



An Overview of the Literature and a Clinical Report on Inflammation Myofibroblastic Tumors of the Maxillary Sinus

AS Murthy*

Division of Plastic Surgery, Akron Children's Hospital, Akron, Ohio, USA.

*Corresponding Author: Murthy AS, Division of Plastic Surgery, Akron Children's Hospital, Akron, Ohio, USA, E-mail: asmurthy@chmca.org

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Editorial Note

Despite being recognized as a clonal neoplasm, the biology of Inflammatory Myofibroblastic Tumours (IMTs) is poorly understood due to their varied appearance. There is no known origin, and they usually manifest themselves as tumor growth mixed with inflammation. IMTs are classified as intermediate malignancies by the World Health Organization. IMTs are characterized as rarely metastasizing in terms of biological potential. IMTs in the maxillary sinus are uncommon, but when they do occur, they can be aggressive locally or even destructive if they enter the orbit. The authors offer a brief clinical report documenting the treatment of a five-year-old girl who had a slow-growing tumor in the right maxillary sinus that extended into the lacrimal sac.

A five-year-old child came with right-eye epiphora. She developed papilledema on the medial orbital margin and a palpable lump. A tumour in the maxillary sinus that extended into the lacrimal sac was confirmed by computed tomography imaging. To acquire tissue for diagnosis, a biopsy was conducted. IMT was suspected after a pathological examination. Immunohistochemistry indicated that smooth muscle actin was expressed uniformly in a 'tram track' pattern, indicating myofibroblastic development. Anaplastic Lymphoma Kinase (ALK) protein expression, S-100 protein, wide-spectrum cytokeratin, and desmin expression were all negative in the tumour. Surgical intervention was suggested as a result of these findings. She had a dacryocystorhinostomy and a medial maxillectomy with frozen-section management. Her surgical recovery went without a hitch. She did exhibit papilledema, which was proven to be Drusen's papilledema following a complete workup that included magnetic resonance imaging and lumbar puncture. At a one-year follow-up with imaging and endoscopy, she was tumor-free.

The World Health Organization (WHO) classified IMTs as separate entities in 1994. IMTs are classified as a clonal neoplasm with moderate malignancy that seldom metastasizes, according to the 2013 classification [1]. Inflammatory pseudotumour, xanthogranuloma, histiocytoma, plasma cell granuloma, inflammatory fibromyxoid tumour, and myofibroblastic proliferation have all been used to describe IMTs in the past. IMTs are most typically found in the lungs throughout the first two decades of life. Extra pulmonary tumors are uncommon, and most are found in the momentum and mesentery [2].

IMTs normally manifest as a solid mass, but in 15% to 20% of instances, systemic symptoms such as fevers, lethargy, and weight loss are also present [3]. IMTs present as a modestly enhancing soft tissue mass with no calcification on computed tomography imaging. Only about 5% of cases have spread to other parts of the body [4].

ALK is only found in the brain, testes, and small intestine, and is not found in normal lymphoid tissue. It was discovered as a fusion partner in anaplastic large cell lymphoma (hence the term "anaplastic lymphoma kinase"). This gene was later shown to be involved in the pathophysiology of IMT. In fact, clonal cytogenetic rearrangements that activate the ALK receptor tyrosine kinase gene on chromosome 2p23 are found in about 30% of IMTs in children. IMTs have an unclear cause. Viruses such as human herpes virus 8 and Epstein-Barr virus have been studied, although the evidence is still ambiguous [5-8]. The ALK gene on chromosome 2 is thought to be fused to one of numerous partners in ALK-positive tumors, such as nonmuscular tropomyosin 3, which can provide the tumor proliferative capabilities [9,10]. Atypical and positive ALK status are more common in aggressive tumours, although negative ALK status is shown in recurring or metastatic tumors, which is contentious.

Inflammatory Pseudo Tumor

IMTs are a type of chronic inflammation that develops as a result of an intermediate neoplastic phase. This explains why corticosteroids, which are used to reduce inflammation, are ineffective in treating malignant tumours. Surgical excision with clear margins is the optimum treatment and important to prevent recurrence in locally aggressive or big tumours, especially if ALK expression is negative. Treatment of IMTs of the maxillary sinus with corticosteroids failed in 66% of instances described in the literature, according to a recent meta-analysis. In a comparable study, all subjects with clear margins who received surgical excision enjoyed tumor-free survival. Despite postoperative radiation or corticosteroid therapy, recurrence was seen in cases of partial resection.

Inflammatory Pseudo Tumor (IPT) are clinic pathologically distinctive but biologically controversial entities, which have been described in the lungs, abdomen, retro peritoneum, and extremities, but rarely affect the head and neck region. It has been described under many appellations including plasma-cell granuloma, plasma-cell pseudo tumor, inflammatory myofibroblastic tumor, inflammatory fibro sarcoma, and most commonly IPT. Exact etiopathogenesis is not known, though it is considered to be an exaggerated inflammatory reaction to tissue injury of unknown cause. Though it has a benign clinical course, it is said that at least a subset of IPTs represents true neoplastic rather than reactive myofibroblastic proliferation.

A 26-year-old man presented with sensitivity in the upper-right back teeth since 1 year, accompanied by pain in the right side of face, forehead, and palate. Pain was severe, getting aggravated during the nighttime and relieved on medication. Diffuse swelling was evident producing mild bifacial asymmetry. On palpation, it was soft in consistency and tender. Right submandibular lymph nodes were also tender on palpation. On intraoral examination, palatal perforation was seen on the right side. Orthopantomograph revealed haziness of right maxillary sinus. Computed Tomography (CT) showed complete pacification of right maxillary sinus with erosion of medial wall and floor. A provisional diagnosis of aggressive lesion of maxillary sinus was considered.

Incisional biopsies revealed fascicles of spindle cells along with chronic inflammatory cell infiltrate predominantly plasma cells and lymphocytes. No fungal organisms were appreciated, both with periodic acid schiff PAS and silver staining. Spindle cells showed positive expression for vimentin and smooth muscle acting. These cells were negative for caldesmon and CD-68. The final diagnosis of inflammatory myofibroblastic tumor was confirmed. The lesion responded very well to corticosteroids and decreased in its size enormously. It was surgically excised, and on follow up, there is no recurrence since last 24 months.

Novel treatments, such as ALK inhibitors, are being studied in preclinical and clinical trials right now. For a minority of individuals with IMT, ALK-directed therapy may prove to be a highly beneficial treatment choice. However, based on current best practises, we urge that IMTs be treated with surgical resection and clear margins.

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