

Primary Extramedullary Plasmacytoma of Maxillary Sinus

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ABSTRACT

A rare case of extramedullary plasmacytoma of maxillary sinus in a 58-year-old man presenting with right-sided nasal obstruction and intermittent blood tinged nasal crusting. Nasal endoscopy revealed that a dark red colored mass arising from the lateral wall of nose. A biopsy specimen was diagnosed as plasmacytoma (Kappa light chain type). Serum and urine electrophoresis failed to detect any myeloma component or Bence-Jones protein. All other screening tests to rule out multiple myeloma were negative. These findings confirmed the diagnosis of extramedullary plasmacytoma. The mass was completely excised by lateral rhinotomy approach. No recurrence has been noted at the end of 6 months follow-up.

Keywords: Extramedullary plasmacytoma, Kappa light chain, Lateral rhinotomy, Multiple myeloma.

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INTRODUCTION

Plasmacytomas are malignant tumors of monoclonal plasma cells. They originate either in bone (solitary osseous plasmacytoma) or in soft tissues (extramedullary plasmacytoma).¹ Extramedullary plasmacytoma is a rare localized tumor. It has no characteristic of multiple myeloma, but the development of multiple myeloma has been noted in 8 to 36% of patients.^{2,3}

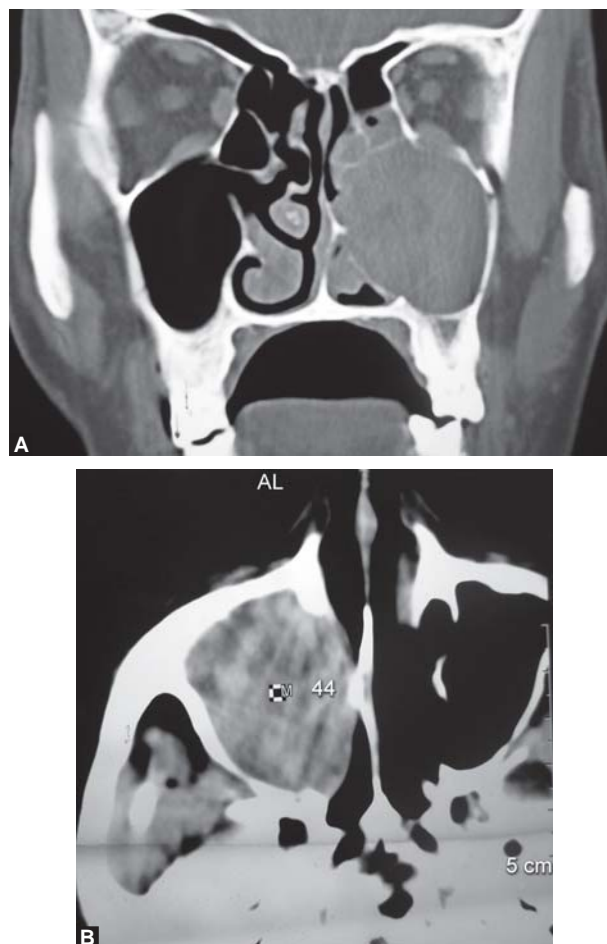
Nearly 80% of all extramedullary plasmacytomas occur in head and neck; most of these arise in upper aerodigestive tract, probably because of the abundance of the lymphatic tissues in this area.^{4,5} Extramedullary plasmacytomas represent less than 1% of all head and neck malignancies⁶ and it accounts for 4% of all nonepithelial tumors of the nasal cavity, nasopharynx and paranasal sinuses.⁷ We report a rare case of extramedullary plasmacytoma arising from the maxillary antrum.

CASE REPORT

A 58-year-old male attended ENT OPD with history of right-sided nasal obstruction and intermittent blood tinged nasal discharge since 6 months and mild proptosis. Nasal endoscopy revealed a dark red colored mass arising from the lateral wall of right side nasal cavity. Left nasal cavity and nasopharynx were normal. No palpable cervical lymph nodes. Computed tomographic (CT) scan of PNS revealed a heterogeneous moderately enhancing soft tissue, density

lesion involving right side maxillary sinus extending to right side nasal cavity, ethmoid sinus and orbital floor (Figs 1A and B). Few areas of bony erosion are seen in floor of orbital wall, inferior turbinate, middle turbinate. Biopsy was taken through nasal endoscopy which on histopathological examination reported to be plasmacytoma (Fig. 2).

Further investigations were undertaken to look for multiple myeloma. These investigations included a CBC, serum calcium and creatinine levels, serum protein electrophoresis and immunoelectrophoresis. Urine analysis for Bence-Jones protein, evaluation of β_2 microglobulin, a skeletal survey with plain films (skull, ribs, humerus and femur), abdominal ultrasonography, bone marrow aspiration and biopsy. All findings were within normal limits. Accordingly, we ruled out multiple myeloma and this confirmed diagnosis of primary extramedullary plasmacytomas of maxillary sinus.



Figs 1A and B: Computed tomographic (CT) scan of PNS showing a heterogeneous moderately enhancing soft tissue, density lesion involving right side maxillary sinus extending to right side nasal cavity, ethmoid sinus and orbital floor

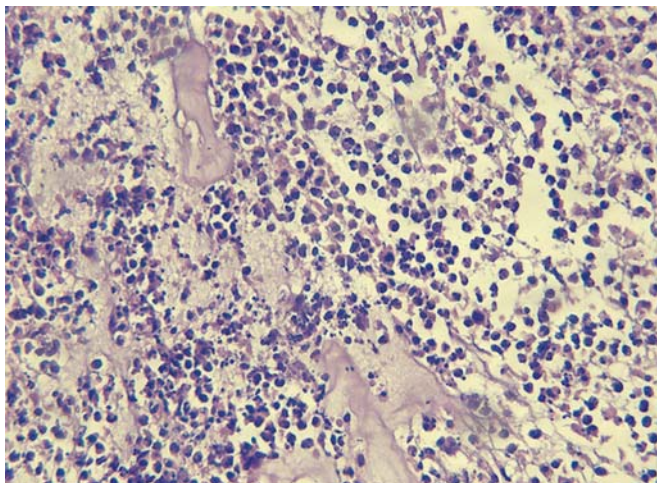


Fig. 2: HP showing sheet like proliferation of plasma cells of varying maturity and atypia and an eccentrically placed nucleus

The mass was excised under general anesthesia by lateral rhinotomy approach. Ant wall of maxilla was removed. Mass was completely removed from maxillary antrum. Mass had eroded the bony orbital floor. However, periosteum was found to be intact. Posterior wall of maxillary antrum was found to be partially deficient. Middle turbinate and superior turbinate were excised. The excised mass was sent for HP study. HP shows sheet like proliferation of plasma cells of varying maturity and atypia and an eccentrically placed nucleus. Immunohistochemical staining demonstrated the monoclonal nature of plasma cells. Patient is now under follow-up. At 6 months no recurrence has been noted.

DISCUSSION

The plasma cell dyscrasias are characterized by the clonal proliferation of plasma cells that produces a homogeneous immunoglobulin protein. The major plasma cell dyscrasias are as follows:

1. Multiple myeloma
2. Monoclonal gammopathy of unknown significance (MGUS)
3. Solitary plasmacytoma of bone
4. Extramedullary plasmacytoma and
5. Heavy chain disorders.⁸

Extramedullary plasmacytoma is a rare soft tissue malignant neoplasm that is made up of monoclonal plasma cells. It can be either primary (without evidence of disease in other foci) or part of a systemic process during the course of multiple myeloma.

The first case of extramedullary plasmacytoma was reported in 1905 by Scrida.⁹ Between then and 1997, more than 400 articles addressing extramedullary plasmacytoma

were published. In 1999, Alexiou et al reviewed all previous reports of extramedullary plasmacytoma and found 869 cases; 714 (82.2%)⁴ of them had occurred in the upper aerodigestive tract. The most frequently affected areas in the upper aerodigestive tract are the nasal cavity and PNS (43.8%) followed by the nasopharynx (18.3%), the oropharynx (17.8%) and the larynx (11.1%).⁴

Extramedullary plasmacytoma is more common in males (ratio 4:1) and 95% of cases occur in patients older than 40 years.¹⁰ Extramedullary plasmacytoma of the upper aerodigestive tract are most often occur as single lesions.^{1,8}

The clinical presentation may vary according to the site of involvement. In sinonasal or nasopharyngeal involvement, the most common symptoms are mass, nasal obstruction and epistaxis.¹¹ Other reported presenting symptoms include nasal discharge, pain, proptosis, cervical lymphadenopathy and cranial nerve palsy.¹⁰

The diagnosis of extramedullary plasmacytomas is usually after HP study. In addition, immunohistochemical study is used to differentiate extramedullary plasmacytomas from benign reactive plasmacytomas as well as other malignant disorders, such as undifferentiated carcinoma, melanoma and esthesioneuroblastoma.¹²

When a plasmacytoma is confirmed histologically, secondary diagnostic procedures must be carried to exclude systematic involvement.⁸

Malignant tumors of the nasal cavity (including extramedullary plasmacytomas) are treated by surgery and/or radiotherapy. Although extramedullary plasmacytoma is known to be radio sensitive, most authors recommended a combination of surgery and radiotherapy for nasal cavity lesions.¹³

The lesion in our patient was localized in the maxillary antrum and there was no sign of invasion into the underlying bone periosteum. We did not consider postoperative radiotherapy.

Local recurrence has been reported to occur in 8 to 30% of adequately treated cases within 3 to 61 months.^{2,3} Conversion of extramedullary plasmacytoma to multiple myeloma has been reported on 8 to 36% of cases. Therefore, long-term follow-up including both local and systemic surveillance is necessary. CT and measurement of serum immunoglobulin and Bence-Jones protein levels may be useful in detecting recurrence or conversion to multiple myeloma.

Although, several reports regarding extramedullary plasmacytoma in the nasal cavity have been published, a finding of such a lesion in the maxillary antrum is quite rare.

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