

Extramedullary Plasmacytoma of Nasal Cavity: A Rare Entity

¹Digvijay Singh Rawat, ²Mohnish Grover, ³PC Verma

¹Junior Resident, Department of ENT, SMS Medical College, Jaipur, Rajasthan, India

²Assistant Professor, Department of ENT, SMS Medical College, Jaipur, Rajasthan, India

³Professor, Department of ENT, SMS Medical College, Jaipur, Rajasthan, India

Correspondence: Mohnish Grover, Assistant Professor, Department of ENT, SMS Medical College, Jaipur, Rajasthan, India
e-mail: drmohnish_aiims@rediffmail.com

Abstract

Extramedullary plasmacytoma is a rare plasma cell proliferative disorder arising outside the bone marrow. It shows a predilection for the head and neck region. They commonly involve the mucosa associated lymphoid tissues of upper airways. Common sites are nasal cavities and paranasal sinuses followed by nasopharynx, tonsils and oropharynx. We report a case of an isolated lesion in the right nasal cavity in an elderly male in which surgical excision is followed by postoperative radiotherapy.

Keywords: Nasal cavity, extramedullary, plasmacytoma.

INTRODUCTION

Depending on their site of development and clinical features, malignant tumors of monoclonal antibody—producing plasma cells are classified as multiple myelomas, solitary plasmacytomas, or extramedullary plasmacytomas. Extramedullary plasmacytoma is a rare localized plasma cell tumor originating in the soft tissues.¹ Almost 80% of extramedullary plasmacytomas occur in submucosal lymphoid tissue in the head and neck. Extramedullary plasmacytoma tends to occur in sixth and seventh decade of life, and it is three to four times more common in men than in women.² The treatment of extramedullary plasmacytoma is radiotherapy. The prognosis depends on tumour size (> 5 cm) and nodal involvement. The 10-year survival rate is 50 to 80%. We are presenting a case of extramedullary plasmacytoma in an 85 years old otherwise healthy male.

CASE REPORT

An 85-year old man presented with a history of right side nasal mass for last two months, slowly increasing in size, with two episodes of minor bleeding. On anterior rhinoscopy there was reddish pink mass in right nasal cavity completely occluding the nasal cavity on right side and septum was pushed to the left side. On probing the mass was arising from the floor of the right nasal cavity. Oral cavity examination revealed bulge in right side of hard palate. No palpable neck node was there.

CT scan of the nasal cavity and paranasal sinuses showed a mass 5 cm × 4 cm × 3 cm in the right nasal cavity (Fig. 1). Biopsy was taken which showed undifferentiated malignant round cells. We performed endoscopic excision of the mass. Histopathological examination of the biopsy specimen came out to be undifferentiated large round cell tumor (Fig. 2). On marker studies the tumor cell expressed CD 138, CD 79a and CD 56 and are kappa light chain restricted (Fig. 3). The tumor cells were immunonegative for CD 20 (a pan-B-cell marker), CD 3 (a pan-T-cell marker) and cytokeratin; no heavy chain expression was detected. Marker study suggested the mass to be extramedullary plasmacytoma. A bone marrow biopsy and 99m technetium scintigraphy did not detect any systemic lesion. Assays for Bence Jones protein in the urine and for serum myeloma protein were negative. Patient was sent for local radiotherapy and was given 40 Gy in 20 settings. Follow up at 6 months showed no recurrence.

DISCUSSION

Plasma cells are mature immunocompetent cells derived from B lymphocytes, and they provide specific noncellular immunity within the immune system. They produce specific antibodies against antigens in different tissues. Large populations are seen in the mucosa of the nasal and paranasal sinuses. Plasmacytomas arises from a neoplastic

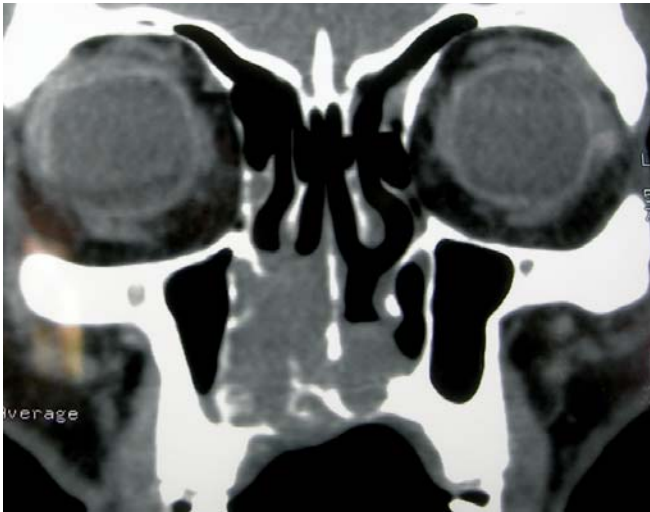


Figure 1: CT PNS – soft tissue density mass in right nasal cavity extending to maxillary sinus and causing bulge in hard palate

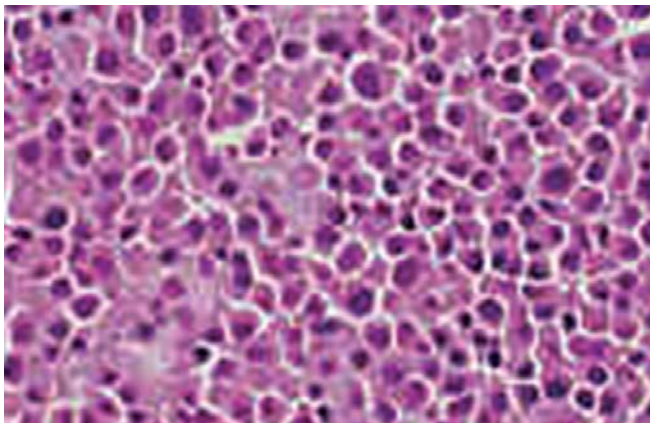


Figure 2: HPE – round cell tumor

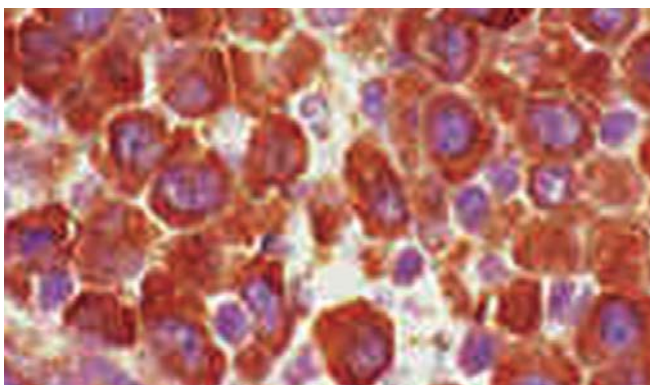


Figure 3: Immunohistochemistry: positive for kappa light chains

proliferation of these antibody—producing plasma cells.³ The three types of plasmacytomas – multiple myelomas, solitary plasmacytomas and extramedullary plasmacytomas are further classified as either localized (stage I), localized

including local lymph nodes (stage II), or generalized (stage III) according to the clinical manifestations.⁴

The first case of extramedullary plasmacytoma was reported in 1905 by Schridde. Extramedullary plasmacytoma is more commonly seen in males (4:1), and 95% of cases occur in patient older than 40 years.⁵ Extramedullary plasmacytomas usually present in the head and neck region. Approximately 80% of cases occur in the mucosa associated lymphoid tissue of the upper respiratory tract where the most frequently involved sites are nose and paranasal sinuses, followed by nasopharynx, tonsils and oropharynx.⁶ Extramedullary plasmacytoma of upper aerodigestive tract usually present as a submucosal, reddish, sessile or polypoidal and are usually solitary tumors that rarely ulcerate.⁷ A systemic method of staging of the disease – including measurement of the complete blood count, renal and liver functions, serum and urinary protein electrophoresis, and serum immunoglobulin levels; a skeletal survey and bone marrow examination; and computerized tomography of the tumor region – must be performed in order to exclude systemic involvement before the diagnosis can be made.³ Immunohistochemical staining will demonstrate the monoclonal nature of the plasma cells and confirm the neoplastic nature of the lesion. In addition, immunohistochemistry study is used to differentiate extramedullary plasmacytoma from benign reactive plasmacytosis as well as other malignant disorders, such as undifferentiated carcinoma, melanoma and esthesioneuroblastoma.⁸ The British society for hematology has issued a set of guidelines for the diagnosis and treatment of extramedullary plasmacytoma (Table 1). The society recommends initial radiation treatment with 40 Gy in 20 fractions with a 2 cm margin for tumors smaller than 5 cm, and 50 Gy in 25 fractions for larger tumors. Regional cervical lymph nodes should be irradiated only if they are directly involved or if there is a high-risk of spread.⁹ 5-year survival rates for extramedullary plasmacytoma are between 30% and 82%. Most deaths occur from development of disseminated multiple myeloma. The rate of conversion of extramedullary plasmacytoma to multiple myeloma has been shown to be from 10 to 36% in various series. Progression to multiple myeloma usually occurs within 2 years of diagnosis, but has occurred up to 15 years later indicating the need for long-term follow-up. This biphasic nature of progression to multiple myeloma suggests that patients with early relapse probably had undetected

myeloma present at the time of diagnosis. The more slowly progressing group may represent patients who developed new primaries in dysplastic marrow. This interpretation has important implications for more intensive screening to be carried out to exclude a diagnosis of multiple myeloma.¹⁰

Table 1: Diagnostic criteria for solitary extramedullary plasmacytoma (Guidelines working group of the UK myeloma forum)¹⁰

<ol style="list-style-type: none"> 1. Single extramedullary mass of clonal plasma cells. 2. Histologically normal marrow aspirate and trephine. 3. Normal results on skeletal survey, including radiology of long bones. 4. No anemia, hypercalcemia or renal impairment due to plasma cell dyscrasia. 5. Absent or low serum or urinary level of monoclonal immunoglobulin.

REFERENCES

1. Bataskis JG. Pathology consultation. Plasma cell tumors of the Head and Neck. *Ann Otol Rhinol Laryngol* 1983;92:311-13.
2. Fu YS, Perzin KH. Nonepithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx. A clinicopathologic study. IX. Plasmacytomas. *Cancer* 1978;42:2399-406.
3. Windufuhr JP, Ott G. Extramedullary plasmacytoma manifesting as a palpable mass in the nasal cavity. *Ear, Nose and Throat journal* 2002;81:100-14.
4. Wiltshaw E. The natural history of extramedullary plasmacytoma and its relation to solitary myeloma of bone and myelomatosis. *Medicine (Baltimore)* 1976;55:217-38.
5. Kapadiya SB, Desai U, Cheng VS. Extramedullary plasmacytoma of the head and neck: A clinicopathological study of 20 cases. *Medicine (Baltimore)* 1982;61:317-29.
6. Webb HE, Harrison EG, Masson JK, ReMine WH. Solitary extramedullary myeloma (plasmacytoma) of the upper part of the respiratory tract and oropharynx. *Cancer* 1962;15:1142-55.
7. Batsakis JG, Medeiros JL, Luna MA, El-Naggar AK. Plasma cell dyscrasias and the head and neck. *Ann Diagn Pathol* 2002;6:129-40.
8. Komisar A, Schetman F, DaSilva M, et al. The histopathologic diagnosis of head and neck tumors by special stains. *Ear, Nose and Throat J* 1989;68:702-12.
9. Soutar R, Lucraft H, Jackson G, et al. Guidelines Working Group of the UK Myeloma Forum; British Committee for Standards in Haematology; British Society for Haematology. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. *Br J Haematol* 2004;124(6):717-26.
10. Michalaki VJ, Hall J, Henk JM, Nutting CM, and Harrington KJ. Definitive radiotherapy for extramedullary plasmacytomas of the head and neck. *British Journal of Radiology* 2003;76:738-41.