

Extramedullary Plasmacytoma: An Unusual Differential Diagnosis Of Nasal Polyp

A Jain, H Chopra, K Dua

Citation

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Abstract

Extramedullary plasmacytomas are neoplasms of plasma cell origin, representing about 0.4% of all head and neck malignancies. We are presenting a patient who was initially diagnosed as glaucoma for complaint of blurring of vision. He underwent glaucoma surgery but did not respond. He later developed proptosis, nasal obstruction, few episodes of epistaxis. Nasal endoscopy revealed grayish polypoidal mass involving left middle meatus, choana. Clinicoradiological and histopathological workup revealed an extramedullary plasmacytoma involving ethmoids, orbit with intracranial extension. Correct diagnosis was achieved one year after the initial complaints of patient, due to delay in otolaryngology consultation and radiological workup. ENT and eye surgeons should keep this entity in the list of differential diagnosis of proptosis with nasal polyp. These patients require multidisciplinary approach for proper management.

INTRODUCTION

Extramedullary plasmacytomas are rare neoplastic lesions characterized by monoclonal proliferation of plasma cells. Eighty percent of extramedullary plasmacytomas are localized in the submucosa of the upper respiratory tract.¹ They are most commonly seen in patients between 50 and 60 years of age, more frequent in men.² The most common locations in the head and neck are the nasal cavity, nasopharynx and paranasal sinuses.³ These tumors have been reported to occur simultaneously at different sites in the head and neck in 10 to 20% of patients.⁴ Symptoms usually include epistaxis, nasal obstruction, nasal discharge, headache, or local pain.⁵ But it is unusual for them to present as diminution of vision, proptosis and nasal polyp. We are describing a patient of extramedullary plasmacytoma who was initially suspected as having glaucoma, but after further investigations correct diagnosis was made.

CASE REPORT

A sixty year old male patient presented in the otolaryngology clinic with complaints of decreased vision of left eye (since 1 year), forward protrusion of left eye (since 1 year), left sided nasal obstruction (since 3 months), few episodes of left sided nasal bleed (since 2 months). Anterior rhinoscopy revealed grayish, polypoidal, non sensitive, friable mass in the left sided middle meatus with mucopurulent discharge in inferior meatus. Mass was

attached superolaterally. In posterior rhinoscopy mass was reaching till choana with normal nasopharynx. Vision was 6/60 in left eye. Right eye was normal. Nasal endoscopy confirmed above findings, mass was soft in consistency, bleeding profusely. Biopsy was taken and sent for histopathological review. MRI (paranasal sinuses & orbit) showed a contrast enhancing mass involving left side anterior, posterior ethmoids, anterior & middle cranial fossa extension with retrobulbar orbital extension. Lamina papyracea and cribriform plate was eroded. His lab reports revealed the following: Hb: 8.7g%, TLC: 8700/cc, DLC: P69, L26, M4, platelets $328 \times 10^3 / L$, ESR 140 mm/hr, Blood Urea: 45mg%, S. creatinine 1.10mg%, S. uric acid 5.1 mg/dL, S. calcium 8.46 mg/dL, total protein 12.50 g/dL, S. albumin 3.32 g/dL, gamma globulin 7.21 g/dL, A:G ratio 0.36. Fungal stains were negative. Histopathological examination of biopsy specimen showed abundant plasma cells suggestive of plasmacytoma. Bone marrow aspiration and biopsy was normal. Serum electrophoresis revealed M band with beta fraction 47.4%. Final diagnosis was extramedullary plasmacytoma. Oncology consultation was taken and patient was started on chemotherapy (oral cyclophosphamide). After one month of therapy there was 40% reduction in tumour size. Therefore patient was planned for further two courses of chemotherapy.

Figure 1

Legend for photograph 1: MRI paranasal sinus (axial section) – Plasmacytoma involving anterior, middle cranial fossa and left orbit.



Figure 2

Legend for photograph 2: MRI paranasal sinus (sagittal section) – Retroorbital extension



DISCUSSION

Dalrymple and Bence Jones in 1846 described neoplastic proliferation of plasma cells. They described a disseminated neoplastic proliferation of plasma cells that was characterised by marked proteinuria and bone pain.⁶ Plasma cell tumours are classified into three categories; multiple myeloma, medullary plasmacytoma and extramedullary plasmacytoma. Medullary plasmacytomas, progresses to multiple myeloma in 85% of patients, extramedullary plasmacytomas usually remain well localised. Only 15% to 20% of cases progress to multiple myeloma or develop regional disease. For the definition of primary extramedullary plasmacytoma, a normal radiographic survey in the absence of bone marrow infiltration is required.⁷ It is essential to rule out multiple myeloma, when evaluating a plasma-cell neoplasm which is done with a normal radiographic skeletal survey, negative serum and urine immunoelectrophoresis, and negative bone marrow biopsy (<5% plasma cells). Determination of clonality of the lesion is felt to be important, as polyclonal EMP tends not to progress, whereas monoclonal EMP can be followed immunologically.⁸

The most common locations in the head and neck are the nasal cavity, nasopharynx, maxillary sinus, thyroid gland, soft tissues of the anterior cervical region, parotid gland, tonsil, oropharynx, larynx, orbit, and choroids of the eye and

eyelid. Cases of solitary bone plasmacytoma have been reported in the sphenoid bone, mastoid, calvaria and skull vault, hyoid bone, temporomandibular joint, maxilla, and mandible. Wiltshaw suggested that bony lesions developed in 81% of patients with disseminated extramedullary plasmacytomas.⁴

The treatment of localized extramedullary plasmacytoma is still a matter of some debate; some clinicians favor radiotherapy,^{4,9} others prefer surgical management,¹⁰. Chemotherapy is advisable only for patients who have a generalized extramedullary plasmacytoma. In our case due to orbital involvement patient was started on chemotherapy. One month of therapy resulted in remarkable improvement. Long term follow up is required to monitor for development of multiple myeloma. Such patient should be managed by multidisciplinary approach.

It is stressed that whenever a patient presents with diminution of vision, proptosis and nasal complaints, otolaryngologist opinion should be taken. There should not be delay in radiological investigations. This will help in clinching diagnosis in early stage of disease.

CONCLUSION

A systematic method of staging this disease--including measurements of the complete blood count, renal and liver function, serum and urinary protein electrophoresis, serum immunoglobulin levels, skeletal survey, bone marrow examination and CT of the tumor region--must be performed in order to exclude systemic involvement before the diagnosis can be made. Patients require lifelong monitoring for local or systemic recurrence, or development of multiple myeloma. The optimal therapy for patients with an

extramedullary plasmacytoma of the head and neck can be achieved only by an multidisciplinary approach.

CORRESPONDENCE TO

Dr Kapil Dua 2190, Krishna Nagar, Near Aarti Chowk, Ludhiana Punjab, India Pin Code 141001 Phone No. + 91 9815544207 E mail: amitkgmc7777@yahoo.com

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Author Information

Amit Kumar Jain, M.S.

Senior Resident, Department Of ENT, Dayanand Medical College and Hospital

Hemant Chopra, M.S.

Professor and Head, Department Of ENT, Dayanand Medical College and Hospital

Kapil Dua, M.S.

Assistant Professor, Department Of ENT, Dayanand Medical College and Hospital