



Ectopic Cervical Thymoma

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Abstract

The thymus is a lymph epithelial organ derived from the pharyngeal pouches, and anatomically located in the anterior superior mediastinum. Ectopic cervical Thymic lesions results from aberrant migration of the thymus during embryogenesis. Ectopic thymoma is a primary epithelial tumour of ectopic Thymic tissue, most commonly located in the anterior mediastinum. It rarely arises from the neck and other regions. A 27 yr old male presented with the history of mass in the neck, without any other significant complaints. The mass lesion was excised and the histopathological diagnosis of the biopsied specimen was ectopic cervical thymoma.

Keywords: *Thymus, Ectopic thymus, Ectopic Cervical Thymoma.*

Introduction

The Thymus plays a prime role in the cell mediated immunity. During early foetal life, the thymus originates as an epithelial outgrowth from the pharyngeal pouches. It descends from the superior neck into the anterior mediastinum. During downward migration into the mediastinum, remnants of thymus may be sequestered in the neck and other sites, resulting in ectopic thymus. The ectopic thymus can either resemble normal Thymic tissue microscopically or give rise to cystic lesions and tumours. The term thymoma is restricted only for the tumours of Thymic epithelial cells. It most commonly originates in the anterior mediastinum comprising 40% of all mediastinal tumours in adults. Ectopic thymoma is a

relatively rare tumour arising from the remnants of maldescended Thymic tissue present in the neck, thyroid, parathyroid, pulmonary hilus and other sites^[3].

Case History

A 27yr old male, presented with the history of a mass in the postero-lateral aspect of the neck, which was initially small in size and gradually progressed. He had no associated symptoms. On examination, the neck mass was mobile with cystic consistency and measured about 8 cm in diameter. Based upon the preoperative investigations, the clinical diagnosis was made as hemorrhagic cyst. The cystic mass lesion was surgically excised. The histopathological

diagnosis of the biopsied mass was confirmed to be ectopic cervical thymoma.

Macroscopy

A single partially skin covered grey brown, grey black well encapsulated cystic mass measuring 8x7x3cm was received for histopathological examination.

On cut section, a uniloculated cyst measuring 5x4 cm, completely filled with friable hemorrhagic material was identified. The thickness of cyst wall varied between 1 to 3cm.

Microscopy

On microscopic examination, a cellular tumour composed of round topolygonal cells with scanty cytoplasm and prominent nuclei, arranged in sheets was identified. Nucleus appeared pale and vesicular. Numerous spaces filled with Proteinaceous fluid along with nests of tumour cells were present. Perivascular spaces were identified. Histological features were consistent with the diagnosis of an ectopic cervical thymoma.

Plate 1 Gross image showing a well encapsulated tumour mass



Plate 2: Cut section- predominantly cystic change along with hemorrhagic areas



Figure 1: H & E stained (10X)
A highly cellular subcutaneous tumour

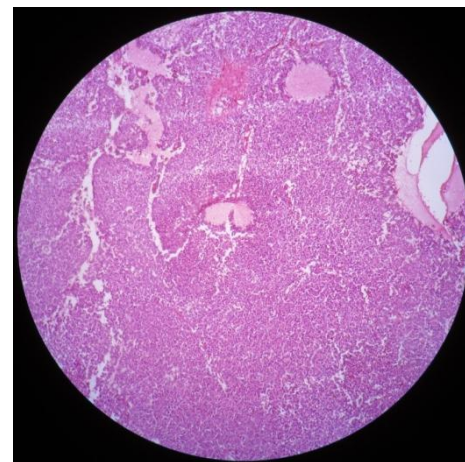


Figure 2: H & E stained (10 X) Thymoma predominantly composed of neoplastic Thymic epithelial cells

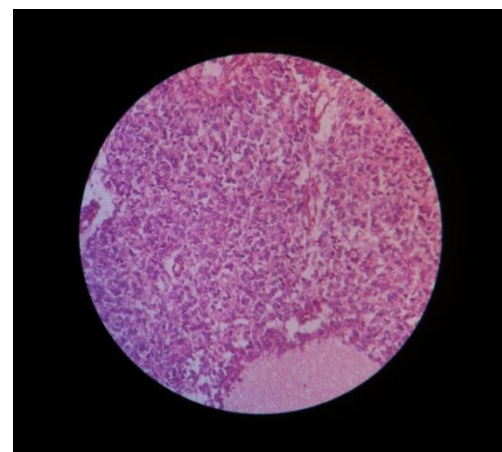


Figure 3: H & E stained (40X) Neoplastic epithelial cells without atypia

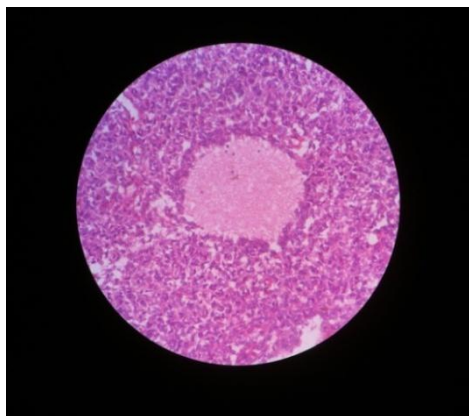


Figure 4: H & E stained (40X) Perivascular spaces filled with Proteinaceous fluid

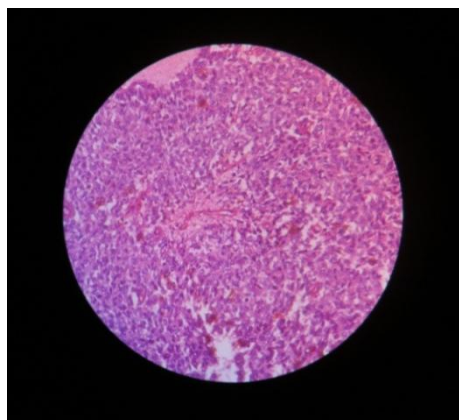


Figure 5: H & E Stained (10X). Perivascular space filled with Proteinaceous fluid



Figure 6: H&E stained (40X)

Perivascular spaces bordered by neoplastic epithelial cells and contains Proteinaceous fluid.

Discussion

The thymus, lymph nodes and spleen are the principal sites of immune response. During early development, T-lymphocytes originating from the bone marrow haematopoietic stem cells migrate to

the thymus and acquire distinctive properties. In 1950, Good first described thymus as an organ with immunological function^[4]. Later, Miller and Martinez established that, intact thymus is essential for cell mediated immunity. Thymus is also considered as a gland, as it secretes polypeptides like thymulin, thymopietin and thymosins with hormonal characteristics and complex endocrine interactions^[5]. Understanding of thymus related diseases has lagged behind because of the inaccessible biopsy site, involution of organ and difficulty in interpretation of morphological changes^[1]. Thymus is now considered as a primary organ of the immune system with a prime function in cell mediated immunity. The thymus is found in all jawed vertebrates with variations in number, size, site among different species. In humans, thymus arises as a paired organ derived from the endoderm of ventral wings of third and fourth pharyngeal pouches on each side of neck along with inferior parathyroid glands. It also receives contribution from the ectoderm of third pharyngeal pouch. During 6th week of intrauterine life, by a process of downward migration, the right and left thymuses come to lie in close apposition, without fusion, in the anterior media stinumas thymus. Upper poles of thymus remain in neck close to the trachea while lower poles lie in the mediastinum, in front of great vessels at the base of heart and pericardium. The two thymuses are enclosed in a fibrous connective tissue capsule. Thymus grows until puberty and achieves a maximum weight of 20 to 50g after which, it undergoes progressive involution. Thymus tends to in volute with age or stress by the process of fatty infiltration and lymphocyte depletion. Fatty change occurs giving the paired organ, a pyramidal shape. Aberrant descent may result in residual Thymic tissue in the neck, and misplacement of Thymic structures in the mediastinum, such as bronchial hilus, pleura or diaphragm and base of skull. Ectopic Thymic nodules have been documented in 20 % normal individuals^[6]. Histologically, the thymus consists of cortex and medulla. In the medullary region,

Thymic lobules are composed of a meshwork of interconnecting epithelial cells in a branching configuration along with the interstitial spaces being filled with small lymphocytes. Hassall's corpuscles with concentric keratinisation or central cavitation is characteristically present. Myxoid cells are also seen rarely in the medulla. These small lymphocytes, namely thymocytes are of T cell origin. The cortex appears dark as it is densely populated by lymphocytes. Tumors of the thymus may be primary or secondary^[1]. Despite several proposed classifications, there is not yet a generally agreed classification of Thymic tumours. However the term thymoma is restricted to primary Thymic tumours in which neoplastic epithelium has a benign or bland appearance. Thymic tumours composed of epithelium with malignant features are termed as Thymic carcinoma^[7].

Tumours arising from striated muscle, neuroendocrine cells, germ cells and haematopoietic cells are placed under separate categories. Thymoma has an incidence of 2-25% of all mediastinal tumours^[8]. Majority are found in the anterior or anterosuperior mediastinum accounting to 94%. Occasionally thymoma can originate from ectopic Thymic tissue with only 4% thymomas arising in the neck and remaining from other sites^[9]. The ectopic thymoma arising in the neck can be unilateral or bilateral. Ectopic cervical thymoma is most commonly located adjacent to thyroid or parathyroid glands; they can even arise from the skin over the neck region giving a clue to branchio-oculo-facial syndrome^[3]. Thymomas can occur at any age peaking in the fifth decade with female preponderance. However for reasons unknown, most of the cervical Thymic lesions are unilateral and common in adolescent males^[10]. Ectopic cervical Thymoma is mostly asymptomatic. Some patients present with mediastinal mass and related symptoms like cough, dyspnoea, dysphagia, hoarseness, recurrent infections. Some patients present with paraneoplastic manifestations like myasthenia gravis, red cell aplasia, acquired

hypogammaglobulinemia. Macroscopically thymomas vary in size and shape and are covered by fibrous capsule. The cut surface is distinctive by septa dividing the tumour into tan-coloured fleshy lobules. Cysts, hemorrhagic foci and calcifications are commonly seen.

Microscopically, the tumour has a jigsaw puzzle like lobules separated by acellular fibrous bands. The diagnostic feature is presence of perivascular spaces, bordered by neoplastic Thymic epithelial cells and filled with Proteinaceous fluid, in which small lymphocytes and RBCs are suspended. Hyaline fibrosis may be present. Rarely, multiple large blood filled cystic spaces called Peliosis thymicis can also be seen. Ectopic cervical thymoma are macroscopically and microscopically similar to thymoma. They are liable to be mistaken for other tumours like lymphoma or carcinoma. Cervical thymoma is also frequently confused with closely related entity known as ectopic hamartomatous thymoma. Ectopic hamartomatous thymoma is a distinctive well circumscribed tumour occurring in the soft tissues of lower neck; and is composed of an admixture of epithelial elements, spindle cells and adipose cells along with perivascular spaces. Invasive thymomas are characterized by invasion of adjacent structures and implantation in the pleura and pericardium. Thymoma is sometimes adherent to the surrounding structures in the mediastinum without any actual invasion. Malignant thymoma is a distinctive entity that can invade locally or metastasize widely. Hence evidence of microscopic invasion is important for differentiation of malignant and benign lesions. Ectopic cervical thymoma has a greatly variable clinical and morphological presentation and it is necessary to differentiate it from other neck lesions. Thymic tissue has a unique echogenicity and hence ultrasound is a sensitive tool for diagnosis. FNAC and flow cytometry may confirm Thymic tissue preoperatively. Since FNA reveals bland cytological characteristics in both benign and malignant Thymic lesions causing diagnostic difficulties, it warrants histopatho-

logical examination for confirmation post operatively^[11]. An intra-operative frozen section is useful to narrow the diagnosis. It also helps in deciding partial or complete resection of the lesion depending upon the tumour presentation. Clinical Staging of the thymoma is the most reliable prognostic indicator than histopathological classification. However, it is determined by the degree of encapsulation, presence of invasion and metastasis, which can be done only by histopathological examination^[11]. Thereby, histopathological examination plays a significant role in confirmation of diagnosis and guides treatment options.

Conclusion

Cervical Thymic anomalies are uncommon and were initially identified at autopsy or by excisional biopsy. They are rarely considered in the preoperative differential of a neck mass. Ectopic cervical Thymic masses have a variable clinical and morphological presentation. They usually present as asymptomatic nodules or incidental neck swellings. As 90% of the lesions have cystic changes, they are clinically misinterpreted as branchial cleft cysts, cystic hygroma, cystic teratoma, lymph proliferative disorders or other tumours. At times they cause debilitating symptoms by encroaching upon adjacent aerodigestive structures. Preoperative diagnosis of ectopic cervical thymus is difficult to establish and is rarely reported, because Thymic vestiges often remain asymptomatic and are mostly unrecognized by clinicians. The specific diagnosis of ectopic Thymic lesions depends largely on histopathologic evaluation. Also, definitive diagnosis, clinical staging, management and prognosis is relied greatly upon the histopathologic examination.

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