Case Report

Cervical Thymic Cyst

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Introduction

Thymic cysts are among the rarest cysts found in the neck [1]. Rarely is cervical thymus a symptomatic cause of lateral neck mass. A spectrum of abnormalities of the thymus is attributed to the embryonic cervical descent of the thymic primordium. Sequestered cystic cervical thymus is found along a normal path of descent, with or without parathyroid glands. It is a cystic version of accessory cervical thymus and may have a fibrous band or a solid thymic cord connection to the pharynx or mediastinum [2]. Due to the rarity of this cyst a preoperative diagnosis is seldom achieved. The purpose of this article is to report a rare form of cervical cyst and to discuss the embryogenesis of this lesion.

Case Report

A 23 year old patient presented with complaints of swelling in the right side of the neck of one month duration. The swelling was insidious in onset, gradually increasing in size without any obstructive symptoms. There was no history of change in voice or difficulty in swallowing. Examination

revealed a 5x4 cm soft cystic swelling, oval in shape, with diffuse margins, part of it being beneath the right sternocleidomastoid muscle. The swelling was non-compressible, not translucent and increased on Valsalva manoeuvre. On indirect laryngoscopic examination, a smooth bulge was noted in the posterior wall of oropharynx extending downwards to the hypopharynx pushing the larynx towards the left and obscuring the visualization of right vocal cord. FNAC showed numerous cholesterol crystals, lymphocytes, granulocytes and few macrophages in a serous background suggestive of branchial cyst. CT scan showed a large hypodense, multi-loculated non enhancing mass in the prevertebral space on the right side extending from C3 to C6 vertebra, medially the hypopharynx was compressed without invasion of the fat planes or destruction of soft tissues, displacing the great vessels laterally and extending up to the glottis (Fig 1). Excision of the mass was done under general anaesthesia. Per-operatively it was found to be a cystic mass with extension in the form of a cord to the mediastinum. It was resected completely and the mediastinal extension was transected at the lower most limit and excised. Histopathology showed multilocular thymic cyst lined by flat cuboidal and at places compressed squamous epithelium. Intervening septum was of variable thickness and composed of fibrocollagenous stroma with presence of lymphocytic infiltrate especially close to the lining epithelium of cyst. Also seen within the septae were islands of thymic tissue (Hassall's corpuscles, lymphocytes) (Fig. 2). The content of the cyst was a homogenous eosinophilic material with some of it having

Fig. 1 : CT scan shows a large hypodense, multi-loculated non enhancing mass in the prevertebral space distorting the right pyriform sinus, extending under the sternocleidomastoid muscle on the right side

Fig. 2 : Section from the wall of multilocular cyst shows collection of lymphocytes, Hassall's corpuscle (thymic tissue) and areas of fibrosis.

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fresh haemorrhage as well as inflammatory changes. Section taken from the extension of the cyst to mediastinum showed thymic tissue with presence of thymocytes, Hassall's corpuscles and lymphoid tissue.

Discussion

Thymic cysts are rare and may be found in the neck or mediastinum. Till date, less than 90 cases of ectopic, aberrant or accessory thymus gland have been reported in literature. Speer [3] while considering cyst formation within mediastinal thymic tissue, postulated many etiologies. These included embryonal epithelial remnants, degenerating Hassall's corpuscles, infectious and inflammatory product and neoplastic, hyperplastic and involutional changes of the epithelial, lymphoid, or connective tissue and vascular elements of the thymus. Two dominant hypotheses seem to have emerged. One relates the pathogenesis of cystic cervical thymus to acquired progressive cystic degeneration (of unknown etiology) in Hassall's corpuscles and the epithelial reticulum of the thymus [4]. A second body of opinion favours cystic change in persistent unincorporated remnants of the thymopharyngeal duct. The latter hypothesis is well accepted and most consistent with the developmental anatomy of the thymus. Zarbo has proposed a unifying scheme and nosology for deciphering the various reported types of solid and cystic cervical thymic abnormalities of normal embryologic migration.

The cervical thymic lesions include the following categories distinguished by anatomic location and the nature of the thymic gland tissue.

Accessory cervical thymus : Solid cervical thymic tissue is sequestered from the main gland, along the normal descent path, with or without parathyroid. Previous terms include aberrant, ectopic, undescended, persistent, or accessory thymus:

Cervical thymic cyst: Sequestered cystic cervical thymus is found along a normal path of descent, with or without parathyroid glands. It is a cystic version of accessory cervical thymus and may have fibrous band or a solid thymic cord connection to the pharynx or mediastinum.

Undescended cervical thymus : This occurs when a solid lobe of thymus fails to descend entirely, with or without a parathyroid complex. It differs from accessory cervical thymus in that only half of the normally lobbed thymus is present in the mediastinum, conceivably it may also become cystic.

Persistent thymopharyngeal duct cyst: This is the same as undescended cervical thymus; however, the thymic duct is cystic. The thymus is solid, with or without parathyroid complex, and probably represents undescended thymus. A variant would be the cervical

cystic duct leading to the mediastinal thymus.

Persistent thymic cord : This is the cervical prolongation of a solid thymic cord which is continuous with the mediastinal thymus. The cystic variant may overlap with the histology and clinical appearance of the cervical thymic cyst if a true connection to mediastinal thymus cannot be documented.

Cervical extension of mediastinal thymus : This appears as low midline solid thymus at the thoracic inlet due to incomplete mediastinal descent. It may transiently present with increased intrathoracic pressure.

Ectopic thymus : This is the rare, solid thymic tissue in abnormal locations, eg., in the pharynx, trachea, or base of skull [5,6]. Ectopic cervical thymic cyst has not been reported.

The increasing number of cervical thymic cysts reported in the last few years probably reflects greater awareness of this condition among pathologists. It is also possible that in the past, many cases of thymic cyst had been missed and diagnosed as brachial cleft cyst because of inadequate sampling of the specimen. The frequent atrophic condition of the thymic remnants may require sampling from various portions of specimen before a diagnosis of thymic cyst could be rendered [7].

Clinically, in most cases, cervical thymic lesions present as a unilateral asymptomatic neck mass, commonly on the left side of the neck and 75% of patients present before 20 years of age. Differential diagnosis includes branchial cyst, thyroglossal cyst, 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 preoperatively [8].

References

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