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An inflammatory myofibroblastic tumor of the soft tissue of the neck: a case report and literature review

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Inflammatory myofibroblastic tumor (IMT) of the soft tissues of the neck is a rare pathological entity. We present the case of a 32-year-old patient who had a painful, slowly enlarging mass in the neck's soft tissues. Radiological examination revealed a well-defined, dense lesion. The mass was surgically removed through local excision. Immunohistological analysis confirmed the diagnosis of IMT. After nearly 2 years of follow-up, there was no evidence of recurrence or distant metastases. In conclusion, although IMT of the soft tissues of the neck is uncommon, it should be considered in the differential diagnosis of neck tumors. Further research is necessary to understand the pathogenetic mechanisms of IMT, which could lead to the development of more effective treatments for this tumor.

Abbreviations: ALK, anaplastic lymphoma kinase; BCL2, B-cell lymphoma 2; EMA, epithelial membrane antigen; IMT, inflammatory myofibroblastic tumor; SMA, smooth muscle actin; TPM-3, tropomyosin-3; TPM-4, tropomyosin-4

Keywords: Anaplastic lymphoma kinase / Case reports / Inflammatory myofibroblastic tumor / Tropomyosin-3 / Tropomyosin-4 / World Health Organization

Case Report and Literature Review

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor, is a rare condition that poses diagnostic challenges due to its non-specific clinical and radiological manifestations. This tumor can occur in both viscera and soft tissues and affects individuals of all ages. The most common sites for IMT in the head and neck region are the orbits, paranasal sinuses, and oral cavity [1]. Clinically, IMT typically presents as a mass lesion, which may or may not be accompanied by non-specific symptoms such as night sweats and fever [2]. Radiologically, the tumor may present as either a hypoechoic or hyperechoic mass, with boundaries that are either ill-defined or well-defined [2]. Several cases of IMT have been documented in the medical literature [1,3]. Here we present a case of IMT involving the soft tissue of the neck in a middle-aged woman. We discuss the clinicopathologic features of this case.

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CASE REPORT

A 32-year-old female patient presented with a slowly enlarging, mildly painful swelling in the soft tissue on the left side of her neck that had persisted for 5 months. Her medical history

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showed no significant findings. A clinical examination identified a 15 mm solid mass at the base of her neck, with no palpable cervical lymph nodes detected. Radiological assessment, including an ultrasound, revealed a single 16 mm solid lesion at the base of the neck, indicative of a neoplastic process.

A computed tomography scan of the neck revealed a well-defined, small, dense lesion on the left side of the cervical root, located at the level of the left trapezius muscle, measuring approximately 15×15 mm. Prominent adjacent subcutaneous vessels were also noted. Several prominent upper and lower deep cervical lymph nodes, ranging in size from 6 to 7 mm, were observed (Fig. 1). The blood vessels in the neck and the thyroid gland appeared unremarkable. Radiographic impressions suggested the possibility of soft tissue hemangiomas or other soft tissue tumors.

Local excision of the mass was performed with safety margins of 0.3 cm and the postoperative recovery was uneventful. Grossly, the mass was 15×15 mm, well-defined, solid, and nonencapsulated with a gray cut section and a firm consistency. No hemorrhage, cystic changes, necrosis, or calcification was noted. Histologically, the mass was composed of spindle cells organized in a bundle pattern, displaying rare mitotic figures (mitotic count: 1/10 high-power field) and signs of regressive changes, including old hemorrhage, cystic degeneration, and fibrosis. The tumor cells tested positive for vimentin, smooth muscle actin, calponin, β-catenin, and B-cell lymphoma 2. The proliferation index of Ki-67 was less than 1%. Stains that returned negative results included anaplastic lymphoma kinase (ALK), cytokeratin (AE1/AE3 cocktail), epithelial membrane antigen (EMA), S100, CD34, CD45, and p53 (Figs. 2, 3). These immunohistological features confirmed the diagnosis of IMT.

The patient completed her nearly 2-year follow-up with no evidence of recurrent or metastatic lesions.

LITERATURE REVIEW

IMT was classified by the World Health Organization in 2002 as a neoplastic entity. It is considered a borderline tumor due to its potential to occasionally recur locally or metastasize [4,5]. Typically, IMT is managed through surgical excision; however, as of now, no safety margin has been established in the literature [1,3]. Approximately 50% to 75% of IMTs exhibit fusions between the *ALK* gene and the genes for tropomyosin-3 (*TPM3*) and tropomyosin-4 (*TPM4*) on chromosome 2p23, leading to ALK overexpression. ALK protein staining is positive in about 40% to 100% of IMT cases [4]. In some IMT/ITPs, the tumor cells are negative for ALK [6,7]. About half of the IMT have genetic changes in the *ALK* gene and ALK protein expression. Nevertheless, the absence of ALK protein expression does not rule out an IMT diagnosis [7].

DISCUSSION

IMT primarily occurs in the lungs and the orbital region, with only about 15% of cases presenting in the nonorbital areas of the head and neck. Instances of IMT in the soft tissue of the neck are extremely rare and have not been well-documented in the literature [1,3]. In this report, we present a case of IMT in the neck's soft tissue. The clinicopathological features of this case align with those described in previous studies [3,4]. Chen et al. [3] studied 14 patients with IMT. Of these, nine had lesions confined to the maxilla, mandible, or neck. Complete

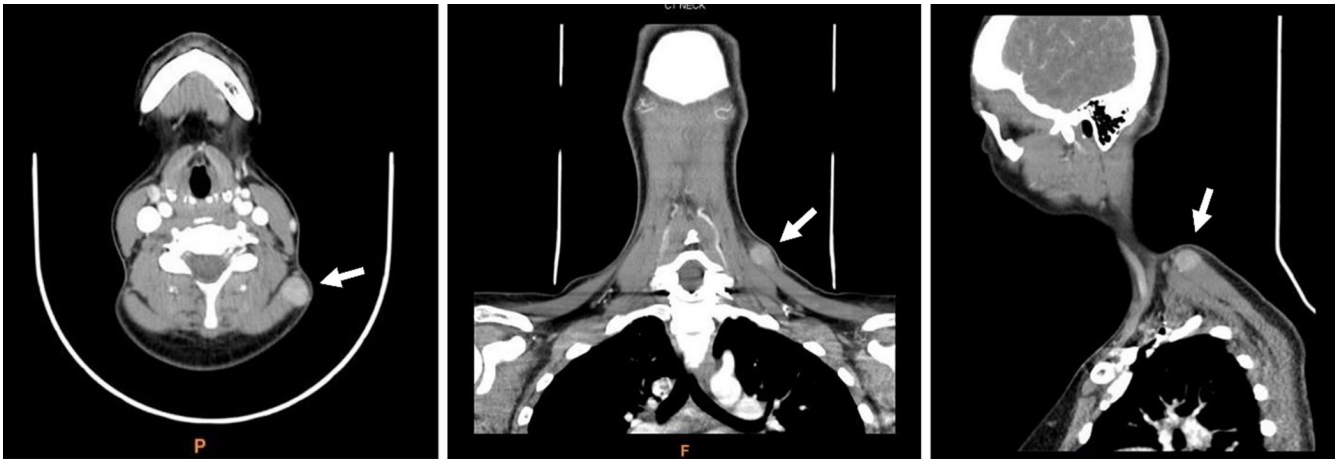

Fig. 1. A slowly growing, mildly painful mass in the soft tissue on the left neck of a 32-year-old woman: A 15 mm solid mass (white arrow) is identified at the base of her neck without palpable cervical lymph nodes on computed tomography. No evidence of breakdown or calcification, or invasion of the overlying skin or the underlying muscle is seen.

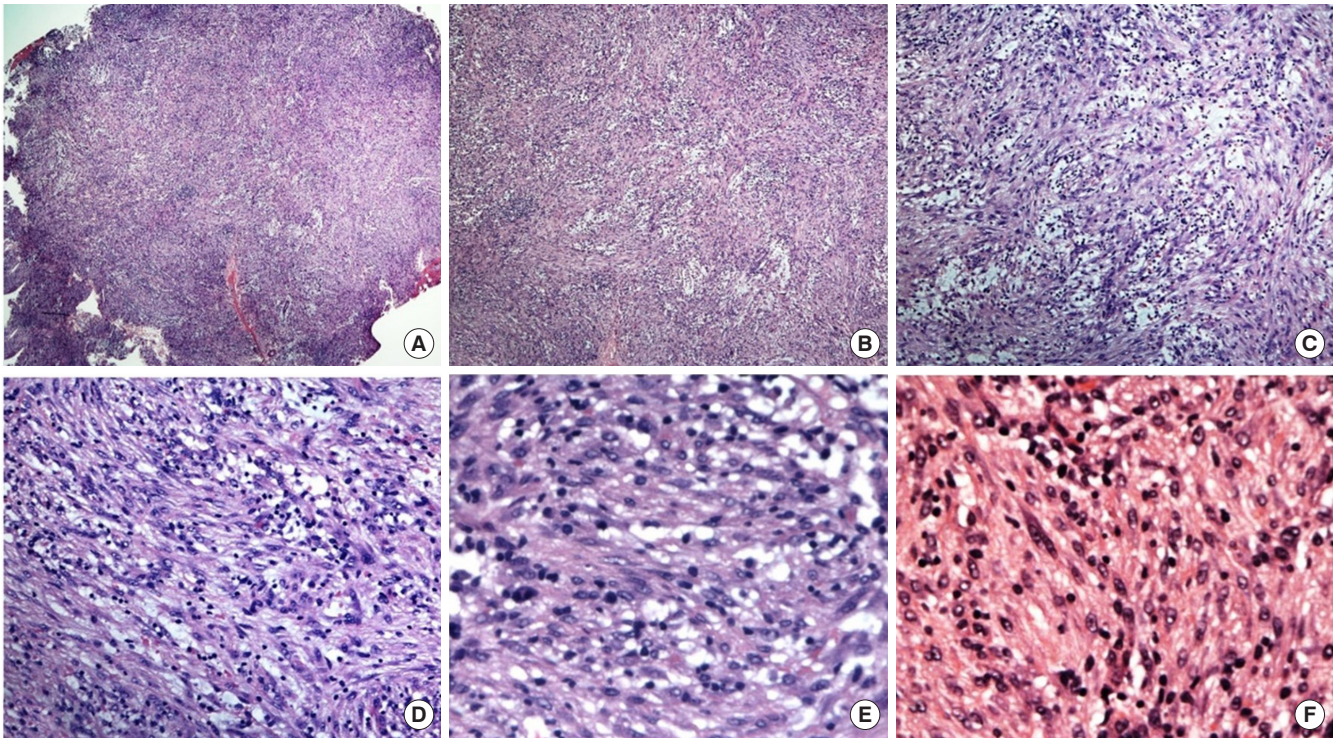

Fig. 2. The histological features of the inflammatory myofibroblastic tumor of the neck. The histologic sections revealed cell proliferation characterized by spindle, oval, and fusiform nuclei arranged in indistinct bundles and storiform patterns. These were interspersed with mixed inflammatory cells, including lymphocytes, histiocytes, plasma cells, and eosinophils, as well as aggregates of xanthoma cells (C). There was no significant nuclear atypia, necrosis, or atypical mitotic figures observed. The sections in all panels were stained by hematoxylin and eosin stain (A: ×20, B: ×40, C: ×100, D: ×200, E: ×400, and F: ×400).

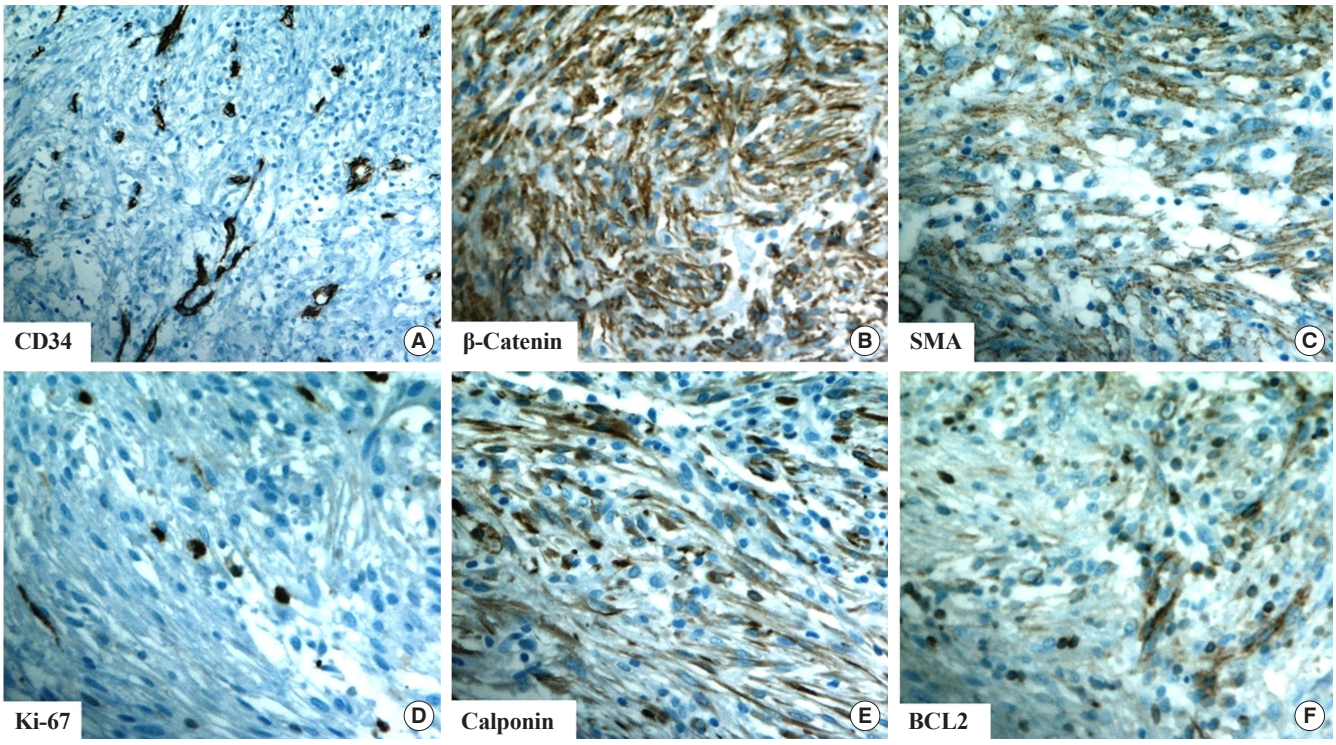

Fig. 3. The immunohistochemical features of the inflammatory myofibroblastic tumor of the neck. The tumor cells tested positive for smooth muscle actin (SMA), calponin, β-catenin, and B-cell lymphoma 2 (BCL2). Negative staining was observed for CD34. The Ki-67 proliferation index was below 1%. The sections in all panels were stained by the immunoperoxidase staining method (all ×400).

cures were achieved through local excision of the mass lesions. However, three cases of maxillary IMT required additional treatments—steroids, radiotherapy, and chemotherapy—yet still experienced local recurrences at 6, 9, and 2 years, respectively. One patient with maxillary IMT developed cervical metastases and succumbed to brain invasion. Another patient, with IMT localized around the common carotid artery, was treated with prednisone and remained disease-free at a 2-year follow-up [3].

The histopathological diagnosis of IMT is challenging due to its varied histological presentations. It is difficult to distinguish from other spindle cell lesions. Grossly, IMT typically presents as a nodular mass, either without a capsule or with a pseudocapsule. Histologically, it is characterized by a mix of myofibroblasts, fibroblasts, and stellate cells, along with an inflammatory cell infiltrate and occasionally some highly abnormal cells [8]. IMT exhibits three distinct histological patterns. The hypocellular fibrous pattern features a keloid-like morphology with a prominently hyalinized stroma [9]. The hypercellular pattern is marked by densely packed spindly cells intermingled with inflammatory cells. In the mucinous pattern, tumor cells are embedded in an abundant myxoid stroma. In the case reported here, the differential diagnosis for IMT included various other spindle cell lesions such as myofibroblastoma (desmin and CD34-positive), nodular fasciitis (ALK-negative), fibromatosis (ALK-negative and β-catenin-positive), inflammatory leiomyosarcoma (desmin-positive), dermatofibrosarcoma (CD34-positive), sarcomatoid carcinoma (cytokeratin and EMA-positive), and spindle cell melanoma (S100-positive) [1,2,8]. A high Ki-67 labeling index and strong, diffuse p53 staining are associated with malignant transformation [10]. In the case presented here, p53 staining was negative, with only a few scattered weakly positive tumor cells, and the Ki-67 labeling index was low, indicating an absence of malignant transformation.

The management of IMT presents significant challenges and variability. Some cases of IMT may exhibit a pronounced inflammatory component and atypical Hodgkin-like cells, mimicking Hodgkin lymphoma [11]. Complete surgical resection is generally the preferred treatment for IMT. However, in rare instances, patients may experience local recurrence or develop distant metastases, necessitating additional interventions such as radiation and chemotherapy [8,12]. In the case reported here, the patient underwent complete surgical resection, and subsequent follow-up showed no signs of recurrence or metastasis.

In conclusion, we report a rare case of IMT in the neck. Although IMT in the soft tissues of the neck is an uncommon neoplasm, it should be considered in the differential diagnosis of patients presenting with a neck mass. There are several key takeaways from this case. Notably, IMT of the soft tissues of the neck is an exceedingly rare pathological entity with distinct immunohistological characteristics and uncertain biological behavior. Approximately half of IMTs exhibit genetic alterations in the *ALK* gene. The neoplastic nature of IMT is evidenced by the fusion of the *ALK* gene with *TPM3* and *TPM4* genes. This tumor poses a risk of local recurrence or even distant metastasis.

NOTES

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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Ethical approval

The study protocol was reviewed, and the need for approval was waived by the Research Ethics Board of the Armed Forces Hospital Southern Region Research Institute.

Patient consent

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

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editing: all authors. Resources: Yahia Awad Alkahtani. Software: Mahmoud Rezk Abdelwahed Hussein. Supervision; Validation: Yahia Awad Alkahtani, Mubarak Mohammed Al-Shraim.

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