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Case Report

A rare case report of Ectopic Cervical Thymoma

Bharathi M.[1], Giri Punja M.[2*], Shilpa K.[3], Geetanjali S.[2]

[1]Professor and Head, [2]PG Student, [3]Associate Professor

Department of Pathology, Mysore Medical College and Research Institute, Mysore, Karnataka, India

*Corresponding author email: giripunja@yahoo.com

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Abstract

The thymus, a lymphoepithelial organ, during the early fetal life originates from the superior neck and descends to the mediastinum. Embryological maldescent may lead to ectopic rests of thymic tissue along the pathway of its descent, neck being the most common site of ectopic thymic rests. Ectopic cervical thymoma is an extremely rare entity, showing a striking female preponderance whereas a mediastinal thymoma has a slight female preponderance. We characterized a case of ectopic cervical thymoma using conventional light microscopy and immunohistochemistry in a 40 years old male patient who presented with a neck swelling simulating a thyroid neoplasm clinically.

Key words

Thymoma, Ectopic, Mediastinum.

Introduction

Thymus originates in the embryo from the ventral ring of 3rd and 4th pharyngeal pouches and ectoderm endoderm of the cervical sinus, as epithelial outgrowths on each side [1, 2]. Being located in the upper anterior mediastinum and lower part of the neck, the thymus is active during childhood and involutes after puberty being replaced by adipose tissue gradually thereafter, although it never disappears completely [3]. Remnants of the thymic tissue may be sequestered in the neck during its migration into the mediastinum, resulting in ectopic thymic tissue, cysts and neoplasms of the cervical thymus. Rarely, an ectopic thymus may lead to the development of malignant thymoma [3].

Case report

A 40 years old male patient presented to the surgical outpatient department with the history of a palpable midline swelling in the neck, just above the supra-sternal notch. There was no associated dysphagia or pain. On physical examination, the mass was 3 cm in diameter, non-tender and was not moving with

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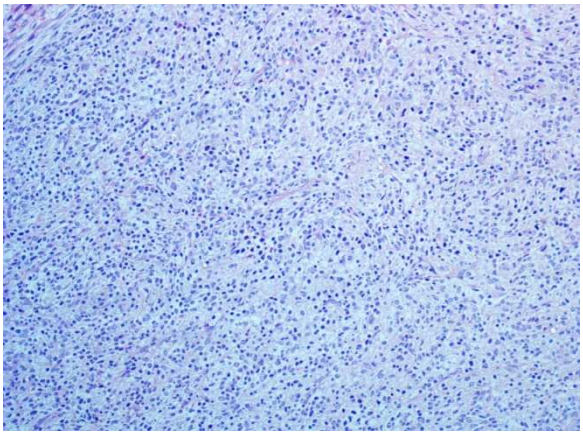
deglutition. He was clinically diagnosed to have ectopic thyroid in supra-sternal region. The patient was euthyroid clinically and biochemically.

A fine needle aspiration exhibited dispersed population of mature lymphocytes, plasma cells with few histiocytes and was opined as non-specific lymphadenitis.

Patient underwent a transverse cervicotomy, followed by excision, and histopathological examination of the excised mass was performed. Gross examination showed a fairly well circumscribed mass measuring 3.2 cm in its greatest dimension. On sectioning of the tumor mass, homogenous gray white areas were identified. Histology revealed an encapsulated tumour mass with fibrous septae extending and dividing the tumor into multiple lobules. The lobules were composed of dual population of cells, spindle cells with oval to spindle, bland vesicular nuclei admixed with sheets of small mature lymphocytes. **(Photo – 1, Photo – 2)** Rosettes, perivascular spaces and glandular structures were noted along with typical hasall's corpuscles. It showed no evidence of cellular atypia or increased mitotic count.

Photo - 1: Microphotograph showing varying proportions of epithelial cells and lymphocytes in thymoma. (100X)



A final diagnosis of mixed thymoma (type AB) was arrived at and confirmed with positive CK14 and CD5 immune markers. **(Photo – 3, Photo – 4)**

Photo - 2: Microphotograph showing oval to spindle epithelial cells with bland vesicular nuclei admixed with lymphocytes. (400X)

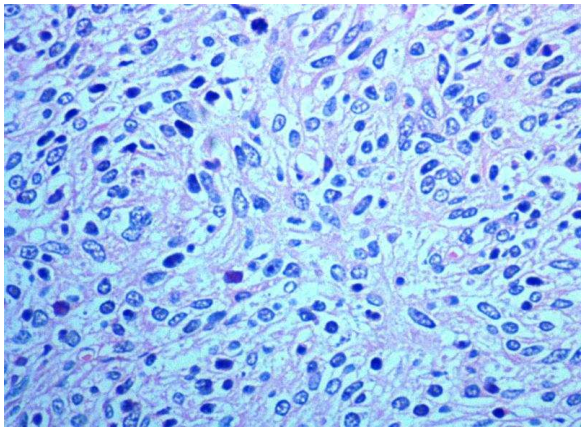


Photo - 3: Microphotograph showing positive immunostain for cytokeratin 14. (400X)

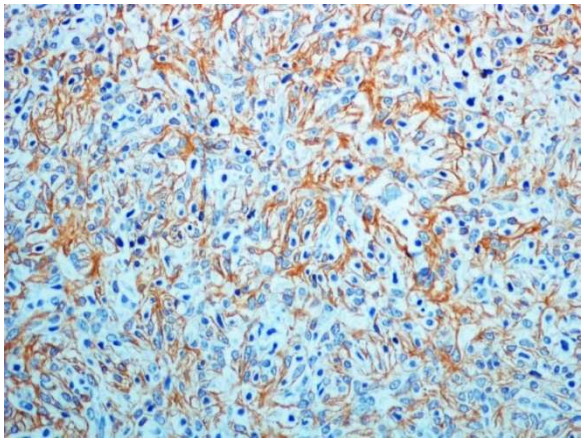
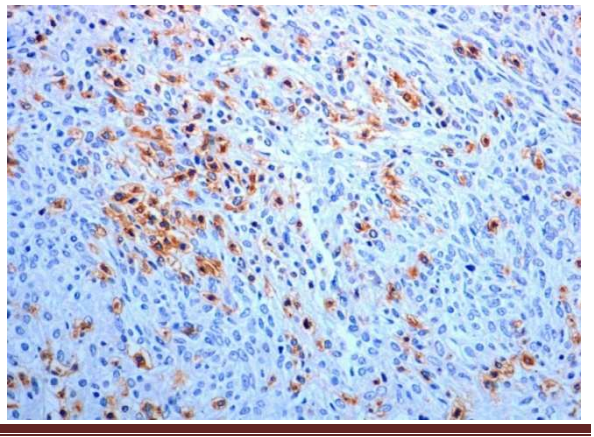


Photo - 4: Microphotograph showing positive immunostain for CD5. (400X)



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Discussion

Thymus was originally believed to be the seat of the soul and near recently to be the "clock for immunologic aging" [4].

Maldescent of the thymus during embryonic life explains the ectopic thymic rests such as in the neck, thyroid, lung, pleura, skull base, root of the bronchus, and in the mediastinum [5]. Majority of the ectopic thymic nodules are located in the neck with close proximity to the thyroid gland, frequently resulting in a mistaken clinical impression of a primary thyroid disorder. Endodermally derived epithelial cells and bone-marrow derived lymphocytes are the two major cell types in thymus. Ectopic thymus can be unilateral or bilateral. About 20% of humans harbour aberrant nodules of thymic tissue; however ectopic cervical thymoma is a rare occurrence [3, 6].

Thymoma is a thymic epithelial neoplasm, accompanied by reactive lymphoid cells in varying numbers, and exhibit organotypic features including medullary differentiation, lobulation, presence of immature T-lymphocytes and perivascular spaces. Thymomas frequently occur in fifth and sixth decades, with a mean age of 49.5years [7]. Although the mediastinal thymoma shows an equal sex incidence or a slight female preponderance, an unexplained and striking preponderance for females is seen in ectopic cervical thymoma. The incidence of ectopic thymoma is about 4% [1]. The most common presenting symptom of an ectopic cervical thymoma is an enlarging neck mass and is frequently misdiagnosed on fine needle aspiration cytology. The diagnosis is difficult to make and has a major diagnostic pitfall [8].

Tumour cells of the thymus grow slowly, look similar to the normal thymus cells and rarely spread to the neighbouring structures. Although thymoma is known to be intricately associated with myasthenia gravis in humans, there is no well documented causative factor for the development of thymoma. Thymoma is also known to be associated with pure red cell aplasia and Good's syndrome.

Thymomas being slow growing tumours have excellent prognosis when diagnosed in their early stages and treated. For thymoma, tumour staging is the single most important prognostic factor. Neoplastic thymic epithelial cells lack CD5 expression and show cytokeratin positivity whereas the non-neoplastic lymphoid cells show CD5 positivity. As in our case, most ectopic cervical thymomas diagnosed as thyroid tumours were removed simply by a neck incision; exact diagnosis was made only after the post-operative histopathological examination.

Our patient underwent a repeat surgery for extended thymectomy and is doing well with 2 years follow up.

Conclusion

In conclusion, clinicians and pathologists must be aware of this rare entity as timely diagnosis and proper management of the patient offers good clinical outcome.

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