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Case Report

Low-grade myofibroblastic sarcoma of the head and neck region: a report of two cases of this extremely rare type of sarcoma and review of literature

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ABSTRACT

Sarcoma of the head and neck are rare and account of less than 1% of all tumors in head and neck region. Low grade myofibroblastic sarcomas (LGMFS) are even rare with only handful of cases ever reported in head and neck. We review 2 cases with LGMFS of maxilla and mandible. Retrospectively reviewed 2 patients who carried histological diagnosis of LGFMS. Previous clinic, operative and referral notes were reviewed along with histopathology slides and radiological studies. A 37-year-old female presented with recurrent right maxillary sinusitis and on CT scan was found to have an expanding mass in right maxillary sinusitis. Biopsy revealed it to be a LGMFS. She underwent total maxillectomy with orbital floor reconstruction, post operative radiation and is disease free for 18 months. A 49-year-old female presented with radiolucent mass on routine radiological examination. Initial enucleation and biopsy revealed it to be a LGMFS and was later treated with segmental mandibulectomy and fibular free graft reconstruction and post-operative radiotherapy. LGMFS of the bone is extremely rare tumor with only 8 cases being reported earlier from the skeletal system in entire body. Most common mode of presentation is asymptomatic mass found incidentally. These tumors are malignant with a rare propensity to metastasize distally. Hence prompt and accurate histological diagnosis followed by wide surgical excision with adjuvant therapy form important tenets of management.

Keywords: Sarcoma, Low grade myofibroblastic sarcomas, Head and neck cancers

INTRODUCTION

Head and neck cancers account for 3% of all the malignancies seen in the US and sarcoma of this region being uncommon represent only 1% of all the head and neck cancers. Myofibroblastic sarcoma is rare histological variant, first described by Vasudev et al but only recently diagnostic criteria were set by Menzel et al.[1,2] Most of LGMFS arise from soft tissue and only 9 cases have been described in English literature that arise primarily from bone.[3] Lack of obvious cytological atypia may result in their being mistaken for reactive fasciitis like lesions or fibromatosis and architectural similarities to fibrosarcoma or leiomyosarcoma may complicate the diagnostic process.[4] Despite these difficulties accurate histological diagnosis become imperative as this variant carries a better prognosis compared to other osseous sarcomas.[3] Herein we describe two cases of LGMFS arising from the bones of the facial skeleton and discuss the presentation, radiological and pathological findings along with treatment and prognosis.

CASE REPORT

Case 1

A 37-year-old white female presented with symptoms of right maxillary sinusitis. After treatment failure with a

course of antibiotics and steroid, a CT scan of the paranasal sinuses revealed the presence of a large expanding mass in the right maxillary sinus (Figure 1). MRI (Figure 2) later confirmed these findings and biopsy performed through a Caldwell Luc approach revealed the presence of an atypical spindle cell proliferation (Figure 3). Immunohistochemistry revealed that the neoplastic cells were focally positive for smooth muscle actin and negative for desmin, cytokeratin, S100, CD34 and CD68 consistent with LGMFS. A PET-CT scan demonstrated increased FDG uptake at the primary site and the absence of any distant metastases.

The patient underwent a right total maxillectomy with reconstruction of the orbital floor with a titanium mesh. Macroscopically the tumor was a 6-centimeter, smooth, white bulbous mass, occupying almost the entire right maxilla with invasion into adjacent tissues. The superior margin near the orbital floor was positive for tumor. The patient received 66Gy of adjuvant radiation therapy to the primary site. She is tumor free for 20 months.

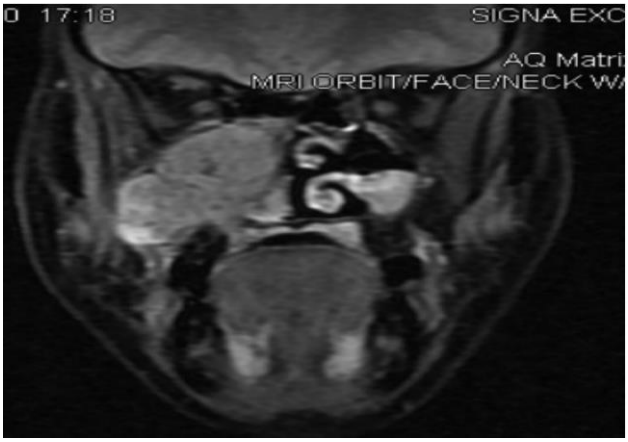

Figure 1: Pre operative MRI scan of case 1.

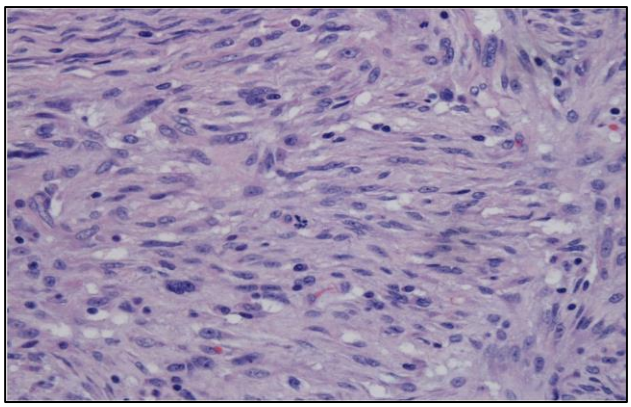

Figure 2: High powered histopathological view from case 1.

Case 2

A 44 year old white female presented with a painless intraoral mass which was suspicious for an odontogenic keratocsyt on dental x-rays. The mass was noted incidentally and was not associated with any symptoms or signs. Initial plain x-ray of the mandible revealed a radiolucent irregular mass in the right mandibular body near the angle (Figure 4). Intraoral biopsy revealed the presence monomorphic proliferation of hyperchromatic, spindled cells arranged in long fascicles, with areas of apparent bony destruction. Immunochemistry showed that the neoplastic cells were positive for smooth muscle actin but were negative for cytokeratin, epithelial membrane antigen and TLE-1consistent with LGMFS. A CT scan with contrast performed revealed the radiolucent mass in the right mandible near the angle with minimal invasion into the surrounding soft tissue. There was no clinical or radiographic evidence of lymphadenopathy. The patient was preoperatively staged asT1N0M0G1 stage-1A according to AJCC staging for bone tumors. The patient underwent a right hemi mandibulectomy with fibular free tissue transfer to reconstruct the mandibular defect. Macroscopically the tumor appeared as a soft gelatinous mass arising within the bone with extensions into adjacent soft tissues. Surgical margins were free of sarcoma. The patient underwent postoperative adjuvant radiotherapy, receiving 50.4 Gy in 28 fractions at 1.8 Gy each using 6 MV photons, custom blocking, and conformal therapy. The patient remains disease free 6 months.

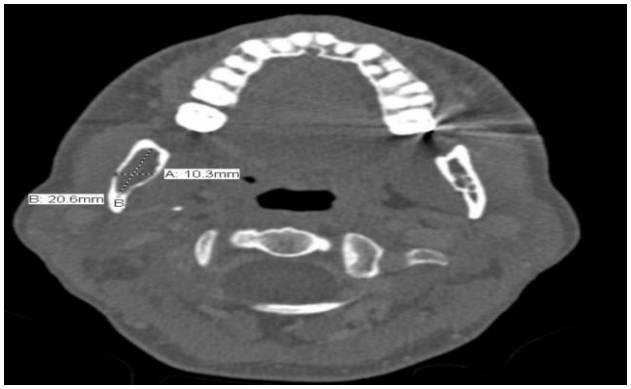

Figure 3: Pre operative CT scan of case 2.

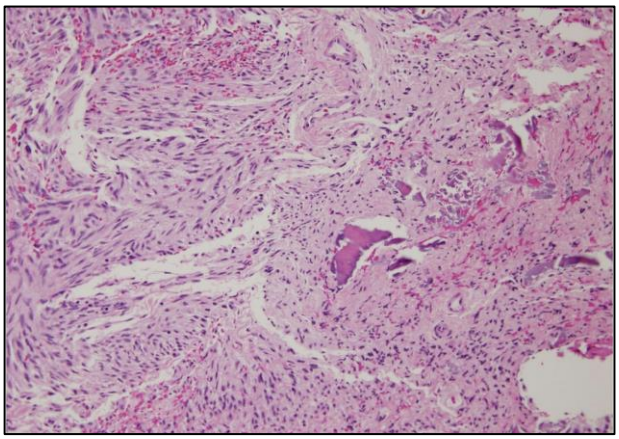

Figure 4: High powered histopathological view from case 2.

DISCUSSION

Myofiboblasts first described by Gabbiani are mesenchymal cells having characteristics of both fibroblasts and smooth cells that are present throughout the body. They secrete numerous cytokines, chemokines and growth factors that play an important role in organogenesis, oncogenesis, repair and fibrosis.[5] The function of myofibroblast in bone appears to be in nurturing stem cells and promoting hematopoiesis.[6]

LGMFS is a very rare spindle cell tumor arising from the myofibroblasts. Because of its rarity and low-grade histologic features make the diagnosis of LGMFS very challenging. Definitive diagnosis requires additional immunochemistry and occasionally electron microscopy. Differential diagnosis includes myofibroblastic sarcomas, inflammatory myofibroblastic tumors and nodular fasciitis.[4] Because of the wide prognostic difference among these conditions differentiating LGFMS from the related tumors becomes imperative.

Histological features of LFGMS include fascicular proliferation of atypical spindled shape cells. Immunohistochemistry reveals positivity to atleast one myogenic markers like smooth muscle actin, desmin, vementin or smooth muscle myosin heavy chain.[2] Ultra structural finding show spindle cells with elongated nucleoli and visible fibrillary centers.[7]

LGFMS most commonly arises from soft tissues and only 9 cases arising primarily from the bone in adult population have been described so far.[2] Only 3 being reported from bones of the facial skeleton. The most common mode of presentation described is a symptomatic enlarging mass, which is similar to the presentation of the disease in our patients. CT scan of both the patients showed an expanding mass in the involved bone with correspondingly increased metabolic activity on PET scan done in one patient. In the previously described report of LGMFS of the mandible, the patient presented with buccal edema and the CT scan showed an expansive lesion in the mandible and infiltrating the adjacent masseter muscle.[8] The radiological findings are nonspecific osteolytic lesion but indolent suggesting a low grade tumor. Both loco regional and distant metastases of LGMFS have been described, and imaging with whole body PET-CT could be recommended to identify them.[2,8]

Table 1 given account of other cases of LGFMS arising from the bone of the facial skeleton described so far.

Table 1: Other cases of LGFMS.

Site of tumor	Authors	Age (In years)/ gender	N	Size of tumor (cm)	Treatment
Mandible	Niedzielska et al[8]	54/M	1	5.9	Enucleation
Maxilla	Bisceglia et al[7]	24/M	1	4.0	Wide resection
Maxilla	Majno[9]	49/M	1	8.0	Wide resection + radiation

Both of our patients had wide local excision of the primary tumor followed by postoperative radiation therapy. Adjuvant form of therapy either radiation or chemotherapy has been advised most authors.[9] The role of adjuvant radiation had been described for sarcomas in general, with potential reduction in loco regional recurrence. This information has been extrapolated in the treatment of this unusual variant. There are no definite pathological or immunohistochemical features that can help predict the biological behavior of these tumors.[10] Given the rarity of this histological variant it would be difficult to determine if adjuvant therapy helps in loco regional or distant control and thereby the overall prognosis.

CONCLUSION

LGMFS is a very rare histological variant with has tendency for locoregional and distant metastases. Diagnosis is mainly rests on histological and immunochemistry evaluation. Wide local excision with adjuvant radiation has been the mainstay of treatment in these rare tumors.

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REFERENCES

1. Vasudev KS, Harris M. A sarcoma of myofibroblasts: an ultrastructural study. Arch Pathol Lab Med. 1978;102(4):185-8.
2. Mentzel T, Dry S, Katenkamp D, Fletcher CD. Low-grade myofibroblastic sarcoma: analysis of 18 cases in the spectrum of myofibroblastic tumors. Am J Surgical Pathol. 1998;22(10):1228-38.
3. Arora R, Gupta R, Sharma A, Dinda AK. A rare case of low-grade myofibroblastic sarcoma of the femur in a 38-year-old woman: a case report. J Med Case Rep. 2010;4:121.
4. Saïji E, Guillou L. Fibroblastic and myofibroblastic tumors of the head and neck. Ann Pathol. 2009;29(4):335-46.
5. Powell DW, Mifflin RC, Valentich JD. Myofibroblasts. I. Paracrine cells important in health and disease. Am J Physiol. 1999;277(1 Pt 1):C1-9.

6. Peehl DM, Sellers RG. Induction of smooth muscle cell phenotype in cultured human prostatic stromal cells. Exper Cell Res. 1997;232(2):208-15.
7. Bisceglia M, Tricarico N, Minenna P, Magro G, Pasquinelli G. Myofibrosarcoma of the upper jawbones: a clinicopathologic and ultrastructural study of two cases. Ultrastructural Pathol. 2001;25(5):385-97.
8. Niedzielska I, Janic T, Mrowiec B. Low-grade myofibroblastic sarcoma of the mandible: a case report Introduction Case presentation Case presentation. J Med Case Rep. 2009;3:8458.
9. Majno G. The story of the myofibroblasts. Am J Surgical Pathol. 1979;3(6):535-42.
10. Fernández-Aceñero M, Sanz-Laguna A, Carrascoso-Arran J, López-Criado P. Low-Grade Myofibroblastic Sarcoma Of The Bone. Internet J Pathol. 2005;4(1):195-9.

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