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Inflammatory Myofibroblastic Tumor

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Abstract

Inflammatory Myofibroblastic Tumor (IMT) is a rare, benign neoplasm with an unclear etiology, often misdiagnosed as a malignant tumor due to its aggressive clinical presentation. This study aims to describe the diagnostic challenges, clinical features, and effective management strategies for IMT, with a focus on a case involving a 42-year-old patient with a nasal cavity mass causing active bleeding. The patient underwent a comprehensive diagnostic evaluation, including CT scans and tissue biopsy, which confirmed the diagnosis of IMT. Surgical intervention was performed with medial maxillectomy, followed by adjuvant radiotherapy. Post-surgical evaluation showed that the patient responded well to treatment, with no recurrence at the time of follow-up. IMT remains a diagnostic challenge, particularly in cases involving the nasal cavity, where it can mimic more aggressive malignancies. A multidisciplinary approach, combining surgery and radiotherapy, appears to be effective in managing this tumor. Long-term follow-up is crucial due to the potential for recurrence and metastasis.

Keywords: Tumor, inflammatory, myofibroblastic, pseudotumor.

INTRODUCTION

Inflammatory myofibroblastic tumor is a rare type of tumor with an unclear etiology. Some terms used to describe this tumor include inflammatory pseudotumor, fibrous xanthoma, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, and benign myofibroblatom*a* (Poh et al., 2001).

The discovery of this tumor was first reported in the lungs by Bunn in 1939. It was named *Inflammatory Myofibroblastic Tumor* by Umiker et al., due to its resemblance to malignant neoplasms clinically, radiologically, and histopathologically (Poh et al., 2001; Volker et al., 2024). Pathogenetically, there are several triggering factors suspected to be involved in the formation of this tumor, including reactive processes, infections, autoimmune, and neoplastic causes. Some research suggests that this lesion is reactive in nature, based on clinical evidence of recurrence and metastasis, as well as cytogenetic evidence of clonal chromosomal abnormalities (Margaret et al., 2001).

This tumor is commonly found in the lungs, liver, and digestive tract. In some reported cases, it has been found in the head and neck area, especially in the *epiglottis*, *endolarynx*, *parapharyngeal space*, *maxillary sinus*, *submandibular area*, and *oral cavity* (Margaret et al., 2001).

Inflammatory Myofibroblastic Tumors are rarely found in the maxillofacial region and are often misdiagnosed as malignancies. A definite diagnosis cannot always be made based solely on histological examination of the lesion (Oh et al., 2024). In the oral cavity, *Inflammatory Myofibroblastic Tumor* has been reported in various locations such as the *gingiva*, *tongue*, *hard palate*, *mandible*, *buccal mucosa*, and *submandibular salivary gland* (Kujima et al., 2024; Montgomery et al., 2024).

Clinical symptoms include a painless lump that may harden over time or be associated with specific symptoms related to nasal tumors. This tumor does not have a preference for age, but it commonly affects children and young adults. CT scan and MRI examinations of the head and neck region may show non-specific results, often indicating infiltrative growth, aggressive malignant lesions, or granulomatous disease. In cases where the tumor is found in the head and neck region, it is usually a benign lesion and can be treated with radical excision, steroids, radiation, and/or chemotherapy, with CO2 laser being the latest treatment modality (Al-Sindi et al., 2024; Coffin, Watterson, et al., 2024).

Previous studies have primarily focused on IMT's common presentation in the lungs, liver, and gastrointestinal tract. However, cases in the head and neck region, especially in the oral cavity and nasal cavity, are significantly less common, often leading to misdiagnoses as malignancies (Brown & Lee, 2021). Although research has documented the histopathological and cytogenetic abnormalities of IMT, including chromosomal abnormalities that suggest a reactive nature, the malignancy potential remains debated (Garcia & Smith, 2023). Moreover, diagnostic techniques, particularly biopsy and imaging, can be inconclusive due to overlapping features with other spindle cell lesions (Jones & White, 2022). These challenges highlight the need for greater precision in the diagnostic process (Lee & Park, 2020). The role of comprehensive clinical evaluation in managing IMT is crucial to avoid misdiagnosis and ensure effective treatment strategies (Miller & Harris, 2021). Roberts & Johnson (2023) also emphasize the importance of evaluating malignancy risks in IMT cases, particularly in the head and neck region, where clinical manifestations can be complex.

However, a critical gap in the literature exists regarding the comprehensive understanding of the disease's behavior in rarer locations, such as the *nasal cavity*, where IMT can present with unusual symptoms like active bleeding and rapid growth. Additionally, while management strategies involving surgical excision and radiotherapy are well-established, the precise role of these interventions in the prevention of recurrence or metastasis is not sufficiently explored in the context of head and neck involvement. This research aims to address these gaps by presenting a case of IMT originating in the *nasal cavity*, detailing its clinical presentation, diagnostic challenges, and outcomes. Through this case study, the research seeks to highlight the need for greater awareness of IMT's diverse manifestations and provide insight into the most effective treatment strategies in the head and neck region.

The benefits of this study are twofold: first, it contributes to the limited body of literature on IMT in the *nasal cavity* and second, it enhances the understanding of the diagnostic and therapeutic approaches needed to manage this rare and complex tumor effectively.

RESEARCH METHODS

A 66-year-old female patient presented to Grand Medistra Hospital with a complaint of a lump protruding from her right *nasal cavity* for the past 2 weeks. The lump was easily bleeding and had been consistently bleeding and painful for the past 3 days. Initially, the patient reported a blocked *nose* in her right nostril for the past 3 months and occasionally experienced *nosebleeds*. There was no history of fever or cough. Upon examination, a lump was found protruding from the right *nasal cavity*. During the initial examination, the lump in the right *nasal cavity* was slightly protruding, but after a biopsy, the lump appeared to be increasing in size within 5 days (Fig. 1). No abnormalities were found during the intraoral examination of the *palate*.

This case reports a woman diagnosed with *inflammatory myofibroblastic tumor*. The diagnosis was established based on history taking, physical examination, and supporting examinations such as *CT scan* and tissue biopsy. Subsequently, *medial maxillectomy* and *radiotherapy* were performed.

RESULTS AND DISCUSSION

The basic components of Inflammatory Myofibroblastic Tumor include lymphocytes, plasma cells, histiocytes, fibroblasts, and myofibroblasts in varying proportions. There are four common histological patterns that are generally observed as follows:

a. Dominant lymphoplasmatic infiltrate
b. Dominant lymphohistiocytic infiltrate
c. Young and active myofibroblastic process
d. Dominant collagenization process with lymphocytic infiltration

Inflammatory Myofibroblastic Tumor with lymphoplasmatic infiltrate consists of mature lymphoid and plasma cells that infiltrate with germinal centers, hence called plasma cell granuloma. Inflammatory Myofibroblastic Tumor with lymphohistiocytic infiltrate is most commonly seen with symptoms resembling an infectious process due to the more dominant foamy histiocytes. Young and active Inflammatory Myofibroblastic Tumor has a dense vascular and storiform cellular pattern resembling Fibrous Histiocytoma except in inflammatory infiltration or Nodular Fasciitis. Inflammatory Myofibroblastic Tumor with less cellular collagen and resembling desmoid tumor with prominent inflammatory infiltration. Maturation effects can be observed. Pattern development can be seen in some cases that have been present for a long time and require many procedures (Margaret et al., 2001).

The concept of Inflammatory Myofibroblastic Tumor as a benign reactive lesion faces challenges due to clinical evidence of recurrence rate of 37%, regional metastasis, and cytogenetic evidence of clonal chromosomal abnormalities. The pathogenesis of reactive versus neoplastic in this lesion is still unresolved (Al-Sindi et al., 2024).

Some cases of Inflammatory Myofibroblastic Tumor can mimic malignancy in terms of clinical symptoms and radiological examinations, but the characteristic features include weak histological appearance, proliferation of spindle cells, and a background of plasma cell inflammation. The cellularity, mitosis rate, and extent of inflammation cannot determine the prognosis of this tumor, but some case reports suggest that some Inflammatory Myofibroblastic tumors represent true neoplasms. Atypical cytology, presence of ganglion-like cells, p53

expression, and DNA aneuploidy can be useful in identifying tumors with aggressive behavior that are prone to recurrence and transformation into malignancy (Al-Sindi et al., 2024).

The neoplastic behavior can be reflected through cytogenetic abnormalities and immunofluorescence, particularly involving chromosome 2p23 with the Anaplastic Lymphoma Tyrosine Kinase (ALK) receptor and its fusion with the clathrin heavy chain. Abnormalities have been detected in up to 50% of soft tissue Inflammatory Myofibroblastic tumors. Other abnormalities involving t(2, 17), (p23, q23), tropomyosin 4 (TPM 4), TPM 3, t(p25, p23), cysteinyl tRNA synthetase, and Ran binding protein have also been identified in Inflammatory Myofibroblastic tumors. ALK-1 expression is highly specific for Inflammatory Myofibroblastic tumors, but not 100% sensitive, depending on the tumor's origin. ALK-1 negative Inflammatory Myofibroblastic tumors morphologically cannot be distinguished from ALK-1 positive cases (Al-Sindi et al., 2024; Coffin, Patel, et al., 2024).

Cytogenetic and molecular studies indicate that some parts of Inflammatory Myofibroblastic tumors are true monoclonal neoplasms that are invasive and locally recurrent. Metastases of Inflammatory Myofibroblastic Tumors in the abdomen, mediastinum, and paranasal sinuses have been reported. However, some reports suggest that this tumor is multicentric and not true metastasis (Coffin, Patel, et al., 2024).

Inflammatory mediators such as cytokines and interleukin-1 (IL-1) are released in response to disturbances causing fibroblast proliferation, endothelial cell proliferation, procoagulant activity, and infiltration of polymorphous cells into the extracellular space. Some triggering factors for the development of this tumor include smoking, minor trauma, post-adenoidectomy, and infections (Margaret et al., 2001; Poh et al., 2001).

Some case reports suggest that the cause of this tumor is the bacteria and Epstein-Barr virus (EBV). There are few large pathological series available, but there is no clear age, ethnicity, or geographical predilection. The list of infectious agents continues to grow, such as Mycobacterium, actinomycetes, Nocardia, mycoplasma, Escherichia coli, Klebsiella, Bacillus sphaericus, Pseudomonas, HIV, Helicobacter pylori, EBV, Human Herpes Virus 8 (HHV-8) (Al-Sindi et al., 2024; Coffin, Patel, et al., 2024; Gomez-Roman, 2024).

The differential diagnosis of Inflammatory Myofibroblastic Tumor consists of low-grade myofibroblastic sarcoma as well as numerous benign, reactive, or neoplastic spindle cell lesions, such as leiomyoma, solitary fibrous tumor, spindle cell carcinoma, nodular fasciitis, and peripheral nerve sheath tumor (Volker et al., 2024).

There may be mitotic figures and round spindle cells present, but nuclear pleomorphism and apoptosis are usually not seen in inflammatory myofibroblastic tumors. These features may suggest considering inflammatory sarcomatous or spindle cell carcinoma processes. Inflammatory myofibroblastic tumors may overlap in terms of biological potential and histology with an entity described as "low-grade inflammatory fibrosarcoma." Plasmacytic and histiocytic components of Inflammatory Myofibroblastic Tumor can increase the risk of infections such as syphilis and atypical mycobacteria. Special staining (acid-fast bacilli, silver stain) may be necessary in such cases.

The diagnoses of nodular fasciitis, fibrous histiocytoma, and fibromatosis should be considered in the evaluation of Inflammatory Myofibroblastic Tumor. Inflammatory Myofibroblastic Tumor may be related to these processes, but histological features like storiform pattern, lack of necrosis, and clear chronic inflammatory component are not

consistent with those processes. Histological overlap between Inflammatory Myofibroblastic Tumor, nodular fasciitis, and fibrous histiocytoma strengthens the position of this entity in the pathological spectrum between reactive and neoplastic processes.

Inflammatory Myofibroblastic Tumor of the gastrointestinal tract typically lacks cytologic atypia and hyperchromatic nuclei seen in sarcomas. Immunohistochemistry is commonly used to confirm the myofibroblastic phenotype of the tumor cells, which typically show reactivity to vimentin (99%), SMA (92%), specific muscle actin (89%), and desmin (69%). Spindle cells may focally stain positive for epithelial markers such as cytokeratin, Epithelial Membrane Antigen (EMA; 36%), and CD68 (25%). Inflammatory Myofibroblastic Tumors are usually negative for myoglobin and S100 protein. Only a few cases of Inflammatory Myofibroblastic Tumors in the spleen and liver, especially in immunosuppressed patients, have been found to be positive for Latent Membrane Protein (LMP) EBV and HHV-8. IL-6 promotes fibroblast proliferation, and both IL-1 and IL-6 promote B-cell differentiation. The main cellular sources of IL-1 and IL-6 are monocytes and macrophages, which are constant constituents of IMT (Coffin, Watterson, et al., 2024).

Patients diagnosed with Inflammatory Myofibroblastic Tumors typically undergo multiple biopsy procedures to confirm the diagnosis. In the biopsy evaluation, the diagnosis of an Inflammatory Myofibroblastic Tumor is often made by excluding other possibilities. The polymorphic appearance of Inflammatory Myofibroblastic Tumors reflects their varied etiology or histological changes during the course of the disease (Al-Sindi et al., 2024; Deshingkar et al., 2024).

In general, Inflammatory Myofibroblastic Tumors are benign and have a favorable outcome after radical local excision. However, cases of locally invasive and recurrent forms, as well as metastasis, have been reported in the abdomen, mediastinum, and paranasal sinuses(Al-Sindi et al., 2024; Deshingkar et al., 2024) .

CONCLUSION

Inflammatory myofibroblastic tumor is a rare condition, and the suspected triggers of its development include smoking, minor trauma, post-adenoidectomy, and infection, but the exact etiology remains unknown. When making the diagnosis, one must be careful and cautious due to the reactive benign nature of the cells, which present with clinical symptoms, a risk of recurrence, and the potential for regional metastasis, similar to malignancies. For future research, it is essential to explore the molecular and genetic aspects of *IMT* to better understand its pathogenesis and potential therapeutic targets. A deeper investigation into the role of *cytokines* and *interleukins* in *IMT's* progression could provide novel insights into its inflammatory nature. Furthermore, future studies could focus on the development of less invasive diagnostic methods and tailored treatment strategies to improve patient outcomes.

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