

# Extramedullary Maxillary Sinus Plasmacytoma: A case report and clinical and radiological features

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## Citation

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## Abstract

### Objectives:

We report a late diagnosis of a rare case of extramedullary maxillary sinus plasmacytoma. It is important to investigate for nasty pathology if the patient has a long history of nasal bleeding even if he had a recent history of nasal surgery.

### Case report:

Although extramedullary maxillary sinus plasmacytoma is a rare presentation, the head and neck region is the most affected area and maxillary sinus is the most of head & neck to be involved. Its clinical and histological diagnosis is difficult. This is a reported case with its clinical, radiological and histological features.

### Conclusion:

If a patient presented with long history of recurrent nasal bleeding, nasty pathology should be excluded even if he had recent history of nasal surgery.

## INTRODUCTION

Plasmacytoma is a mass of neoplastic monoclonal plasma cells. It is classified into medullary if arised in bone or extramedullary if arised in soft tissue. Extramedullary plasmacytoma (EMP) represents 3% of all plasma cell tumors (1), 1% of all head and neck malignancies and 0.4% of upper respiratory tract malignancies (2). Eighty percent of them affect the head and neck (3). The main sites of involvement are the nasal cavity, paranasal sinuses, nasopharynx and the oral cavity. Men are affected 3 times more than women (4) with average age presentation of 60-70 years (5). Local lymph node affection occurs in 10 to 20 % of cases (6). It is reported that 15-20% of extramedullary plasmacytoma is converted to multiple myeloma (6). Metastasis occurs in 35 to 50% of cases (7). Its diagnosis depends on histology and by immunocytochemistry (8). EMP microscopically consists of sheets of plasma cells which may be monomorphous or pleomorphic (9). By immunohistochemical demonstration of one light chain monoclonal staining and one heavy chain class, most EMP can be differentiated from reactive plasma cell infiltrates with polyclonal staining (9). It is divided into 3 grades

according to its cellular atypia (10). Low, intermediate and high grade. Multiple myeloma should be excluded by serum and urine protein electrophoresis and immunoelectrophoresis, skeletal survey, bone scan and marrow biopsy (4). It should be differentiated from other destructive diseases in the maxillary sinus such as olfactory neuroblastoma, lymphoma, anaplastic carcinoma and metastatic tumours (11). Extramedullary plasmacytomas are highly radiosensitive, so radiotherapy is the best choice of treatment (1). Fifty percent of EMP patients survive more than 10 years (5). However, maxillary sinus EMP is associated with poorer prognosis than other sites of head and neck especially if it is associated with bone destruction (2). Long –term follow up is mandatory, as local recurrence and dissemination can occur many years after the original lesion has been treated (12). Chemotherapy may be added to treatment if there is recurrence or metastasis. This is a report of solitary EMP in the maxillary sinus in a 50-year old man.

## CASE REPORT

A 50-year old patient presented to Al-Jabr Hospital, ENT department in Al-Ahsa, Saudi Arabia with left nasal block and facial pain of one month duration and left facial swelling

and upper lid partial closure of 2 weeks duration in November 2007. He had history of nasal bleeding on and off during the last year after he had surgical correction for nasal trauma after car accident in another hospital. He is smoker working in Oil Company exposed to gaseous chemicals (H2S). No history of allergy, medical disease or drug intake.

On examination, he was a generally fit patient with blood pressure of 120/80, pulse of 80/minute. Chest and heart were clinically free. Nasal examination revealed left fleshy red mass attached to the lateral wall, bleeds on touch. Left cheek swelling with edematous left lower lid and left proptosis. On palpation the swelling was fleshy, painless and mobile under cheek skin. The left eye examination revealed incomplete eye closure with limited downward and inward mobility. There was also obliteration of the left upper buccoalveolus sulcus and bulging of the left hard palate with intact mucosa covering (Figure 1). There were no palpable lymph nodes in the neck.

**Figure 1**

Figure 1: Left hard palate bulging with intact mucosal covering



Hard palate bulging with intact mucosal covering

Ophthalmic examination showed no papilloedema, clear cornea, conjunctiva and pupils. His visual acuity was 6/6 on both eyes.

Blood investigations showed no abnormalities. Normal complete blood count.

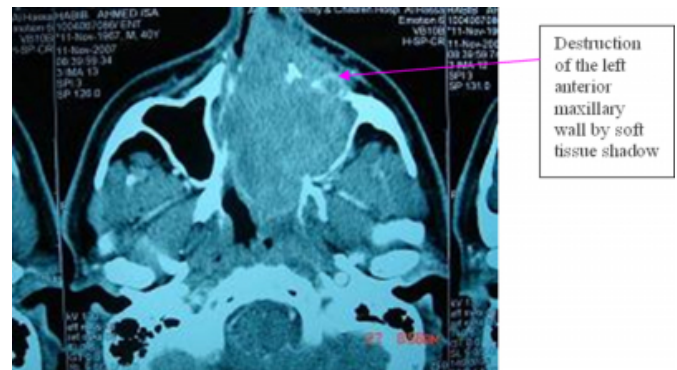
WBC 7000/mm, RBC 7.19x106/ml, Hb 14.5 gm%, MCV 66.1 LFL, MCH 22.2 LPL, MCHC 30.6 gm/dl, platelets 380000/mm, polymorphs 53%, lymphocytes 38%, monocytes 4%, eosinophils 5%. AST 41 HI, ALT 46, TBil 1.7, DBil 1, TP 68, ALB

Urine gravity was 1015; there was no protein in urine. Bence Jones protein test was requested and was not available in the laboratory. We checked it in tube & there was no protein. Protein electrophoresis was requested.

CT scan on nose and paranasal sinuses, coronal and axial sections, bone and soft tissue windows was done and revealed 5X5 cm soft tissue mass with mild to marked heterogeneous enhancement occupying the left maxillary sinus extending to the left ethmoid sinuses, left orbit destructing the lamina papyracea and filling the nasal cavity (Figures 2-4). It was also eroding the anterior wall of left maxillary sinus extending under the facial skin.

**Figure 2**

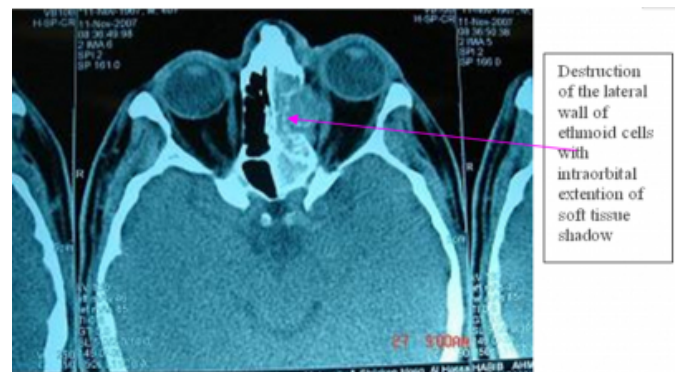
Figure 2: Axial CT shows soft tissue in the left maxillary sinus destructing medial wall, anterior wall and the septum



Destruction of the left anterior maxillary wall by soft tissue shadow

**Figure 3**

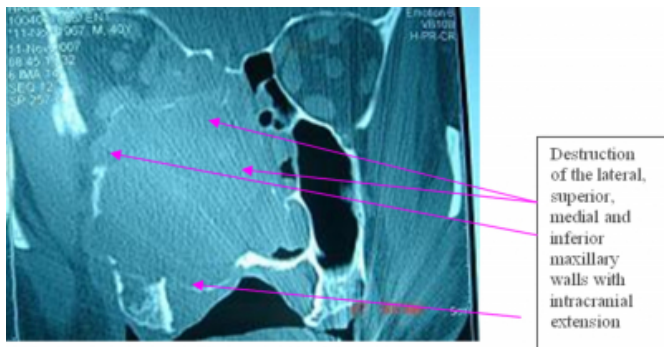
Figure 3: Axial CT shows the mass involving the left ethmoidal air cells destructing the lamina papyracea extending into the orbit pushing it antero-laterally



Destruction of the lateral wall of ethmoid cells with intraorbital extension of soft tissue shadow

**Figure 4**

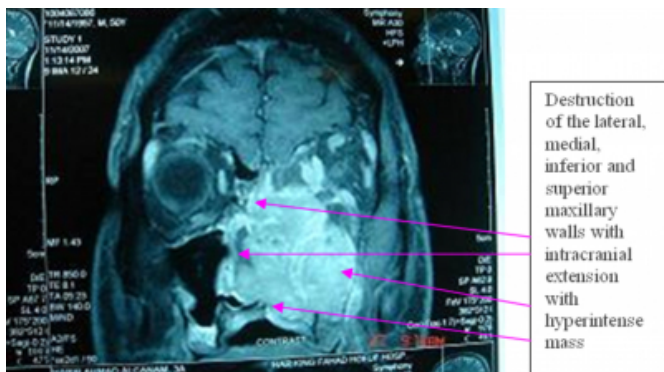
Figure 4: Coronal CT involving the left maxillary sinus destructing the medial wall, lateral wall and the septum and superiorly into the ethmoids and fovea ethmoidalis and medially into the orbit



MRI paranasal sinuses pre and post Gd DPTA was done and revealed a large expanding mass in the left maxillary sinus isointense at T1 and iso- to hyperintense in T2 with homogenous enhancement is seen after G injection. The lesion was seen extending medially occupying left nasal cavity obliterating it and perforating the nasal septum compressing the right nasal cavity with bowing the right maxillary sinus medial wall extending superiorly with orbital extension with mild left exophthalmos (Figures 5-6). It was also extending to the left ethmoid sinuses. It was extended inferiorly bowing the left side of the hard palate with erosion of the left side of the alveolus margin with extension posteriorly into the nasopharynx. Superadded sinusitis was seen involving sphenoid sinus and right maxillary sinus (bright T2 & low T1). The scanned parts of the brain showed no evidence of signal abnormalities with no evidence of metastasis.

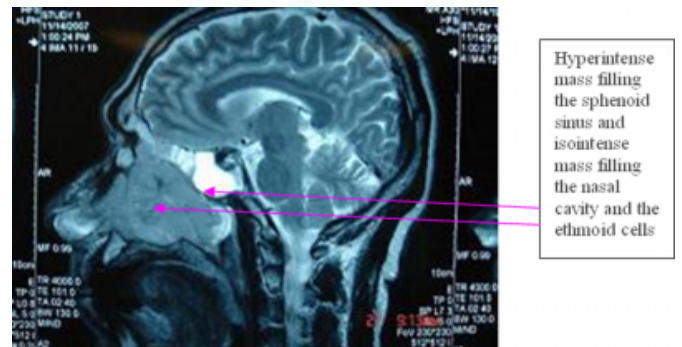
**Figure 5**

Figure 5: MRI, coronal section shows hyperintense mass destructing medial wall, superiorly into ethmoid sinuses and extending intracranially and intraorbitally and inferiorly into the hard palate and laterally into the cheek muscles.



**Figure 6**

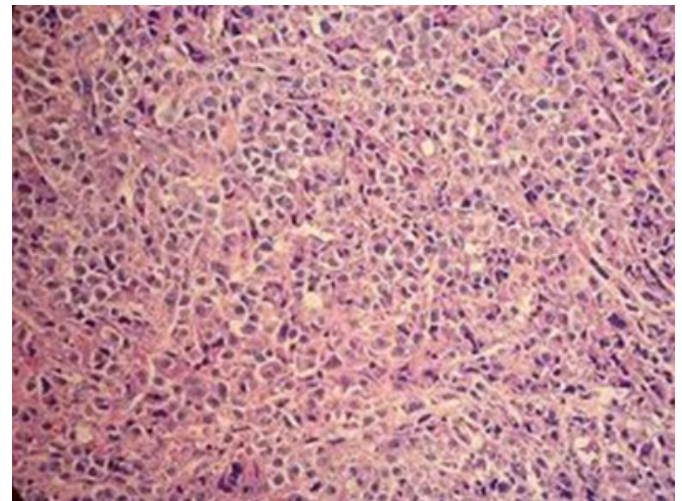
Figure 6: MRI sagittal section shows isointense mass extending into the anterior ethmoid air cells with hyperintense shadow in the sphenoid sinus and posterior ethmoid cells



Punch biopsy was taken from the fleshy nasal mass and was sent for histopathological analysis. The report came with diagnosis of extramedullary plasmacytoma. Histopathological examination showed tissue infiltrated by cells with eccentric nuclei, condensed nuclear chromatin with cart wheel appearance, inconspicuous nucleoli and abundant basophilic cytoplasm. There was absence of leukocytes (Figure 7).

**Figure 7**

Figure 7: photomicrograph shows tissue infiltrated by cells with eccentric nuclei, condensed nuclear chromatin, inconspicuous nucleoli and abundant cytoplasm

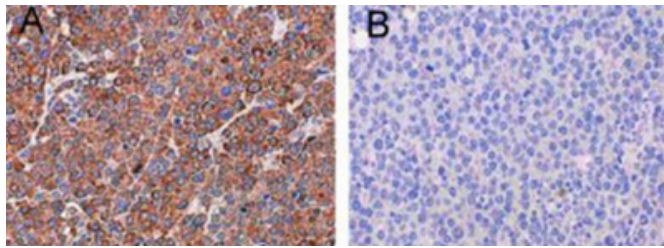


There was plasma cell dyscrasia. In immunohistochemistry, Paraffin embedded tissue with Anti- KAPPA and Anti- LAMBDA antibodies shows strong positivity in lambda light chain restricted population (Figure 8A) and negativity for Kappa (Figure 8B) which is consistent with plasmacytoma has been demonstrated.



**Figure 8**

Figure 8: Immunohistochemistry shows strong positivity in lambda light chain restricted population (A) and negativity for Kappa (B)



**DISCUSSION**

Schridde in 1905 was the first to describe extramedullary plasmacytoma. Eighty percent of the extramedullary plasmacytoma of the head and neck arise from the submucosa of upper aerodigestive tract (6). Solitary extramedullary plasmacytoma of the paranasal sinuses are uncommon and of B lymphocyte origin (3). To diagnose solitary EMP, one year of follow up from the original diagnosis should pass without evidence of skeletal radiological deposits as EMP may long precede occult but nevertheless disseminated myelomatosis (13). This is a rare case of left maxillary sinus extramedullary plasmacytoma. The patient presented with recurrent epistaxis for one year, nasal obstruction for one month and protrusion of his left eye with left facial swelling for 2 weeks. There was bulging of the hard palate of the same side due to invasion. The patient had history of trauma one year earlier to presentation with surgical correction. There was no history of pain or fatigue. Computed tomography revealed soft tissue swelling filling the maxillary sinus with erosion of its bony walls and extension laterally intraorbital, anteriorly subcutaneous, superiorly to ethmoid sinuses, medially perforating the septum extending to the opposite nasal cavity and inferiorly bulging the hard palate. On MRI, T1 and T2 cuts showed isointense swelling in T1 and hypointense in T2 filling the maxillary sinus extending to the ethmoid sinuses, orbit, subcutaneous with associated sinusitis in sphenoid sinus and opposite maxillary sinus. There was no evidence of intracranial extension. Microscopic examination of the biopsy revealed strong cytoplasmic staining for the plasma cell. On immunocytochemistry, lambda light chain restricted consistent with plasmacytoma has been demonstrated.

There was a case of right maxillary sinus extramedullary plasmacytoma in a 53-year old woman reported in 2002 (6). In this case, maxillary destruction was much less than in our case although the patient also presented with proptosis and

recurrent epistaxis. The authors did not present the MRI findings of the mass. The late discovery of the diagnosis of our case might be due to the thinking of physicians following the patient, the recurrent nosebleeds is related to the surgery performed to the patient earlier.

Gromer and Duvall reported 7 cases of EMP, 2 of them in the left maxillary sinus, one in the right maxillary sinus, 2 in the nasal cavity (12). One of them had local cervical node metastasis, another one converted to multiple myeloma and one with distant metastasis. Singh et al. reported 3 cases of EMP, one of them in the left maxillary sinus which recurred one year after total maxillectomy and one nasal case (13). There was report of a case of right maxillary and nasal cavity EMP destructing the bony structures in all directions with a history of trauma and surgery for the nose 6 months earlier to its presentation (14). It is same like our presenting case. Out of 22 cases of EMP reported in Birmingham in the period 1963- 1973, 2 cases involved the left maxillary sinus in which recurrence occurred 7 years after maxillectomy in one of them (15). Another report of 3 nasal cases of EMP was published one of them involved the ethmoid sinus and another one converted into multiple myeloma after one year (9).

**CONCLUSION**

This is a rare case of maxillary sinus extramedullary plasmacytoma. Computed tomography and MRI are presented. Any patient with history of recurrent epistaxis after surgery should be thoroughly investigated for nasty pathology.

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