Case Report

Maxillary Sinus Inflammatory Myofibroblastic Tumors: A Review and Case Report

Chase C. Hansen, 1 Colby Eisenbach, 1 Carlos Torres, 1 Suzanne Graham, 2 and Fred Hardwicke 3

1 Department of Radiation Oncology, Texas Tech University Health Sciences Center, Lubbock, TX 79430, USA
2 Department of Pathology, Texas Tech University Health Sciences Center, Lubbock, TX 79430, USA
3 Department of Internal Medicine, Texas Tech University Health Sciences Center, Lubbock, TX 79430, USA

Correspondence should be addressed to Chase C. Hansen; chase.hansen@ttuhsc.edu

Received 5 December 2014; Accepted 30 January 2015

Academic Editor: Nurdan Tacyildiz

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An inflammatory myofibroblastic tumor (IMT) is an immunohistochemically diverse entity demonstrating neoplastic and nonneoplastic qualities first described by Brunn in 1939 [1, 2]. Until recently, the umbrella term “inflammatory pseudotumor” has been used to describe these lesions, which share a common histological appearance. A variable degree of spindle cell proliferation within a background of myxoid/collagenous stroma and a significant inflammatory infiltrate composed of lymphocytes, histiocytes, plasma cells, neutrophils, and eosinophils is descriptive of such lesions [3, 4].

Although the term inflammatory myofibroblastic tumor was coined by Umiker and Iverson in 1954 [2], the prominent histologic variance, erratic neoplastic, and inflammatory characteristics of these lesions have led to the development of a diverse nomenclature including, but not limited to, fibroxanthoma, plasma cell granuloma, histiocytoma, and inflammatory fibrosarcoma. Differences in cytological makeup and vast variations in the cytogenetic expression of immunohistochemical markers and inflammatory mediators present a challenge to providing accurate diagnosis, a process which has not been determined. Although IMTs can arise in any area of the body, lesions arising in certain sites, namely, the nasal cavity, paranasal sinuses, and pterygopalatine fossa, demonstrate a heightened neoplastic and invasive potential [5–12].

Treatment, in like manner to pathological diagnosis, is often a quandary for surgeons and oncologists alike. To date, complete surgical excision or corticosteroid therapies of IMTs are the gold standards of treatment [13]. However, in refractory, recurring or nonresectable cases, systemic and novel therapies have arisen, including chemotherapy, radiotherapy corticosteroids, NSAIDs, COX inhibitors, and kinase inhibitors [14, 15]. Despite case specific complete tumor regression and disease remission in response to such pharmacotherapeutics, a subset of IMTs remains resistant...
2. Case Presentation

A 34-year-old Hispanic female presented with complaints of progressive nasal congestion and mass in the right nasal cavity. Initial biopsy of the mass was felt to be benign. A year later, her right nasal mass recurred and again was reexcised with pathology reported as benign. A few months after her second surgery the mass had rapidly grown in size filling her mouth and causing right facial swelling (Figure 1(a)). She had difficulty eating and swallowing. CT scan of the neck showed an extensive destructive mass measuring 5.5 cm x 5.3 cm x 7.1 cm with extension into the medial wall of maxillary sinus and extending to the inferior orbit (Figures 2(a) and 2(b)). She had evidence of metastases to her right neck but there was no intracranial extension.

The lesion was biopsied and sent to two referral centers for assistance in making the diagnosis. The first referral center determined that the lesion consisted of a “polypoid sinonasal mucosa with ulceration, with acute and chronic inflammation and spindle cell proliferation, favoring reactive changes.” Immunohistochemical stains including smooth muscle antibody (SMA), desmin, S100, and keratin were all negative.

The patient went to second referral center for further treatment. At this institution, the report was slightly different, determining the sample to be consistent with an IMT, but found it had scattered positivity for ALK-1 and SMA within spindled cells. This is significant, because a diagnosis of IMT is quite difficult; however, one would expect samples from the same tumor biopsy to show similar SMA staining. The complexity of this tumor is further noted in that another biopsy, three years following the initial pathology report, differed from both of the previous reads.

Most recently, the second referral center reported immunohistochemistry stains demonstrating a profile different than the prior biopsies, with rare tumor positivity for SMA, and negativity for S100, pan-keratin, p63, and ALK-1. Next-Generation Sequencing- (NGS-) based analysis for detection of somatic mutations revealed TP53 and KRAS mutations. It was felt that pathology was consistent with inflammatory myofibroblastic tumor (Figures 3(a) and 3(b)). They decided to initiate therapy using crizotinib, an ALK-1 and ROS-1 inhibitor, given the refractory status of the tumor and prior positivity for ALK-1. Given the history, there was a chance this tumor would respond. Unfortunately, this patient did not respond to the targeted therapy and was eventually sent to her hometown hospital to seek further treatment due to unanticipated insurance complications.

In the beginning of July 2014, the patient was seen at our institution. At a multidisciplinary tumor board conference, it was decided that the best courses of action would be combination radiation and chemotherapy. She received a total dose of 60 Gy in 6 weeks using IMRT. She was also treated with ifosfamide, dacarbazine, and mesna as well as celecoxib. She recently completed four cycles of this regimen and a follow-up CT demonstrated overall tumor regression to a measurement of 4.9 cm at its greatest dimension as compared to 7.1 cm at initial presentation (Figures 1(b) and 2(c)).

3. Discussion

Since its first known appearance in medical literature in 1939, the inflammatory myofibroblastic tumor has garnered the attention of pathologists, surgeons, and oncologists, alike, due to its marked idiosyncrasies in immunohistology, pathophysiologic behavior, and therapeutic response [13]. In past years, a relative dogma has developed regarding the benign nature of IMT. Many cases have arisen, much like that presented above, which call such notions into question: thus, shedding light on the malignant and aggressive variant of this fascinating neoplasm. Due to the variable, and often case specific, behavior of many IMTs, it is fitting to delve somewhat into the generalities applied to this case and thereafter discuss the unique findings. The ultimate goal is to
Figure 2: (a) A computed tomographic scan of the maxillofacial region with contrast showing the maxillary IMT in axial section at initial presentation July 2014. (b) A coronal section of the same CT scan highlighting extent of invasion. (c) A CT scan of the head without contract to rule out an intracranial hemorrhage October 2014. Image demonstrates tumor regression when compared with (a) and (b).

Figure 3: Pathologic findings by hematoxylin and eosin staining of maxillary tumor tissue samples. (a) Whorled and fascicular spindle cells with moderate nuclear atypia and mitotic activity are heavily infiltrated by mixed chronic inflammatory infiltrate at 125x magnification. (b) Similar findings at 500x magnification.

advance the management and care of individuals affected by IMTs.

The definitive etiology of IMT has yet to be fully elucidated, but a great deal of proponents agree its development is largely multifactorial involving both inflammatory and chromosomal aberrations [16–23]. Undeniably, vast variations in the cytogenetic expression of immunohistochemical markers and inflammatory mediators present a challenge to providing accurate diagnosis and uncovering the concerted symphony that must take place for an IMT to develop.

To date, many cellular markers have been identified, including desmin, vimentin, smooth muscle actin, cytokeratin, and ALK-1 that aid in the pathologic diagnosis of IMT. Of particular interest is anaplastic lymphoma kinase or ALK-1 as well as its essential role in differentiating IMT from other spindle cell neoplasms that fall within the broad category of "inflammatory pseudotumors." According to studies conducted by Lawrence in 2000 and Coffin in 2001, greater than 50% of soft tissue IMTs possess chromosomal translocations involving the short arm of chromosome 2 and the ALK tyrosine kinase receptor locus [16–19, 24].

The diagnostic value of ALK-1 positivity is evident, considering most of the neoplastic counterparts of IMTs including desmoid fibromatosis, nodular fasciitis, calcifying fibrous tumor, myofibromatosis, and infantile fibrosarcoma are negative for ALK. Despite the obvious identification value of ALK-1 status in these neoplasms, the clinical and prognostic significance remains uncertain with some researchers suggesting unsubstantial value in ALK-1 status. However, adequate evidence exists to demonstrate the role of ALK-1 in conveying metastatic and invasive potential to IMTs. In fact, several recent retrospective studies have shown a marked increase in metastasis and recurrence in IMTs that are ALK-1 negative when compared to ALK-1 positive lesions [4, 25, 26].

In light of such information, ALK-1 negativity alone, as seen in the case presented above, demands an aggressive therapeutic approach and increased vigilance for distant metastases or local recurrence. Additionally, as with any
unregulated cell growth resulting in tumor formation, chromosomal aberrations leading to cellular atypia, nuclear pleomorphism, abnormal mitotic rate, DNA aneuploidy, and tumor suppressor inactivation have been shown to be beneficial in predicting IMTs [23]. This heightened neoplastic capability gives IMTs potential for particularly aggressive clinical behavior with local recurrence or malignant transformation.

Despite the IMT predilection for developing in the lung, abdominopelvic, retroperitoneal, and extremities in adolescent and pediatric populations, this rare tumor has been shown to occur in a vast age range and in a great number of locations [3, 24]. These include, but are not limited to, the orbit, liver, paranasal sinuses, and bladder [25, 27–30]. Just as certain cell signaling and chromosomal mutations enhance adverse disease behavior, a large degree of case specific evidence, in addition to that found in the case presented above, exists to support the idea that lesions arising in the nasal cavity, paranasal sinuses, and pterygopalatine fossa demonstrate a heightened neoplastic and invasive potential [5–12, 31]. Contiguous spread of such lesions has been shown to result in destruction of surrounding muscles, fat, nerves, and bone. Common presenting symptoms include nasal congestion, swelling, epistaxis, pain, parasthesias, proptosis, and headache [6, 7]. Large lesions, or those arising in areas not amenable to surgical removal, have been shown to have a higher degree of local recurrence and an increased risk for distant metastasis [32].

Of particular interest regarding the etiology of IMT is the role both the immune system and its host of inflammatory mediators play in the development and persistence of these rare entities [19, 20, 33–35]. Historically, IMTs were first described as benign, reactive, postinflammatory lesions arising primarily in children and adolescents [1, 2]. The variable degree of spindle cell proliferation within a background of myxoid/collagenous stroma and a variable inflammatory infiltrate is a testament to such a description.

In the face of such early evidence and the relatively consistent presentation profile within pediatric intrapulmonary lesions, no surprise IMTs have struggled to shed their recurrent inclusion into the misnomer of inflammatory pseudotumor, even in recent years. However, due to the work of Meis and Enzinger in 1990 and Coffin in 1995 involving intra-abdominal and retroperitoneal tumors [3, 28], the invasive and metastatic potential of the newly coined inflammatory myofibroblastic tumor was made evident. Thus, the dichotomy of inflammatory versus neoplastic behavior in IMTs was born. In recent years, the idea that one etiologic mechanism is involved has been largely abandoned and a multifactorial school of thought now predominates.

Just as erratic immunohistochemical characteristics seem to define IMT, differences in the cytological makeup of IMTs have been described and categorized into four cellular variants, largely modified from the three stromal classifications first described by Coffin et al. in 1995 [3, 4]. The fusion of these two classification systems yields the following cellular and stromal combinations:

1. Spindle cells within a vascularized myxoid stroma and an inflammatory infiltrate of neutrophils and eosinophils.
2. Compact spindle cells within a collagenized stroma and storiform architecture and an inflammatory infiltrate of plasma cells and lymphocytes often forming germinal centers.
3. Elongated spindle cells within a hypocellular highly collagenous stroma and a variable inflammatory infiltrate of lymphocytes, plasma cells, and eosinophils.
4. Lymphohistiocytic variant consisting of myofibroblastic spindle cells and foamy histiocytes. This is thought to represent the most inflammatory variant.

With regard to the case in question, it is important to point out the fact that significant overlap in cellular populations can occur and the phenomenon of maturation and zonation within a single tumor has been described: thus, adding to the complexity of affective histological classification and ultimate diagnosis [4]. Of interest, the occurrence of zonal expressivity and variable cellular differentiation within a single tumor has been shown to promote the need for multiple perioperative biopsies or complete sample procurement.

As for the case at hand, two prominent institutions obtained differing pathology reports and scattered reactivity of the previously discussed immunohistochemical markers within the same tumor sample before being seen at our institution. The occurrence of zonation and maturation explains this discrepancy. Additionally, and perhaps most importantly, the awareness of such a phenomenon within IMTs is paramount to successful diagnosis and treatment of inflammatory myofibroblastic tumors. It is likely that differential expression within the same IMT lesion explains disease resistance to some degree, as well.

Although the mainstay for successful treatment of IMT has been complete surgical excision, cases like that presented above often prove problematic for surgeon and oncologist, alike. Despite the difficulty, standard and novel pharmacotherapies, including NSAIDs, COX inhibitors, corticosteroids, and kinase inhibitors, are readily available for treatment of refractory, recurring, or nonresectable disease as evidenced in the literature on IMTs (Table 1). Variable reactivity to similar chemotherapeutic agents is common knowledge in IMT therapy. Nowhere is this more evident than in the previously presented case.

Recall that crizotinib, an ALK-1 inhibitor, was used by second referral center as therapy after unsuccessful surgical excision. This, no doubt, was initiated in hopes the scattered ALK positivity initially present in histologic sections would be inhibited, leading to regression and death of the tumor. The resistance of this particular lesion to such treatment, again, sheds light on the tremendous zonal and maturative expressivity profiles IMTs can possess. A second unique and fascinating characteristic of IMT is the often astounding regression when COX inhibitors like celecoxib and systemic steroids have been administered, a fact of which oncologists involved in the above case were well aware [14]. This highlights the vital role inflammatory mediators and immune...
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<th>Treatment</th>
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<tr>
<td>22/F</td>
<td>Epistaxis and protrusion of left eye</td>
<td>Paranasal sinuses and L orbit expansion</td>
<td>POS: vimentin, SMA, NEG: desmin, ALK-1</td>
<td>Resection, RT, CS, Chemo</td>
<td>Death</td>
<td>[32]</td>
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<tr>
<td>39/M</td>
<td>Nasal obstruction, supraorbital headaches</td>
<td>Vomer and ethmoid plate</td>
<td>NDA</td>
<td>Resection</td>
<td>24-month NED</td>
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<td>16/F</td>
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<td>Sinus walls</td>
<td>NDA</td>
<td>(1) CS (2) CS</td>
<td>(1) Initial regression; recurrence 2 months later; (2) 2.5-year NED</td>
<td>[10]</td>
</tr>
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<td>39/M</td>
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<td>Orbital floor</td>
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<td>27/F</td>
<td>R orbital swelling, trismus, diplopia</td>
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<td>POS: SMA</td>
<td>(1) Resection, (2) CS, (3) methotrexate</td>
<td>NDA</td>
<td>[11]</td>
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<td>29/M</td>
<td>L. facial numbness; swelling and pain in L. maxillary sinus and upper teeth</td>
<td>L. maxillary sinus and mild bony destruction</td>
<td>POS: vimentin, SMA, ALK-1 NEG: desmin, pancytokeratin, S100</td>
<td>NDA</td>
<td>NDA</td>
<td>[36]</td>
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<td>38/M</td>
<td>Headache, R exophthalmia, R 6th nerve palsy</td>
<td>Invasion of right cavernous sinus, sphenoidal sinus</td>
<td>POS: SMA NEG: ALK-1</td>
<td>(1) CS, (2) RT (20 Gy)</td>
<td>2-year NED</td>
<td>[12]</td>
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<td>NDA</td>
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<td>NDA</td>
<td>8 cases: 7+ vimentin; 5+ SMA, desmin; 2+ S-100</td>
<td>No recurrence in surgical patients</td>
<td></td>
<td>[37]</td>
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<td>88/M</td>
<td>Nasal obstruction and foul smelling discharge</td>
<td>Nasal septum, infraorbital wall, L maxillary antrum</td>
<td>NEG: melanocytic and epithelial markers</td>
<td>Resection</td>
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<td>[38]</td>
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<td>2/F</td>
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<td>NDA</td>
<td>NDA</td>
<td>Arterial embolization</td>
<td>5-year NED</td>
<td>[39]</td>
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<td>Pain in L maxillary molars, swelling of L. cheek, pulp necrosis of L 2nd molar</td>
<td>Lateral and superior L maxillary sinus walls</td>
<td>POS: SMA, b-catenin NEG: ALK-1, CD34</td>
<td>Resection</td>
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<td>26/M</td>
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<td>R medial wall and floor of maxillary sinus</td>
<td>POS: SMA, vimentin NEG: caldesmon, CD-68</td>
<td>(1) CS, (2) resection</td>
<td>(1) Regression, (2) 24-month NED</td>
<td>[41]</td>
</tr>
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<td>7/F</td>
<td>NDA</td>
<td>Expanding tumor without skull destruction</td>
<td>NDA</td>
<td>Resection</td>
<td>2-year NED</td>
<td>[42]</td>
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<tr>
<td>25/?</td>
<td>Pressure behind R eye, pain and swelling in R maxilla</td>
<td>NDA</td>
<td>NDA</td>
<td>Resection + CS (x2)</td>
<td>6-month NED</td>
<td>[43]</td>
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<td>63/?</td>
<td>Pain and swelling of L face, numbness</td>
<td>L posterolateral wall</td>
<td>POS: vimentin NEG: SMA, S100</td>
<td>Resection, RT (50 Gy), Chemo → recurrence; RT (50 Gy)</td>
<td>Death</td>
<td>[44]</td>
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<td>Age/sex</td>
<td>Presentation</td>
<td>MRI/CT reads</td>
<td>IHC stains</td>
<td>Treatment</td>
<td>Outcomes</td>
<td>Citation</td>
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<td>54/M</td>
<td>Swelling of L maxillary sinus and lower eyelid</td>
<td>Anterior maxillary sinus and infraorbital wall</td>
<td>POS: SMA, vimentin NEG: CD68, p53, S100</td>
<td>Resection → recurrence, resection</td>
<td>4-month NED</td>
<td>[45]</td>
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<td>64/F</td>
<td>Nasal obstruction, epistaxis</td>
<td>Medial sinus wall remodeling</td>
<td>NDA</td>
<td>Resection</td>
<td>24-month NED</td>
<td>[46]</td>
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<td>73/F</td>
<td>Vertigo, dysphagia, R retromolar swelling</td>
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<td>Incomplete resection + CS</td>
<td>Stable disease</td>
<td>[47]</td>
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<tr>
<td>6/F</td>
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<td>Maxilla</td>
<td>NDA</td>
<td>CS</td>
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<td>[48]</td>
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<td>42/F</td>
<td>Nasal polyps</td>
<td>Orbital floor, lateral sinus wall</td>
<td>NDA</td>
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<td>Progression</td>
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<td>41/M</td>
<td>Persistent necrotizing infections</td>
<td>Medial sinus wall</td>
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<td>NDA</td>
<td>NDA</td>
<td>[50]</td>
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<td>63/M</td>
<td>R facial pain, diplopia</td>
<td>Infraorbital wall, maxillary remodeling</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>[50]</td>
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<td>67/M</td>
<td>Epistaxis</td>
<td>Ethnomaxillary plate</td>
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<td>NDA</td>
<td>NDA</td>
<td>[50]</td>
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<tr>
<td>58/M</td>
<td>Epistaxis, L cheek swelling</td>
<td>Infraorbital wall</td>
<td>NDA</td>
<td>CS</td>
<td>1-month regression</td>
<td>[50]</td>
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<td>15/M</td>
<td>R eye pain, R facial swelling, trismus</td>
<td>Orbital floor, medial wall</td>
<td>(1) CS + RT, (2) resection</td>
<td>(1) Stable disease, (2) NDA</td>
<td>[50]</td>
<td></td>
</tr>
<tr>
<td>48/M</td>
<td>L nasal obstruction</td>
<td>Orbital floors, sinus remodeling</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>[50]</td>
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<tr>
<td>15/M</td>
<td>R eye pain, R facial swelling, epistaxis</td>
<td>Invasion of medial/lateral sinus walls</td>
<td>NDA</td>
<td>CS</td>
<td>2-month minimal regression</td>
<td>[51]</td>
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<td>32/F</td>
<td>Facial pain, R cheek fullness</td>
<td>Invasion of anterolateral sinus wall</td>
<td>NDA</td>
<td>Resection</td>
<td>1-month NED</td>
<td>[52]</td>
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<td>13/F</td>
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<td>Invasion of bone</td>
<td>NDA</td>
<td>CS + resection</td>
<td>33-month stable, residual disease</td>
<td>[53]</td>
</tr>
<tr>
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<td>NDA</td>
<td>No invasion evident</td>
<td>NDA</td>
<td>CS</td>
<td>Stable disease</td>
<td>[54]</td>
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<td>NDA</td>
<td>Sinus, orbit, anterior cranial fossa invasion</td>
<td>NDA</td>
<td>CS</td>
<td>Stable disease</td>
<td>[54]</td>
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<tr>
<td>NDA</td>
<td>NDA</td>
<td>Sinus, orbit, anterior cranial fossa invasion</td>
<td>NDA</td>
<td>CS</td>
<td>Stable disease</td>
<td>[54]</td>
</tr>
<tr>
<td>36/M</td>
<td>Obstruction, trismus</td>
<td>Lateral and posterior walls of nasopharynx</td>
<td>NDA</td>
<td>CS</td>
<td>7-month w/o symptoms, residual pain</td>
<td>[55]</td>
</tr>
<tr>
<td>18/M</td>
<td>Obstruction</td>
<td>Invasion into nasal septum and inferior turbinate</td>
<td>Polyclonal kappa and lambda light chains</td>
<td>(1) Resection → recurred, (2) RT (40 Gy)</td>
<td>27-month NED</td>
<td>[56]</td>
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<td>40/M</td>
<td>NDA</td>
<td>Nasal cavity, ethmoid sinus</td>
<td>NDA</td>
<td>Resection</td>
<td>1.5-month stable, residual disease</td>
<td>[57]</td>
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<td>83/M</td>
<td>NDA</td>
<td>Pterygomaxillary fossa</td>
<td>NDA</td>
<td>Resection</td>
<td>26-month NED</td>
<td>[58]</td>
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</table>
dysregulation play in the survival and growth of these lesions [20, 33–35, 60, 61]. Surely the remarkable susceptibility shown in some IMTs when compared with the above resistant case should lead us to question the current standard of surgical excision with corticosteroid adjuvant therapy, especially when approaching a case that demonstrates all of the hallmarks of likely resistance and the potential for invasion. In such cases, sufficient evidence exists to support radiation therapy as first line adjuvant therapy. Indeed, most IMT cases involving successful treatment with radiation were of a resistant, refractory, or recurring nature [8].

In the age of detailed cytogenetic analysis, refined imaging techniques, and precisely targeted therapeutic regimens a greater degree of time should be dedicated to discovering a unique cytogenetic “profile” for each IMT case. There is no denying this complex neoplasm demonstrates variation in the form of zonal expressivity, and overcoming this phenomenon will continue to be the challenge posed to all providers dealing with this rare tumor. Likewise, the role of specific cellular proteins such as ALK-1 will begin to serve as markers or the use of targeted therapy. Thus, physicians should emphasize effective determination of the cytogenetic profile of all IMTs, as well as a systematic and aggressive approach to IMTs presenting in areas shown to be refractory to many types of treatment, that is, paranasal sinuses. All things considered, the essential nature of taking a multidisciplinary approach with pathologist, surgeon, and medical and radiation oncologist providing concerted and comprehensive care is the foundation of proper IMT management.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

Acknowledgment

The authors thank Dr. Eduardo M. Diaz Jr., M.D., at MD Anderson Cancer Center, for his assistance in collecting the significant histology images for this case.

References


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Table 1: Continued.

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<td>67/M</td>
<td>Dysphagia</td>
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<td>CS</td>
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<td>63/F</td>
<td>Lower cheek swelling</td>
<td>Bone invasion of maxillary sinus</td>
<td>NDA</td>
<td>(1) RT (50 Gy), (2) CS, (3) cytoxan</td>
<td>Partial regression</td>
<td>[59]</td>
</tr>
<tr>
<td>55/M</td>
<td>Hypesthesia lower lip and jaw, progressive trismus</td>
<td>No bone or muscular invasion</td>
<td>NDA</td>
<td>Resection</td>
<td>1-year NED</td>
<td>[59]</td>
</tr>
</tbody>
</table>

NDA: no data available; RT: radiotherapy; NED: no evidence of disease; CS: corticosteroids.


[44] M. Karakok, E. Ozer, I. Sar et al., “Inflammatory myofibroblastic tumor (inflammatory pseudotumor) of the maxillary sinus mimicking malignancy: a case report of an unusual location (is...


