Case Report

Extramedullary plasmacytoma of the nasal cavity: A rare case report

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Abstract

Introduction: Extramedullary plasmacytoma is a rare plasma cell proliferative disorder arising outside the bone marrow. It shows 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. **Aim:** This study reports a case of an extramedullary plasmacytoma of the nasal cavity and provides a literature review on the topic. **Case Report:** A 32-year-old male presented to our hospital with a 6-month history of progressive left sided nasal obstruction and self-limiting epistaxis. Examination revealed a large pale-pinkish mass within the left nasal cavity. Surgical excision of the mass was done and sent for HPE, which showed features of plasmacytoma. this was confirmed further by immunohistochemistry which was positive for CD138 suggestive of plasmacytoma. Further assessment revealed the solitary nature of the condition, consistent with extramedullary plasmacytoma.
Keywords: Nasal cavity, extramedullary, plasmacytoma.

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INTRODUCTION

Plasma cells are mature B lymphocytes typically found in various tissues and organs and in inflammatory foci in general. Their main function is the production of immunoglobulins or antibodies. Plasma cell neoplasams refer to a group of lymphoproliferative disorders are characterized by neoplastic proliferation of a single clone of plasma cells, producing a monoclonal immunoglobulin[1-4]. Plasma cell neoplasms can present as a single lesion (solitary plasmacytoma) or as multiple lesions (multiple myeloma). Solitary plasmacytomas present as solitary plasmacytoma of bone (SPB), and extramedullary plasmacytoma (EMP)[5,6]. EMP is a rare localized plasma cell tumour originating in soft tissuses. Extramedullary plasmacytoma corresponds to less than 10% of all plasmacytic tumors[1,2], representing less than 1% of all head and neck tumors[4] and less than 0.5% of tumors of the aerodigestive tract[3]. Almost 80% of EMP occur in submucosal lymphoid tissues in head and neck, commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity but may also occur in the gastrointestinal tract, urinary bladder, gland, lymph nodes, and skin[7-9]. In nasal cavity, they represent approximately 4% of tumors. It accounts for approximately 1 – 3% of human malignancies, with median age of 55 to 60 years, and approximately two-thirds of patients are male[7,8]. This extramedullary tumor was first described in 1905 by Schridde[10]. The estimated global incidence of the disease is 1 case per 500,000 people[1,2] The etiology of this disease remains unknown, but chronic irritation from inhaled irritants and viral pathogenesis has been suggested[6]. Most patients present with symptoms such as epistaxis, nasal discharge (rhinorrhea) or nasal obstruction[7,8] It is important to distinguish EMP from other plasma cell tumors for the purposes of prognosis and treatment. The main differential diagnosis is multiple myeloma and Waldenstrom macroglobulinemia. The evaluation of a patient with a suspected EP should include a biopsy of the suspected lesion for tissue histological confirmation, a unilateral bone marrow aspirate and biopsy, and laboratory studies. The treatment of choice for EP is

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surgery and radiation therapy (RT) with dose of 40 to 50 Gy over a four-week period, the disease is highly radiosensitive. Small lesions may be cured with surgery alone and no adjuvant RT is indicated unless a residual local disease is suspicion[11]. Herein, we are presenting a case of extramedullary plasmacytoma of the nasal cavity in a 32 year old male patient.

CASE REPORT

A 32yr old male patient, who is mechanic by occupation, presented to our department with the complaints of left nasal obstruction which was gradually progressive in nature and bleeding from nose which was intermittent in nature since 6months. On examination, externally, bulge present over the dorsum of the nose at the junction of left nasal bone and the upper lateral cartilage area .On anterior rhinoscopy, single pale pinkish mass was seen in the left nasal cavity, occluding the whole of the nasal cavity extending up to the inferior turbinate. On probing, mass was soft in consistency, it was insensitive to touch and mass bleed on probing. Probe could be passed all around except laterally. Posterior rhinoscopy was normal. CT PNS showed features suggestive of left nasal cavity polypoidal mass (fig 1). Hence the patient considered to be a case of sinonasal polyposis and was posted for endoscopic excision of the mass. During surgery the mass was not consistent with the inflammatory polypoidal mass (fig 2) and also bleeding was more than anticipated .As a part of routine tissues was sent for Histopathological examination. HPE report showed features of plamacytoma (fig 3). So, Immunohistochemistry was done to confirm the diagnosis, it showed immunopositivity for the CD138 which is sugggestive of plasmacytoma. Postoperatively, on follow up for 6 months no recurrence of the disease was noted.

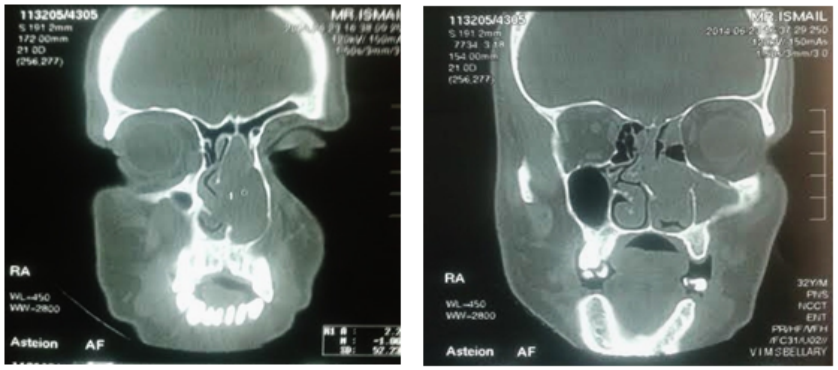

Figure 1

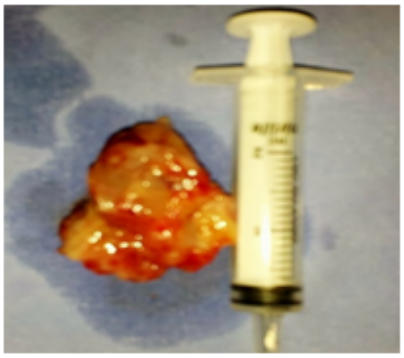

Figure 2

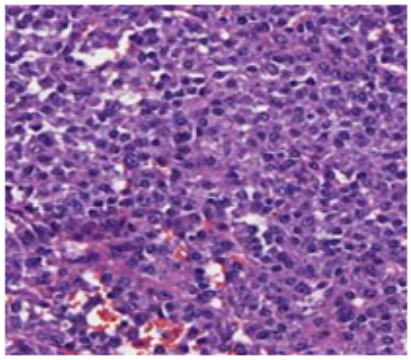

Figure 3

Legend

Figure 1: CT PNS showing radiopacity in the left nasal cavity and maxillary sinus

Figure 2: Mass excised from the left nasal cavity

Figure 3: HPE showing round cell tumour

DISCUSSION

Plasmacytoma is a rare solitary mass of neoplastic monoclonal plasma cells, first described by Schridde in 1905[9]. Extramedullary plasmacytoma originate from plasma cells with a single class of heavy and light chains in a monoclonal proliferation of B cells. The commonest immunoglobulin expressed by the tumor cells is IgG with kappa chain restriction. Eighty percent of EMPs arise in the soft tissues of the head and neck region. The nasal cavity, paranasal sinuses, and nasopharynx are the most

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common sites[12,13]. Extramedullary plasmacytoma represent 3% of plasma cell neoplasms. Tissue biopsy, serum electrophoresis and radiological skeletal survey with bone marrow study are necessary for diagnosis. The treatment of EP is surgical resection and radiotherapy[14]. Chemotherapy may be considered for patients with refractory or relapsed disease[11]. Follow-up radiological and serum electrophoresis is required after treatment to detect recurrences and progression to myeloma. Approximately 10% of EP has multiple sites of involvement[5]. The rate of progression to multiple myeloma is lower than in solitary bone plasmacytoma (SBP), ranging from 11% to 30% and is associated with a poorer prognosis[7]. In pathology, plasma cells are present with a morphologic spectrum ranging from mature forms with abundant cytoplasm and perinuclear halo to highly atypical cells with large nuclei, hyperchromatic clumped chromatin, and prominent nucleoli with scant cytoplasm. Amyloid deposition may be seen in 15%–38% of extramedullary plasmacytoma. Immunohistochemistry is used to establish clonality[6]. Wiltshaw classified soft-tissue plasmacytoma into 3 clinical stages, as follows[14]:

Stage I – Limited to an extramedullary site
Stage II – Involvement of regional lymph nodes
Stage III – Multiple metastasis

The 10-year survival rate is 70%. Plasmacytoma should be differentiated from other lesions having plasma cells such as plasma cell granuloma, chronic granulomatous inflammation and rhinoscleroma. Plasma cell granuloma shows an admixture of inflammatory cells including plasma cells, histiocytes, eosinophils and fibroblasts. A chronic granulomatous inflammation is characterized by the presence of epithelioid histiocyte, granulomas and giant cells. Lesions with monoclonal plasma cells are considered neoplastic, whereas lesions with multiclonal plasma cells are inflammatory nature[9].

CONCLUSION

Extramedullary plasmacytoma can present as a polypoidal mass of the nasal cavity. Hence routine Histopathological examination of the excised polypoidal mass of the nasal cavity can detect these rare occurrences

REFERENCES

1. Fernandes AM, Podovani JA, Maniglia JP. Plasmocitoma extramedular de nasofaringe: relato de um caso e revisão da literatura. Rev Bras Otorrinolaringol. 1998; 64:296-8.
2. Sousa RMA, Costa EG, Takahashi GM, Butugan O, Miniti A. Mieloma múltiplo e manifestações raras dentro da otorrinolaringologia. Rev Bras Otorrinolaringol. 1993; 59:2848.
3. Granato L, Petitto JW, Prospero JD. Plasmocitoma extramedular do aparelho respiratório Apresentação do caso. Rev Bras Otorrinolaringol. 1977; 43:214-23.
4. Kuppersmith RB. Extramedullary plasmocytoma of the head and neck. [Cited 1996].
5. Dores GM, Landgren O, McGlynn KA, Curtis RE, Linet MS, Devesa SS. Plasmacytoma of bone, extramedullary plasmacytoma, and multiple myeloma: incidence and survival in the United States, 1992-2004. Br J Haematol. 2009; 144(1):86–94. doi: 10.1111/j.1365-2141.2008.07421.
6. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group. Br J Haematol. 2003;121(5):749–57
7. Bachar G, Goldstein D, Brown D, Tsang R, Lockwood G, Perez-Ordonez B, et al. Solitary extramedullary plasmacytoma of the head and neck--long-term outcome analysis of 68 cases. Head Neck. 2008; 30(8):1012–9. doi: 10.1002/hed.20821.
8. Kitamura A, Yamashita Y, Hasegawa Y, Kojima H, Nagasawa T, Mori N. Primary lymphoma arising in the nasal cavity among Japanese. Histopathology. 2005; 47(5):523–32. doi: 10.1111/j.1365-2559.2005.02265
9. Soutar R, Lucraft H, Jackson G, Reece A, Bird J, Low E, et al. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. Clin Oncol (R Coll Radiol). 2004; 16(6):405–13.
10. Shridde H. Weitere Untersuchungen uber die Kornelunger der Plasmazellen. Centralbl Allg Pathol Anatol. 1905;16:433-5
11. Creach KM, Foote RL, Neben-Wittich MA, Kyle RA. Radiotherapy for extramedullary plasmacytoma of the head and neck. Int J Radiat Oncol Biol Phys. 2009; 73(3):789–94. doi: 10.1016/j.ijrobp.2008.04.077
12. Manganaris A, Conn B, Connor S, Simo R. Uncommon presentation of nasopharyngeal extramedullary plasmacytoma: a case report and literature review. B-ENT. 2010; 6(2):143–6.
13. Micozkadioglu SD, Erkan AN, Kocer NE. Extramedullary plasmacytoma of the septum. B-ENT.2009; 5(3):169–71.
14. Wiltshaw E. The natural history of extramedullary plasmacytoma and its relation to solitary myeloma of bone and myelomatosis. Medicine (Baltimore). 1976; 55(3):217–38.

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