyroid, 3(6%); peritracheal, 5(10%); retrotracheal, 1(2%), right phrenic nerve, 2(4%); left phrenic nerve, 14(28%); right recurrent laryngeal nerve, 2(4%); left recurrent laryngeal nerve, 2(4%) and periaortic, 0.^[1]

CASE REPORT: A 30-year lady with myasthenia gravis (nonthymomatous) presented with difficulty in swallowing and breathing for the last 1 year. She was under medical treatment but with little improvement. Thyroid functions were in normal range.

The titer of acetylcholine receptor antibody was 3.70 nmol/L.

NSS (Neuropathy Symptom Score) was done and it showed of incidence of early sensory neuropathy.

RNST (Repetitive Nerve Stimulation Test): Decremental response present in deltoid, trapezius, nasalis and oris muscle.

CECT Thorax: Thymus gland size was within normal limit.

She was operated on in the Department of Cardiovascular and Thoracic Surgery on July 2012 underwent extended thymectomy: A standard median sternotomy approach was used after which an en bloc resection of the anterior mediastinal fat tissue, including the thymus was performed. Dissection was performed from pericardium and pleura. The adipose tissues around the upper poles of thymus, around both brachiocephalic veins, and on the pericardium were resected meticulously. The borders of resection were the diaphragm caudally, the thyroid gland superiorly, and the phrenic nerves laterally.

Grossly the soft tissue specimen taken from near left lateral area of heart, was fibrofatty tissue measuring 4 × 2 × 1 cm. Microscopically isolated thymic tissues were seen interspersed among the fatty tissue. The thymic tissue is composed of mature lymphocytes and epithelial cells. Few Hassall's corpuscles were observed.

Thymus specimen was within normal histological limits.

DISCUSSION: The embryologic development of the thymus originates primarily from the ventral wing of the third pharyngeal pouch high in the neck in early foetal life when

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the fusion is complete the thymus and the inferior parathyroid lose their connection with the pharynx and migrate caudally in the anterior mediastinum.

The level of the thyroid gland is the most common site for ectopic thymic tissue. Hyperplastic or neoplastic changes can occur in ectopic tissues.^[2]

The cortex is composed mainly of lymphocytes, whereas the medulla with epithelial cells. Hassall's corpuscles are characteristic of thymus.^[3]

Klimek and colleagues reported the distribution of thymic tissue in the individual as; retrothyroid, 3(6%); peritracheal, 5(10%); retrotracheal, 1(2%), right phrenic nerve, 2(4%); left phrenic nerve, 14(28%); right recurrent laryngeal nerve, 2(4%); left recurrent laryngeal nerve, 2(4%) and periaortic, 0.^[1]

In the same study, the ectopic thymic tissue was found in 32 out of 50 cadavers (64%). In 10(20%), was accessible for the standard surgery, and in 22(44%) of them was not accessible for standard surgery as it was frequently located along phrenic nerves, especially on the left side.^[1]

The frequent presence of ectopic thymic tissue limits the efficacy for surgical treatment of myasthenia. To ensure complete removal, the adipose tissue at the anterior mediastinum as well as the gross thymus should be removed and even the cervical adipose tissues are removed at times.^[2] Myasthenia gravis treated medically has been reported to have remission rates as low as 15%. Thymectomy on the other hand has been reported to have clinical remission rates up to 80% and therefore has become the accepted mode of treatment. Many studies have reported greater improvement in female patients.^[4]

Behnam and colleagues did a study in the distribution of mediastinal ectopic thymic tissue in patient without thymic disease and found that 70.85% had ectopic thymus. Left pericardiophrenic being the most common site.^[5]

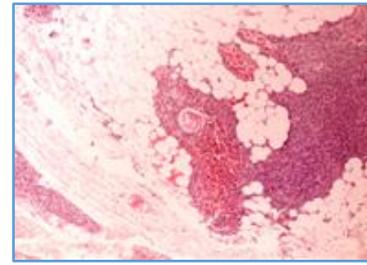
In another study, women benefitted more than the male patients from thymectomies and age older than 50 years and presence of thymoma were potential predictors for bad outcomes.^[6]

The probability of obtaining complete stable remission at 5 years after transsternal extended thymectomy was significantly higher in patients without ectopic tissue was observed by Ponseti and colleagues.^[7]

Trans-sternal thymectomy was found to be beneficial to all patients of mild-to-moderate myasthenia gravis, with 70.2% patients showing improvement postoperatively.^[4] Subsequently, this procedure was accepted as a standard method in patients with not only nonthymomatous, but also thymomatous MG in many centres.

Kirby J Scott reported a case of ectopic thymic tissue in a 13-month-old boy presenting as a neck mass.^[8]

The presence of thymic tissue in the mediastinum outside the lobes of thymus is evident. Hence, extended thymectomy where adipose tissue in anterior mediastinum and gross thyroid is removed is an excellent operative procedure for myasthenia gravis patients. It enhances the therapeutic yield of thymectomy.



Thymic tissues with Hassall's Corpuscles seen in the mediastinal adipose tissues

CONCLUSION: The embryologic development of the thymus at times result in tissues migrating caudally in the anterior mediastinum. The level of the thyroid gland is the most common site for ectopic thymic tissue. The ectopic thymic tissue located along phrenic nerves, especially on the left side is the most difficult area for standard surgery. Myasthenia gravis treated medically has been reported to have remission rates lower than those who went for thymectomy with remission rates up to 80% in the latter and therefore has become the accepted mode of treatment. Trans-sternal thymectomy was found to be beneficial to most patients. Subsequently, this procedure was accepted as a standard method in patients with not only nonthymomatous, but also thymomatous MG in many centres. The presence of thymic tissue in the mediastinum outside the lobes of thymus is evident. Hence, extended thymectomy where adipose tissue in anterior mediastinum and gross thyroid is removed is an excellent operative procedure for myasthenia gravis patients. It enhances the therapeutic yield of thymectomy.

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