Ectopic Cervical Thymic Cyst In Elderly: A Case Report With Review Of Literature

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Citation

Abstract
Cervical thymic cysts are rare lesions, and in adults, they are extremely rare. We are reporting a case of a 58-year-old female with an asymptomatic anterior cystic neck mass, clinically diagnosed as a lymph cyst that was surgically excised. She was diagnosed as having an ectopic cervical thymic cyst based on histopathological findings. To the best of our knowledge, our patient is the oldest reported case of cervical thymic cyst to date. As malignant change has been reported in adult cervical thymic cysts, it should be considered in the differential diagnosis of cystic masses in the neck of adults, followed by complete surgical removal and histopathological confirmation for definitive diagnosis. A case report with a review of the literature along with relevant embryogenesis and pathogenesis has been presented in this article.

INTRODUCTION
Ectopic cervical thymic cysts in adults are rare(1,2). Most of the cases have been reported in the paediatric age group. Their prevalence is less than 1% of all cervical masses, and they are usually noted in childhood(1). Being uncommon, they are rarely included in the clinical diagnosis of cervical cystic masses and are misdiagnosed by surgeons as branchial cysts, lymphatic malformations, epidermoid cysts, dermoid cysts or thyroglossal cysts, lymphadenitis or neoplastic masses(3). Malignancy in thymic cysts, though rare, has been mentioned in literature (4,5). Therefore the surgeon should be aware of this possibility, and complete surgical removal is the treatment of choice, followed by thorough histological examination to ensure that a possible malignant change is not overlooked.

Histopathologically, thymic cysts are diagnosed by the presence of remnants of thymic tissue, represented by Hassall’s corpuscles, in the cyst wall.

The purpose of publishing this case is to acknowledge the rarity of an ectopic thymic cyst in the cervical region in an elderly female of 58 years old, this being older than any patients currently reported in the literature, and to discuss the embryology and pathological features of this rare entity.

CASE REPORT
A 58-year-old female with history of slowly growing painless cystic mass, first noticed 3 years ago on the left side of the anterior triangle of neck in the cervical region, was admitted to our outpatient department of Surgery. A clinical diagnosis of a lymph cyst was considered. Intraoperatively, the cystic mass was found to be anterior to the sternocleidomastoid muscle and did not extend to mediastinum. Patient underwent excision biopsy. We received an ovoid cystic specimen measuring 3x2 cm. Upon cutting it open, the cyst was found to be unilocular with clear serous fluid and smooth luminal surface.

Histopathological examination showed a cyst wall lined by flat to cuboidal lining [fig.1]. The wall was composed of fat and fibro-collagenous tissue showing focal aggregation of lymphoid cells with Hassall’s corpuscles inside [fig.2]. Based on these classical findings, the diagnosis of ectopic thymic cyst of the cervical region was given.
Figure 1
Figure 1: Shows cyst wall lined by flat to cuboidal lining and composed of fibrocollagenous.

Figure 2
Figure 2: Cyst wall shows lymphoid aggregate with Hassall’s corpuscles within it (arrow).

DISCUSSION
Thymic cysts are rare lesions of the neck and anterior mediastinum (3). They are embryonic remnants arising from the endoderm of the third pharyngeal pouch along the course of thymic migration caudally in the neck or the anterior mediastinum, which may result in cervical masses in children that are rarely detected in adults (6). Early in development, this pouch is cut off from the pharyngeal wall. It consists of a thinner cranial part, which forms the cervical part of the thymus, and a broader caudal part, which enters the thorax in the anterior mediastenium (7). Eventually, the cranial part gets regressed. Failure to do so may later lead to the formation of a thymic cyst. Thymic cysts are more common in children, in contrast to ectopic cervical thymus, which is more common in adults (8).

Thymic cysts are of two distinct types, the most common of which are unilocular thymic cysts, which are of developmental origin as described above and can be found anywhere from the angle of the mandible to the manubrium sternum (9). They are located in the neck more often than mediastinum (10). The lining epithelium can be flattened, cuboidal, columnar or rarely squamous. Another type of cyst is the acquired type, which is multilocular and most frequently located in the mediastinum (11). It is by definition multilocular and is the result of an acquired cystic dilatation of medullary duct epithelium-derived structures, induced by an acquired inflammatory reaction (11,12). It is always accompanied by inflammation and fibrosis. These may be associated with neoplasm, most commonly thymoma or thymic carcinoma, lymphoma, or germ cell tumor (11).

In our case, the cyst was unilocular, located in the neck along the path of descent, with flattened to cuboidal lining and wall showing lymphoid aggregate with Hassall’s corpuscles. There was no evidence of inflammation or fibrosis indicating its developmental origin.

The histological diagnosis of a cervical thymic cyst is made if thymic epithelial elements or, more commonly, Hassall’s corpuscles can be found adjacent to the cyst cavity. These histopathological features of thymic cysts are diagnostic (6). Cholesterol granulomata can also be identified (13,14).

Clinically, a patient with a cervical thymic cyst complains of a painless, enlarging mass in the neck. In adults, because thymic cysts are a rare cause of benign neck masses, they are usually not considered in the differential diagnosis preoperatively (1). This is because various types of cysts that originate in embryonal remnants may be observed in the neck. Among these, branchial cleft cysts and thyroglossal duct cysts are more commonly observed, whereas thymic cysts are rare (1,3,6,15). In older age groups, thymic cysts at mediastinal locations are more common than cervical thymic cysts (16). In the mediastinum, they represent 1-2% of all mediastinal masses, and in the neck, their prevalence is less than 1% of all cervical masses (1,17). Age at presentation ranges from the neonatal period to adulthood. Most cases are asymptomatic; few cases are associated with symptoms of dysphagia or dyspnoea, especially in children (16,18). In the literature, a majority of cases have been reported in the paediatric age group ranging from newborn infants to 12 years (18,19). They usually present in their 1st decade after the age of 2 years, possibly because the thymus attains its greatest development before puberty (14). Most cases have been described in the age group of 4-7 years, fewer in younger patients (3,18,20,21, 1,22,23). In adults, Ertugrul has mentioned six patients with thymic cysts diagnosed in
the anterior mediastinum with ages ranging from 34 to 64 years (mean age, 46 years) and equal sex predilection (17). In a case series of cervical thymic cysts, the mean age reported was 36 years with a male/female ratio of 4:5 in adults (1). Two cases of multilocular cervical thymic cysts in adults presenting as asymptomatic swellings in the neck, possibly as an acquired disease, have been reported (24). To the best of our knowledge, our patient is the oldest reported to date for an ectopic cervical thymic cyst.

Because of the high incidence of pathological changes, surgical removal of the ectopic thymus is the treatment of choice and has been performed in all reported cases (3). Malignancy in adult thymic cysts can occur, though extremely rare, and very few cases have been reported in literature. The first documented case was reported by Leong in 1984 as a squamous cell carcinoma arising in a thymic cyst (4). Moran et al. reported 3 cases of primary carcinoma arising in multilocular thymic cysts in the neck in 2004 (5). Another case of incipient adenocarcinoma in the thymic cyst of a 38-year-old woman was reported in 2008 by Morrisi (25).

CONCLUSION

Cervical thymic cysts are rare lesions seen in the paediatric age group. They are very rare in adults. The possibility of a cervical thymic cyst should be considered in the differential diagnosis of cystic masses in the neck in elderly patients. Also, rarely, malignant transformation has been reported in these cystic lesions in adults. Treatment is complete excision followed by histopathological confirmation.

References

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