

Mast Cell Carcinoma of Maxillary Sinus

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Received: July 10, 2021; Published: July 21, 2021

Clinical Case

A 66 year old man presented to our ENT department with left chronic nasal obstruction and left jugal swelling without pain. The left jugal fossa was enlarged with a firm and unlimited mass 40X25 mm. Testing of trigeminal nerve (V) was normal. Nasal endoscopy showed deformity of left nasal lateral wall without endonasal tumor. Facial CT Scan revealed a soft tissue mass of left maxillary sinus with osteolysis (anterior pars of sinus). RMN a low intensity (T1) intra sinus mass enhancement after gadolinium. Biological evaluation note normochromic anemia (hemoglobin 8, 9 g/dl) and a normal WBC and platelet count, blood glucose 1, 05 g/l. The rest of the patient's workup liver and kidney function was negative. A Rouge Denker technique (sub labial) approach was chosen with tumor extirpation and medial maxillectomy. The diagnosis of mast cell sarcoma was confirmed after immunohistochemical who's expressed CD117 and CD 68. Radiotherapy (50 Gy) has been proposed. The follow-up is well after 14 month later.

Mast cell sarcoma is a rare form of mastocytosis characterized by destructive infiltration and metastatic potential. The most organ involved was bone (78%), gastro intestinal tract (37%), nodes (30%), skin (30%) spleen (26%), liver (20%).

A few cases of ENT localisation were reported: laryngeal (sub glottic), ear, tonsillar and lip.

Mast cell sarcoma is generally manifested by solid tumor and may be associated with fever, flushing, diarrhea and malaise.

Biological features note anemia and increased serum tryptase levels in the majority of cases. Imaging (CT and RMN) was performed to screening the tumor and loco regional extending. FDG-PET Scan can to be useful for diagnostic and therapeutic assessment [1].

Surgery is indicated especially in localized disease (total exision or debulking). Radiotherapy also has been proposed for localized disease 40 to 59 grays or in post operative with temporary efficacy. Corticosteroids improve clinical symptoms with transient effect. Interferon-alpha, cladribine, 2-chlorodeoxyadenosine, tyrosine kinase inhibitors and targeted therapies (imatinib, dasatinib, midostaurin, masitinib) may to procure positive response in some cases. Combination chemotherapy may to benefit in some patients [2, 3]. Prognosis is poor and mast cell leukemia is noted in 50% of cases.

Conflicts of Interest: No conflicts of interest regarding the publication of this article.



Figure 1: CT tumor of left maxillary sinus.

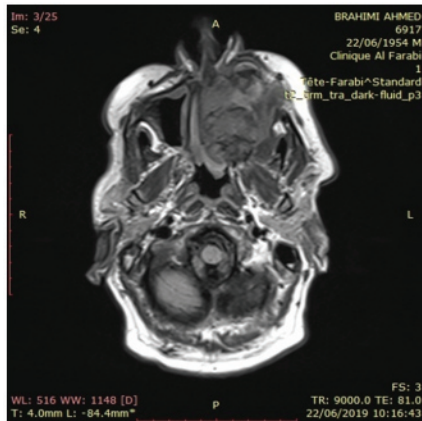


Figure 2: Rmn Iso Intense Mass of Left Maxillary Sinus.

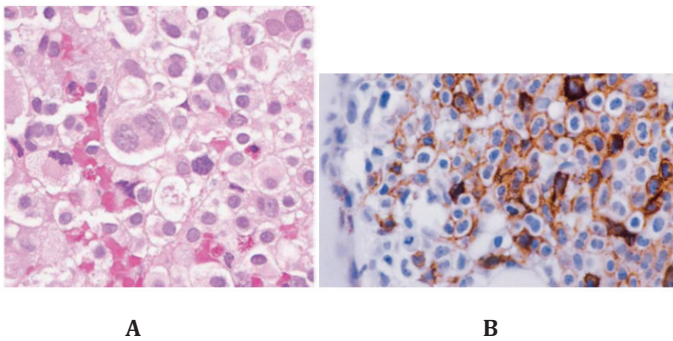


Figure 3: Mast cell sarcoma Histopathologic view (a).
Immunochemical test cd 117 (b).

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