

**CERVICAL ECTOPIC THYMUS:
A CASE REPORT AND REVIEW OF THE LITERATURE**

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ABSTRACT

Ectopic cervical thymic tissue is rarely reported in medical literature, but it should be included in the differential diagnosis of neck masses, especially in children. The authors present the case of a young male with a soft right mid-cervical mass. The patient underwent complete excision of the lesion and histological examination showed an ectopic thymic cyst. The embryological development, clinical presentation and management of ectopic thymic lesions are discussed together with a review of the literature.

Keywords: ectopic cervical thymus, neck masses, children.

INTRODUCTION

Ectopic cervical thymic tissue is a rare cause of neck masses. It may be found at any level of the pathway of normal thymic descent, from the angle of the mandible to the superior mediastinum. Seldom considered in the differential diagnosis of neck masses, its presence is often revealed by

pathologic examination of an excised specimen. Adolescents and children comprise the majority of the typically asymptomatic patients. As such, the presence of a mediastinal thymus must be confirmed prior to the surgical resection of a neck mass to prevent inadvertent total thymectomy with possible negative effects on the developing immune system. We report the clinical presentation, diagnostic evaluation and therapeutic management of one case of ectopic cervical thymic tissue, accompanied by a short review of the relative literature.

CASE-REPORT

A 6-year-old child was referred to the Otolaryngology Clinic of S.Orsola-Malpighi Hospital, Bologna, Italy, for the evaluation of a large right mid-cervical mass with the suspicion of lymphadenopathy. The mass was asymptomatic and was noted by the parents in conjunction with a previous upper respiratory tract infection. Although the patient followed an antibiotic therapy regime, the lesion failed to change significantly. The family history and past medical history were unremarkable. Physical examination revealed a soft, mobile and poorly circumscribed neck mass of about 4 cm in diameter. No overlying skin changes or pits were apparent, and the infant's neck position and mobility were not limited by the presence of the mass. Endoscopic laryngoscopy revealed normal laryngeal mobility and an integral pharyngeal mucosa with only a slight medialization of the pharyngeal wall at the pharyngeal-epiglottic fold. An ultrasound of the neck

suggested a multi-loculated cystic mass, while an MRI indicated a right-sided, anteriorly-located 6 cm cervical mass beneath the sternocleidomastoid muscle, lateral to the carotid sheath and inferior to the parotid gland. The mass also presented a slight extension into the retropharyngeal space (Fig.1-2). Following the clinical and MRI findings, surgical excision revealed a multi-loculated cystic mass (Fig.3). In this case, the blunt dissection technique was employed to conserve the integrity of the external capsule of the cyst, respecting the integrity of the pharyngeal-laryngeal-tracheal wall and of the surrounding nervous and vascular structures. The final pathology report confirmed the presence of an ectopic cervical thymic cyst. The child's post-operative course was unremarkable, without neurological defects or injuries. Furthermore, clinical and echographic six-month follow-up was negative.

DISCUSSION

Cervical thymic anomalies may occur as a consequence of an arrest in descent during the ninth week of embryonic growth, a sequestration of thymic tissue during descent or a failure of involution [1-4]. Of the more specific origins proposed [5], the two favored theories for the development of cervical thymic cysts are the persistence of thymopharyngeal ducts (congenital) and the degeneration of Hassall's corpuscles within ectopic thymic remnants (acquired) [6].

A cervical thymic cyst is usually a soft, unilocular or, more frequently, multilocular mass, 1 to 15 cm in width. The lesion occurs on the left side in 60-70% of patients, on the right side in 20%-30% of patients, and in the midline or the pharynx in the remaining 5-7% of patients [7,8]. Nests of thymic tissue may be found anywhere along the path of descent of the thymic primordia from the angle of the mandible to the mediastinum. They are usually located anteriorly and deep to the middle third of the sternocleidomastoid muscle, adhere posteriorly to the carotid sheath and often extend into the retropharyngeal space [2,9]. Approximately 50% of all cervical thymic masses may be continuous with the mediastinal thymus by direct extension or by connection to a vestigial remnant or a solid cord [3].

The diagnosis of cervical thymic tissue is rarely made preoperatively [6,9,10,11,12]. An estimated 80-90% of these lesions are asymptomatic, only 6% have symptoms, such as stridor, dyspnea and/or dysphagia due to compression of the trachea and/or esophagus [7,13]. The ectopic thymic tissue appears to cause symptoms relatively more common in children than in older patients with over 50% of children cases experiencing respiratory and feeding difficulties [10]. Airway compromise and feeding disturbances are caused by compression of the parapharyngeal space or vagus nerve and by the rapid expansion of the cyst attributed to fluid accumulation or to hemorrhage. Some patients report small fluctuations in lesion size, whereas others experience rapid enlargement after minor trauma, vaccination or

upper respiratory tract infection [13]. Routine diagnostic evaluation for a neck mass includes a thorough history, physical examination, ultrasound of the neck and chest X-ray. Bloodwork, including a CBC with a differential, is usually obtained, and an empiric course of antibiotics is often utilized.

CT scan findings of an ectopic thymic mass usually reveal a homogenous mass intimately attached to the carotid sheath. A useful additional sign is a lack of mass effect on the airway or vessels when the mass is present during infancy. MRI can help in the diagnosis of ectopic cervical thymus as its tissue plane definition is often able to demonstrate a connection between the cervical mass and the mediastinal thymus or to determine if the mass's density is similar to normal thymic tissue. In thymic cysts, an irregular enhancement within the mass is possible due to the presence of septal architecture.

The differential diagnosis of a cervical mass in children includes thyroglossal duct cyst, brachial cleft cyst, cervical lymphadenopathy, benign tumors (dermoid, epidermoid, hemangioma and lymphangioma) and malignant tumors (lymphoproliferative, soft tissue sarcomas and other metastatic lesions) [4,12].

For both diagnostic and therapeutic reasons, surgery is the treatment of choice for most neck masses, including cervical thymic lesions [4,6,12]. At this time, there is no preoperative test that can accurately identify a neck mass as cervical thymic tissue, diagnosis being made from the histology of

the resected specimen. Most cervical thymic lesions may be adequately excised via a transverse cervical incision and, even in the presence of a caudal extension, may be removed from the thoracic inlet by blunt dissection without the need for a sternotomy. At surgery, the mass may be found to be adherent to surrounding structures, such as the carotid artery, the jugular vein and/or the vagus, hypoglossal, phrenic and recurrent laryngeal nerves. Great care must be taken during the surgical procedure as to avoid rupture of innominate vessels and adjacent structures in the high mediastinum. In most cases, removal by blunt dissection is all that is required, and complete excision is curative. There has been no reported recurrence and long term prognosis is excellent.

An accurate evaluation of the presence of normal thymus should be undertaken prior to the surgical removal of a cervical mass. If a normal thymus is not present, removal of cervical thymic tissue may leave the patient athymic. This would pose no problem in the adult, but in a child dependant on thymic tissue for immunological function, long term observation is strongly recommended.

CONCLUSION

Ectopic cervical thymus cysts are uncommon but should be included in the differential diagnosis of neck masses, especially in the young. Such anomalies are rarely diagnosed pre-operatively and can be easily confused with other neck lesions. Once diagnosed, surgery is the definitive treatment

if the mass is symptomatic and cosmetically unbecoming. Prior to surgery, the presence of a mediastinal thymus should be confirmed to prevent the risk of a total thymectomy. The prognosis after removal of ectopic cervical thymus is excellent and no cases of recurrence have been reported.

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