Malignant fibrous histiocytoma of maxillary sinus- a diagnostic challenge

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Abstract Malignant fibrous histiocytoma (MFH) is a malignant mesenchymal tumor uncommonly seen in head and neck region and even rarer in maxillary sinus with less than thirty cases reported in literature. Microscopic diagnosis at this rare site may be challenging because of the spectrum of features, which frequently overlap with other benign and malignant tumors. We herein report a case of malignant fibrous histiocytoma of the maxillary sinus in a 40-year-old man who presented with epistaxis. The initial biopsy appeared benign whereas histopathology of wide surgical excision was consistent with MFH. In view of the deep location, the diagnosis may be missed on a small biopsy, which can appear deceptive leading to diagnostic errors. This case report highlights the histopathological difficulties and pitfalls of this tumor at this rare site.

Keywords Malignant · Tumor · Maxillary sinus · Sarcoma

Introduction

Malignant fibrous histiocytoma (MFH) is a high grade soft tissue sarcoma which uncommonly involves upper aerodigestive tract and rarely maxillary sinus [1]. A case of MFH of maxillary sinus is described with emphasis on diag-

nostic difficulties. The patient was successfully treated with surgical excision and radiotherapy.

Case report

A 40-year-old man presented with history of epistaxis and left cheek swelling for five months. Intraorally, a fleshy growth involving whole of left buccal mucosa was seen extending up to superior and inferior gingivo-buccal sulcus. Small cervical lymph nodes were palpable at level Ia and Ib. Rest of general physical examination was normal. Patient was a chronic smoker but nonalcoholic. There was no history of trauma or irradiation. Both Computed tomography and Magnetic Resonance Imaging (MRI) of paranasal sinuses revealed a soft tissue mass measuring $6 \times 4.3 \times 4$ cm completely filling the left maxillary sinus extending into buccal space, pterygoid space, alveolar process and infratemporal fossa with bony destruction of medial, lateral, posterior and superior walls of maxillary sinus and erosion of the orbital floor. (Figure 1a) A diagnostic biopsy was performed which showed bland appearing spindle cells lacking atypia and mitosis and was suggestive of benign mesenchymal tumor. (Figure 1b) In view of intraosseous location and infiltrative nature, possibility of non-ossifying fibroma was considered and wider resection was advised. Patient underwent left total maxillectomy with supraomohyoid neck dissection. Grossly, maxillary bone with tumor measured $5 \times 5 \times 3.5$ cm. Cut surface was grey white, fleshy with infiltration of bone. Microscopy showed a variably cellular lesion composed of spindle cells intimately admixed with many chronic inflammatory cells. Focal storiform pattern was seen. Many areas showed bland looking spindle cells similar to biopsy. (Figure 1c)

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Fig. 1 a: MRI of paranasal sinuses: a large mass in the left maxillary sinus (*bold arrow*) extending into the buccal space, pterygoid space and erosion of the orbital floor. **b** Microscopy of biopsy showing bland appearing spindle cells admixed with inflammatory cells. (H&E, 10x). **c**

Histopathology of the main resected specimen showing foci of spindle cell lesion with bland appearing morphology similar to the initial biopsy (H&E, 10x). **d** Cellular areas with atypia and necrosis (*arrow*) (H&E, 20x). **e** Tumor infiltrating the bone (H&E, 10x)

Focal areas were more cellular with moderate to marked nuclear atypia. Increased mitoses including atypical mitoses and focal necrosis were present. (Figure 1d) Tumor was infiltrating into bone and soft tissue. (Figure 1e) Cervical lymph nodes were free. On immunohistochemistry, tumor cells were positive for Vimentin and focally for CD68. They were negative for SMA, S-100, myogenin, Anaplastic Lymphoma Kinase-1 (ALK-1) and anti-cytokeratin antibodies. Ki-67 shows increased proliferative activity (10–15 %). The findings were consistent with malignant fibrous histiocytoma. Patient underwent radiotherapy and is currently well at one year follow up without any recurrence.

Discussion

Malignant fibrous histiocytoma is a high-grade sarcoma first described by O'Brien and Stout occurring

commonly in extremities and trunk. [2] In current WHO classification, pleomorphic MFH is called undifferentiated pleomorphic sarcoma and is considered a diagnosis of exclusion. Incidence in head and neck region is low varying from 3 % to 10 % and affects sinonasal tract, craniofacial bones, larynx, and the soft tissues of neck. [3] No definitive etiology has been identified although role of previous radiation exposure, fracture and bone infarcts is implicated. [4] Involvement of the mandible, maxilla and maxillary sinus particularly intraosseous variety is uncommon. [3] It commonly presents with local swelling, nasal obstruction or discharge and epistaxis. Radiologically, soft-tissue invasion and bone destruction are common. [5] According to the current WHO classification. The microscopic subtypes include undifferentiated high-grade pleomorphic sarcoma, myxofibrosarcoma, undifferentiated pleomorphic sarcoma with giant cells and undifferentiated pleomorphic

Table 1 Diagnostic features of infiltrating fibrous lesions in head and neck

Characteristic	MFH	Dedifferentiated liposarcoma	Carcinoma	Lymphoma	Melanoma	Fibrosarcoma	Fibromatosis	IMT	Fibrous dysplasia	Osteosarcoma (MFH like variant)
Diagnostic histological feature	Mixture of fibroblasts and histiocyte-like cells	Atypical lipoblasts				Herringbone pattern, Occasional storiform pattern	Poorly circumscribed	Fascicles of bland spindle cells admixed with inflammatory infiltrate of plasma cells, lymphocytes.	Curvilinear trabeculae of woven bone; No osteoblastic rimming	Malignant osteoid (MFH like variant)
	Storiform pattern		Epithelioid/spindle tumor cells with bland stromal cells	Round to oval cells with scant cytoplasm and hyperchromatic nuclei	Nests of tumor cells Melanin	High cellularity	Paucicellular; Bland morphology		Hypocellular, fibroblastic stroma	Haphazardly arranged spindle cells
	Variable cellularity(at least focally high)						Atrophic and regenerating muscle fibers at periphery			
	Pleomorphic& multinucleated tumor cells									
Mitosis	+ Atypical + Moderate to severe	+ Atypical + Moderate to severe	+ Atypical + Moderate to severe	+ Atypical + Moderate to severe	+ Atypical + Moderate to severe	+ Atypical + Moderate to severe	Rare Atypical -	+ Atypical -	-	+ Atypical + Moderate to severe
Nuclear pleomorphism								Mild	Mild	
Necrosis	+	+	+	+	+	+/-	-	-	-	+
Infiltration	+	+	+	+	+	+	+	+	-	+
Specific IHC	Focal CD68+	MDM2, CDK4 +	CK+	LCA+	HMB45+	SMA focal +	SMA (variable)	SMA focal+ ALK-1(in some)		

IMT: Inflammatory myofibroblastic tumor; MFH: Malignant fibrous histiocytoma

sarcoma with prominent inflammation. The histopathologic diagnosis is characterized by admixture of fibroblastic and histiocyte-like cells. The cell morphology may vary greatly from well differentiated to anaplastic [4]. Thus, small biopsy and peripheral sampling may result in diagnostic pitfalls as seen in the present case. Correct diagnosis is usually possible on wider resections [6]. In the head and neck region, a wide variety of benign and malignant lesions enter the differentials which include other malignant mesenchymal tumors [4] spindle cell carcinoma, lymphomas and other infiltrating fibrous mesenchymal lesions (Table 1). In the present case, the infiltrative nature, marked pleomorphism, tumor giant cells, presence of necrosis and mitosis in resection specimen confirmed the malignant nature. Surgery with wide excision remains the principal treatment followed by radiotherapy. MFH of head and neck particularly in sinonasal tract is an aggressive tumor with relative worse outcome. Review of previously reported cases at this site shows frequently a poor prognosis despite radiotherapy with many of the patients dying from frequent local recurrences and early metastasis particularly to lungs and bones [4].

Definitive diagnosis can be made by biopsy, which should be taken at multiple sites to avoid misdiagnosis. [7] When evaluating small biopsies, radiologically destructive and invasive tumors should be viewed with caution as these may have a highly variable morphologic pattern with bland appearing morphology. MFH should be included in the differential diagnosis in

destructive masses of sinuses with recommendation of wide excision.

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