



Ectopic thymic cyst in neck: a case report and review of literature.

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Abstract

Cervical thymic cyst is not common pathology encountered in medical literature. It should be included in the differential diagnosis of neck masses especially in children. Our case presentation is of a 10years male child with a left upper cervical soft mass. The patient underwent complete excision of lesion and histological examination showed an ectopic thymic cyst. The diagnosis of thymic cyst is not possible prior to histological examination. Therefore in children the disorder should be considered as a differential diagnosis of neck tumors in children; thought it is rare.

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Introduction

Ectopic cervical thymic tissue is a rare cause of neck masses. It may found at any level of the pathway of normal thymic descent, from the angle of mandible to the superior mediastinum. Seldom considered in the differential diagnosis of neck masses; its presence is often revealed by pathological examination an excised specimen only. Children and adolescent comprises the majority of typically asymptomatic patients. Due to rarity of this cyst a preoperative diagnosis is seldom achieved. [1]

The purpose of this article is to report a rare form of cervical cyst and to discuss short review of relative literature.

Case: 10years old male child referred to the clinic for evolution of left upper cervical mass since 5-6 months. The swelling was soft to cystic non reducible, situated at angle of mandible 4x3 cm. The mass is non tender, soft with diffuse borders and smooth external surface. No overlying skin changes were apparent.

On clinical examination the lesion diagnosed with **d/d** as Branchial cyst, cystic hygroma and lymphadenopathy. Although the patient followed an antibiotic therapy regime, the lesion failed to change significantly. Family history and past history were unremarkable. Endoscopic examination revealed no any abnormality. The ultrasound of neck suggest encapsulated cystic (lesion filled with echoes seen in left submandibular region) mass 25x20mm with diagnosis of branchial cyst. All other neck structure was normal. By incision through the neck cyst was dissected entirely. Surgical specimens send for histopathology. Grossly: Irregular soft tissue mass 5x4x3cm,

lobulated cut section showed multiple small cystic areas (Multilobulated cyst) with necrotic.

Microscopically showed multiple cystic spaces lined by columnar squamoid epithelium. Thymic tissue with Hassall's corpuscles was found in the wall of cysts fig. which gives origin of cyst final histological diagnosis was multi cystic thymic cyst.



Fig. 1: Thymic cyst showing multiple lobulated mass externally

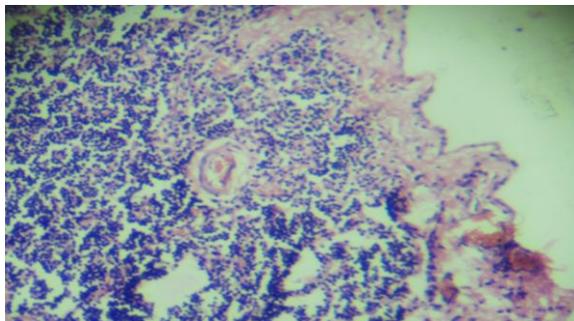


Fig.2: Cyst wall lined by columnar epithelium with lymphocytes

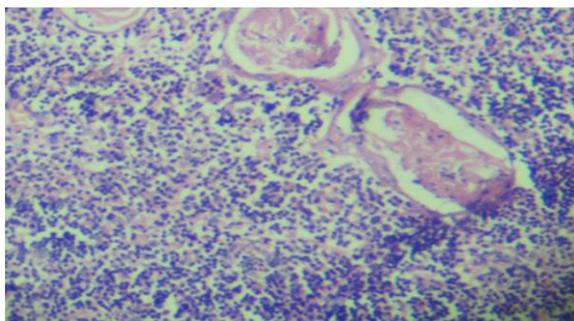


Fig.3: lymphoid tissue with Hassall's corpuscles

DISCUSSION

The thymus is embryologically derived from the ventral sacculation of 3rd pharyngeal pouch during the 6th week of development. Formed paired thymic buds on each side begin to migrate caudally to form a thymopharyngeal duct. Migration continues and the duct becomes separated from the pharynx. Cellular proliferation gives rise to paired solid masses by 8th week, which descend into mediastinum where they fused to form bilobuled thymus. During 3rd fetal month, cellular differentiation and continuing proliferation separate cortex from medulla [2]. The epithelial cords persists as the branching canalicular structures which are seen in cross section as Hassall's corpuscles. Nests of thymic tissue may be found anywhere along the path of descent from the angle of mandible to the mediastinum. [3]

The two favored theories for the development of cervical thymic cyst are persistence of thymopharyngeal tract (congenital) and degeneration of Hassall's corpuscles within ectopic thymic remnants (acquired) [4]. The presence of thymic tissue in sites other than normal characterized as ectopic or aberrant thymus. Thymic cysts are uncommon lesions and approximately only 150 cases have been reported [5]. However asymptomatic ectopic thymic tissue in neck is not such rare condition if we consider results of autopsies but the absence of clinical manifestation is responsible for it's not being diagnosed [6]. On the other hand, cystic thymic tissue is more likely to be diagnosed because it's clinical presentation (7). That was the case present report. Cervical thymic cyst is usually a soft unilocular or more frequently multilocular 1-15cm in size. The lesion occurs on the left side in 60-70% of patients, on the right side in 20-30% of patients and in midline or the pharynx in remaining 5-7% of patients [8, 9]. Amongst 80-90% of the patients are asymptomatic & have only painless swelling [8]. Respiratory symptoms such as dyspnoea, hoarseness, stridor and dysphagia are reported in 6-13% of patients [10]. The ectopic thymic tissue appears to cause symptoms relatively more common in children than in older patients with over 50% of children

cases presenting with respiration and feeding difficulties [11].

The differential diagnosis of cervical mass in children includes thyroglossal duct cyst, bronchial cleft cyst, cervical lymphadenopathy, benign tumors (dermoid, epidemoid cysts, haemangioma and lymphangioma) and malignant tumors (lymphoproliferative lesions; soft tissue sarcomas and other metastatic lesion) [12, 13]

Histological investigation of the excised specimen is the only definitive means of diagnosis. Macroscopically a thymic cyst is a soft, unilobular or more frequently multilobular mass. It is commonly elongated with one or both ends tapered to tract or cord. The cystic fluid may be clear or yellowish, brown or even purulent [14].

Microscopically show the presence of thymic tissue remnants with the pathogenic Hassall's corpuscles within the cyst wall. Lymphocytes, cholesterol crystals, giant cells, histocytes, inflammatory cells and naemogiderin have also been described. The cyst wall lining may be spindle, cuboidal, columnar, stratified, pseudostartified, ciliated or nondiated. Malignant transformations have not been reported in children. [15]

The cyst may adhere to surrounding structures such as vagus nerve, carotid artery, jugular vein, phrenic hypoglossal and recurrent laryngeal nerves. The preoperative differentiation of thymic cyst from other cystic masses in neck was different. For example second bronchial cleft cysts have similar physical exam and radiographic findings to thymic cyst. Bronchial cleft cyst tend to occur more commonly in the upper portion of neck where as thymic cyst tend to occurs more frequent in inferior portion of neck.

Thymic cysts are also difficult to distinguish from lymphatic malformation. Their presentation differs however, in the 90% of lymphatic malformation, occurs in patients less than 2years of age, where as thymic cysts occurs most commonly between 2-13years. Further 50% of thymic cysts have mediastinal

connection while this occurs only 5-10% of lymphatic malformation [15]. The only definitive diagnostic test for thymic cyst is histological examination. This will reveals Hassall's corpuscles and or cholesterol granuloma one of which required for diagnosis. The malignant transformation has been reported in adults but not in children, possibly due to fact that a thymic cyst contains no active solid thymic tissue. There has been no report of recurrence in a child.

Conclusion

Ectopic cervical thymic cysts are uncommon but should be included in the differential diagnosis of neck masses especially in the neck. Such anomalies are hardly diagnosed preoperatively & can easily confuse with other neck lesions. The only definitive diagnostic test for thymic cyst is histopathological examination. The prognosis after removal of ectopic cervical thymus is excellent and no cases of recurrence have been reported.

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