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Case Report

Pathology Section

Ectopic Thymic Cyst Presenting as Lateral Neck Mass in a Young Boy: A Case Report

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ABSTRACT

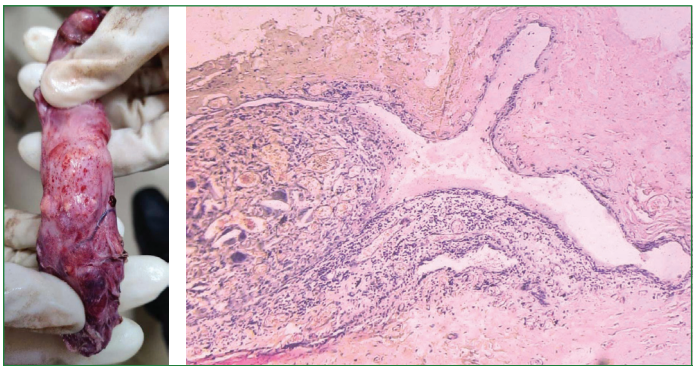
The thymus is derived from ectoderm and endoderm of third pharyngeal pouches. It descends down along the cervical region to anterior mediastinum and involutes by increasing age. The ectopic thymic tissue in neck is a rarity and few mentioned in literature are described as sporadic case reports. Ectopic thymic cyst accounts for less than 1% of all cystic neck masses in children and even more rare in adults. Worldwide literature states that there are about 150 cases of congenital ectopic thymic tissue. It can occur anywhere along the path of its descent, from mandible to mediastinum. This entity being unusual is infrequently considered in the differential diagnosis of cervical cystic masses. Histopathology forms the mainstay in confirmation of diagnosis. Here, we report a case of 12-year-old boy who presented with right-sided neck mass. The excised specimen revealed presence of lymphoid tissue with Hassalls corpuscles in the wall of cyst. This hallmarking feature is the pointer to arrive at a correct diagnosis of thymic cyst and to exclude its closest differential at this anatomical location including branchial cyst, cystic hygroma, lymphadenopathy and epidermoid cyst. The literature is reviewed and differentials of ectopic thymic cyst in cervical region are being discussed.

Keywords: Benign cystic neck lesions, Branchial cyst, Cervical lymphadenopathy, Cystic hygroma, Thymopharyngeal cyst

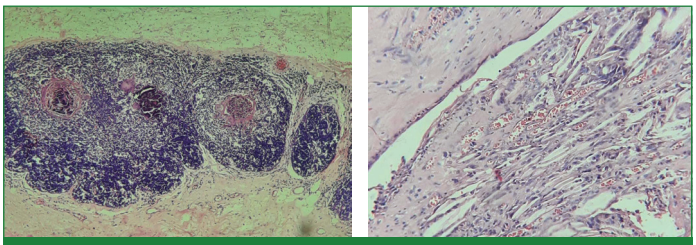
CASE REPORT

A 12-year-old school going boy attended Ear Nose Throat (ENT) Outpatient Department with parents. The history provided was of a swelling on right side of neck noticed since birth. Initially it was, 2×1 cm which gradually increased over a period of 12 years to attain the present size of 4×3 cm. There was no history of associated pain. No history of swellings in any other location in body. On examination, it was soft, mobile, non tender, soft swelling with normal overlying skin. Ultrasonography (USG) revealed a hypoechoic mass suggestive of benign cystic lesion. The Computed Tomogrphy (CT) scan showed 7×3×3 cm enhancing thick walled cystic lesion in right carotid triangle posterior to sternocleidomastoid muscle and extending up to the confluence of right and left brachiocephalic vein in anterior mediastinum abutting the thymus. Radiological impression was cystic hygroma, thymic cyst.

Complete resection of the mass was done and we received an excised specimen comprising of elongated mass of size 10×4×3 cm (Gross: [Table/Fig-1]). Externally, it was greyish pink, soft, nodular. The head end on cutting was solid cystic that yielded around 1ml of shiny clear fluid and thick whitish pink solid area. The other narrow end consisted of elongated cord like tubular structure. On histopathology examination, the mass revealed an elongated cystic structure lined by single layer of flattened to attenuated squamous epithelium [Table/Fig-2]. The underlying wall showed fibrocollagenous tissue along with foci of thymic tissue which were seen as variable sized lymphoid follicle with Hassall's corpuscles in the centre. At places, the lymphoid tissue revealed degenerated Hassall's corpuscles [Table/Fig-3]. The wall of the cyst also displayed plenty of cholesterol crystals, sheets of histiocytes and numerous foreign body giant cells resembling cholesterol granuloma [Table/Fig-4]. The histopathological diagnosis rendered was thymic cyst.



[Table/Fig-1]: Elongated tubular, greyish pink 10×4×3 cm mass. Cut surface solid cystic, pinkish white, soft in consistency (Gross).
[Table/Fig-2]: Cyst wall lined by flattened squamous epithelium. The wall focally shows foreign body cells and cholesterol clefts (H&E; 10X). (Images from left to right)



[Table/Fig-3]: The cyst wall shows numerous lymphoid follicles with Hassall's corpuscles (H&E; 20X).
[Table/Fig-4]: Sheets of histiocytes and foreign body giant cells (H&E; 40X). (Images from left to right)

DISCUSSION

Cervical thymic cyst is a rare abnormality that accounts for 0.3% of all congenital neck cysts in children [1,2]. Till date very few cases of congenital ectopic thymus have been described in the world literature [3]. The ectopic thymic cyst can be present along the normal path of normal thymic descent, from angle of mandible to superior mediastinum. These unusual lesions clinically present in the 1st decade of life [4].

The thymus gland develops mainly from ventral part of paired 3rd pharyngeal pouch and occasionally from 4th pouch in neck. It descends down as thymopharyngeal tracts along the course of carotid sheath to final position in superior mediastinum during 6th to 9th weeks of gestation to form bilobed thymus gland by the third month [3,5]. The ectopic thymic cyst develops due to an arrest in migration of thymic tissue or sequestration and persistence of thymic vestiges along the line of descent of thymopharyngeal tract.

This is followed by degeneration of Hassall's corpuscles and/or epithelial component of aberrant thymic tissue. It mainly involves the left side of neck, seen in young children and adolescents and present with mediastinal extension in about 50% cases [6,7]. They are closely related to sternocleidomastoid muscle and carotid sheath contents. However, patient presented with gradually increasing neck mass occupying right carotid triangle posterior to sternocleidomastoid muscle and extending up to the confluence of right and left brachiocephalic vein into anterior mediastinum. Evaluation through imaging modalities can provide an important guidance, the diagnosis confirmation of ectopic thymic cyst is only made by pathologic examination of an excised specimen [8,9].

The other usual reasons for lateral neck masses in children can be branchial cyst, cystic hygroma, lymphadenopathy, haemangioma [5]. The clinical manifestations such as age, size and anatomical location may help in correct interpretation of lesion [5]. Branchial cyst is one of the closest differentials and is derived from second branchial cleft. It appears clinically as a slow-growing, lateral neck mass seen at the anterior border of sternocleidomastoid muscle. Thus, both these lesions share a common anatomical location and presence of lymphoid tissue; however ectopic thymic cyst appears at a relatively earlier age [3]. In thymic cyst, the lymphoid tissue display Hassel's corpuscles that may at times undergo degeneration as evident in present case. However, lymphoid tissue associated with branchial cysts often exhibits germinal centres and these cysts are lined by cuboidal, columnar or stratified squamous epithelium. Presence of cholesterol granuloma in the wall of cyst which consisting of numerous cholesterol crystals admixed with sheets of histiocytes and foreign body giant cells as observed in this case can be common to both the cystic lesions [2]. The other differential can be cystic hygroma which arise due to failure of lymphatic system to communicate with venous system in the neck and is most frequently found in lateral cervical region along the jugular chain of lymphatics including posterior triangle to supraclavicular region. It presents as a multilocular cystic mass with septation of variable thickness [2,3]. The median age of diagnosis being 3 years, that is slightly younger than that of thymic cyst. It is considered to originate from failure of the lymphatic system to communicate with venous system in neck. The cystic spaces are lined by endothelial cells and filled with eosinophilic proteinaceous fluid and separated by thin fibrous wall. Large lesions may extend downward into the mediastinum [2]. The treatment of choice for ectopic thymic tissue is surgical excision with a single cervical incision only after confirming the presence of normal thymus [5-7]. Rarely, may it present as a fast-growing cervical mass necessitating prompt attention due to risk of mass effect on adjacent vital structures. The spontaneous haemorrhage in mass can be one of the reasons for sudden enlargement [10]. Increasing number of cases that are recognised in last few years may indicate the greater awareness of this unusual entity among Pathologists. There might also be a possibility that, many cases of thymic cyst probably had been missed and misdiagnosed as brachial cleft cyst in the past due to inadequate sampling of specimen. The atrophic thymic remnants mandates extensive sampling before a definitive diagnosis of thymic cyst could be rendered [11]. The possibility of thymoma arising in ectopic thymic cyst although unusual is documented in literature as case report [12]. The authors report this rare case to highlight the importance of thymic cyst as one of the considerations for differential diagnosis of cystic lateral neck mass. In similar lines with literature reviewed, the thymic cyst in our child had shown a fibrous cord that can be traced inferiorly upto the superior mediastinum. It may therefore be believed that the ectopic thymic cyst could have arisen from thymopharyngeal duct remnant, which is rarity [13,14].

CONCLUSION(S)

Cervical Ectopic thymic cyst, though rare should be included in the differential diagnosis of paediatric cystic neck swellings. The definitive opinion should always be based on the Histopathological examination of tissue specimen. Surgery is the main stay of treatment if mass is symptomatic and cosmetically unbecoming. Evaluation for the presence and preservation of normal thymus should always be considered prior to surgical removal of ectopic thymic tissue.

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