International Surgery Journal
Balmoori NR et al. Int Surg J. 2025 Oct;12(10):1822-1824
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20253038

Case Report

Ectopic cervical thymoma: a rare entity in an unusual location

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Received: 08 July 2025
Accepted:10 September 2025

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ABSTRACT

Background: Ectopic cervical thymoma is an extremely rare occurrence, with few reported cases in literature. Thymomas typically arise in the anterior mediastinum, and their presence in the cervical region poses significant diagnostic challenges. Given its rarity, it is often misdiagnosed as other neck masses, such as metastatic lymphadenopathy, neurogenic tumors, or thyroid lesions. This report presented a case of a 57-year-old male who presented with a right upper neck mass, which was progressively increasing in size over several months. The patient underwent thorough clinical and radiological evaluation but the results were inconclusive. Ultrasound guided core needle biopsy was suggestive of spindle cell tumour. Neck exploration and surgical excision was performed. Intraoperatively, a well-encapsulated lesion was identified in the right level II neck region, closely adherent to surrounding structures. Complete excision was achieved without complications. Histopathological examination, supplemented by immunohistochemistry (IHC), confirmed the diagnosis of an ectopic cervical thymoma. Ectopic cervical thymomas are rare and often pose a diagnostic dilemma due to their unusual location and overlap with other neck masses. Preoperative identification is difficult, making histopathology and IHC essential for definitive diagnosis. Complete surgical excision remains the mainstay of treatment, offering good prognosis. Increased awareness of this rare entity can aid in early recognition and appropriate management.

Keywords: Ectopic cervical thymoma, Neck mass, Anterior mediastinum, IHC, Rare cervical tumour, Thymoma

INTRODUCTION

Although thymomas arise almost exclusively in the anterosuperior mediastinum, ectopic variants account for <4% of all thymomas, and fewer than 2% occur in the neck, giving rise to ectopic cervical thymoma (ECT).[1,2] Since 1896, only 112 individual ECT cases have been documented, with patients ranging from adolescence to octogenarians and showing a modest female predominance.[1,3]

Clinically, ECT appears as a slow-growing, painless paramedian neck mass and is frequently mislabelled as metastatic lymphadenopathy, neurogenic tumour, parathyroid or thyroid nodule; fine needle aspiration and frozen section are often non-specific.[4] Imaging typically reveals a well-demarcated, mildly enhancing solid lesion. Histologically, most tumours are World Health Organisation (WHO) type A or AB, and up to 10% are accompanied by paraneoplastic syndromes such as myasthenia gravis, which often improve after excision.[5,6] Complete surgical resection remains the cornerstone of management, and long-term prognosis is excellent in most series.[6]

CASE REPORT

A 56-year-old man presented with a right level II cervical mass. Pre-operative work-up included 18F-flourodeoxyglucose positron emission tomography/ computed tomography (18 F FDG PET CT), and ultrasonography-guided core biopsy. Histology and immunohistochemistry (IHC) were reviewed by two

consultant pathologists, applying the 2021 WHO thymic tumour criteria.

Staging followed the modified Masaoka–Koga system. The patient subsequently underwent neck exploration with en bloc excision of the lesion; the specimen was serially sectioned, photographed and processed for routine H&E. Post-operative recovery was uneventful.

Imaging by PET CT revealed an FDG-avid, defined soft tissue lesion with areas of necrotic component noted involving the right level II cervical region, with maximum dimension measuring 4.4×3.3×3.5 cm; SUVmax 6.3. The lesion was seen closely abutting the right submandibular gland anteriorly and jugular and carotid vessels medially, with preserved fat planes in between. No evidence of infiltration was noted. No abnormal FDG uptake/lesion was noted in the bilateral cervical and supraclavicular regions. Rest of the head, neck and chest structures appeared metabolically inert.

Figure 1: Gross image depicting solid and cystic, lobulated, tan white lesion.

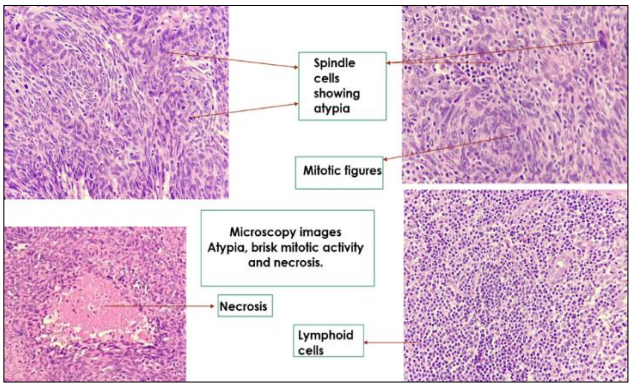

Figure 2: Microscopic images showing atypia, brisk mitotic activity and necrosis.

Core biopsy revealed spindle cell proliferation admixed with mature lymphocytes and rare squamoid whorls. IHC was positive for pan cytokeratin, p40 and p63 in spindle cells, while CD20 and CD5 highlighted background lymphocytes. TdT was negative, and PAX8 showed scattered positivity, favouring ectopic thymoma A/AB type.

Surgical excision yielded a lobulated, tan-white, partially cystic nodule encapsulated by fibro-adipose tissue. Histology confirmed type AB thymoma with atypical type A component, featuring 10–12 mitoses/10 HPF and focal coagulative necrosis, but no capsular or angiolymphatic invasion (modified Masaoka stage I). Surgical margins and surrounding adipose tissue were free of tumour. The patient recovered uneventful.

DISCUSSION

Ectopic cervical thymoma (ECT) is the rarest anatomic subset of thymic epithelial tumours, arising from aberrant descent or sequestration of thymic primordia originating in the third pharyngeal pouch.[2,6] ECTs comprise <2% of all thymomas and <0.5% of adult neck tumours, with only 112 cases documented to date.[3] Their indolent presentation as painless neck nodules often lead to misdiagnosis as metastatic nodes, neurogenic tumours or thyroid/parathyroid lesions. While fine needle or core biopsy can suggest thymic differentiation, the diagnosis hinges on a constellation of spindle cell morphology, dual epithelial–lymphoid architecture and a CK+/p40+/CD5+ immunophenotype.[4]

Our case adds to the small pool of atypical variants, showing hypercellularity, brisk mitoses and necrosis—features first highlighted by Nonaka et al. in "atypical type A thymoma," now recognized as an aggressive spectrum within type A/AB lesions.[9]

Despite these worrisome features, the absence of capsular invasion placed the tumour at stage I, where complete surgical resection is considered curataive.[6,10] Neck exploration allows en bloc excision with low morbidity; transcervical thymectomy is unnecessary unless concurrent mediastinal disease is proven. Adjuvant radiotherapy is generally reserved for stage II–III mediastinal thymomas. Extrapolating from those data, most authors omit radiotherapy for stage I ECTs if margins are negative.[10]

Long-term outcomes were excellent: Nemiroff et al reported 95% five-year disease-specific survival for ectopic lesions versus 86% for mediastinal counterparts.[1] Nonetheless, late recurrences—including lung and brain metastases—have been described up to 15 years post resection.[9] Hence, prolonged surveillance is prudent.

CONCLUSION

In summary, this case underlines the value of thorough histopathological and IHC evaluation in neck spindle cell tumours, reinforces surgery as the cornerstone of treatment, and enlarges the literature on atypical type AB ectopic thymoma.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Balmoori NR, Sreekanth K, Arun SRAR, Sai CR, Manjula VK. Ectopic cervical thymoma: a rare entity in an unusual location. Int Surg J 2025;12:1822-4.