



Isolated Extramedullary Nasopharynx Plasmacytoma: Neglected Otolgia

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Authors' contributions

This work was carried out in collaboration between both authors. Both authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Primary isolated plasmacytoma is an uncommon malignancy. However, occurrence of isolated plasmacytoma involving head and neck region is approximately 80% of cases. In head and neck isolated plasmacytoma, symptoms reported includes nasal obstruction, local pain, epistaxis, ear fullness, hoarseness. Occasionally, isolated plasmacytoma may present in the nasal cavity. We report a case of isolated nasopharynx plasmacytoma presenting as otalgia and literature review on presenting symptoms and management of isolated nasal cavity plasmacytoma.

Keywords: Nasopharynx; extramedullary plasmacytoma; otalgia.

1. INTRODUCTION

Plasmacytoma is a rare tumour that can present as either a widespread disease or localized disease. Localized plasmacytoma can be solitary medullary or solitary extramedullary depending on its origin from bone or soft tissue, respectively. Eventhough it is rare, solitary

extramedullary plasmacytoma occurrence is approximately 80% in the head and neck region which constitutes less than 1% of head and neck tumour [1].

Here, we present a case of neglected ear pain with incidental finding of a nasopharyngeal mass which was diagnosed as isolated extramedullary

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plasmacytoma and we review the clinical features, imaging and histopathologic findings, and treatment options of this rare disease.

2. CASE PRESENTATIONS

A 48 year old man presented to us with 3 months history of persistent right ear pain. He had sought prior treatment in general practitioner and was treated as otitis externa with topical ear drops, however, his symptoms persisted. There was no other significant ear, nasal and throat symptoms. Otoscopic examinations showed mildly retracted tympanic membrane with no other remarkable findings. Rigid endoscopy revealed small irregular non fungating mass at right fossa of Rosenmuller (FOR) (Fig. 1). No cervical lymph node palpable. Pure Tone audiometry result showed Bilateral normal hearing and bilateral tympanogram type A.

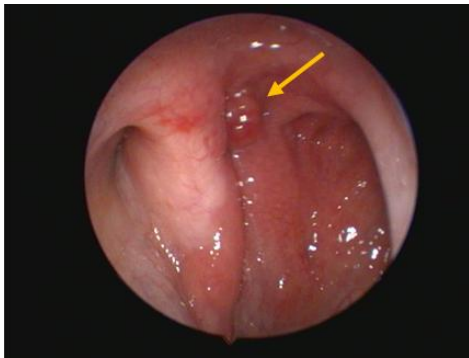


Fig. 1. Endoscopic view of right nasopharynx. Arrow showing small right upper FOR mass

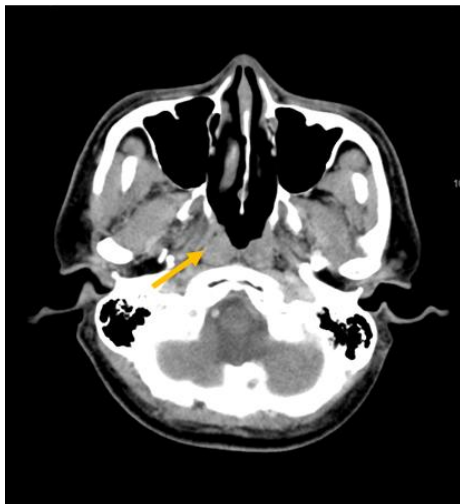


Fig. 2. Axial view: Arrow showing slight asymmetric thickening of the right roof of the nasopharynx with no local invasion

Biopsy of the nasopharynx was performed endoscopically under local anaesthesia in clinic setting. The mass was friable however, only minimal bleeding at biopsy site which stopped after ice compression. Results of the right FOR showed lymphoid stroma with prominent germinal centres with a focal area of plump cells with eosinophilic cytoplasm. Some of these cells show plasmacytoid appearance, exhibiting round, eccentrically placed nuclei with clock face appearance. The plasma cells are positive for CD138 and negative for MNF 116 pancytokeratin immunohistochemistry. Skeletal survey revealed no primary lesion. Computed tomography (CT) of paranasal sinus and neck revealed asymmetric thickening of the right roof of the nasopharynx with no local invasion (Fig. 2) with non significant left upper cervical lymphadenopathy.

A diagnosis of isolated nasopharynx plasmacytoma was established and further evaluation of multiple myeloma was not significant. Serum and urine protein electrophoresis was normal and showed no sign of Bence Jones protein. Skeletal survey done showed negative for lytic and blastic lesions and bilateral iliac bone marrow biopsies results were normal.

Due to the rarity of disease, multidisciplinary discussion decided for radiotherapy 45-50 gy /25 fractions for this patient.

3. DISCUSSION

Solitary extramedullary plasmacytomas accounts 3% of plasma cell malignancies. Approximately 75% of solitary plasmacytoma involve the nasal cavity, paranasal sinuses, and the nasopharynx where others were found in the tonsils, oropharynx, and larynx [2].

Extramedullary plasmacytoma of the head and neck region is a rare tumour which usually occurs in the submucosa of the upper aerodigestive tract. Susnerwala et al. [3] reported 25 cases of extramedullary plasmacytoma of the head and neck, where the commonest sites of presentations are the nasal cavity and sinuses, followed by nasopharynx, tonsils, larynx.

It appears that most patients with solitary extramedullary plasmacytoma in the nasal cavity presented with nasal obstruction and epistaxis [4-13]. Few cases present with epistaxis only [14-17] or with associated symptoms of facial pain and swelling [6,18,12,19]. Other unusual

presentations reported were sudden expectorated polypoidal mass. Lomeo et al. [6,14] and visual loss [20]. There were also cases reported as confined nasopharynx solitary plasmacytoma with presentations of hemoptysis reported by Mangaris et al. [7] and symptoms of nasal obstructions [14,21].

As per our knowledge, there was no report on otalgia as presenting symptoms in solitary nasopharynx plasmacytoma. A possible reason for otalgia is the mass effect and proximity of the tumour to the eustachian tube leading to middle ear effusion and ear pain. Clinically, this patient had mildly retracted tympanic membrane, which may be due to middle ear effusion, however, hearing test showed no significant results.

It is of paramount important to rule of nasopharynx pathology especially if patient presented with unresolved/chronic otalgia with normal ear examinations. Even though without a significant nasal symptoms, assessment and early referral to otorhinolaryngologist is crucial to avoid mismanagement and late diagnosis.

To the best of author knowledge, there are no randomized studies regarding the best management of solitary extramedullary plasmacytoma in the nasal cavity, due to the rarity of this case. Reported solitary extramedullary diseases originating from the nasal cavity, nasopharynx or sinonasal region, were mainly treated with surgical resection alone [4,5,8,22,23,13,19,20] and few cases were managed with combined surgical and radiotherapy [6,7,14,24,9,10,16,25,17]. In most cases, the surgical resections were done via transnasal endoscopic approach [4,26,27,9,25,12]. There were 3 cases who undergone radiotherapy alone [15,21,28] and 2 cases reported failed radiotherapy and were proceeded with surgical resection later [2,27,12]. Likewise, cases of solitary extramedullary plasmacytoma originating from nasopharynx [7,14,21], has no specific treatment, 1 case each was managed by surgical resection only [14], radiotherapy only [21] and combined surgical and radiotherapy [7]. Our case was managed with radiotherapy only as per multidisciplinary discussion.

Nevertheless, generally the treatment of choice for solitary extramedullary plasmacytoma is radiotherapy [29], although its efficacy has been tested only in small retrospective series [30]. Vento et al study of 25 patients with sinonasal or nasopharyngeal extramedullary plasmacytoma

supports radiotherapy as a treatment of choice, however, for small tumours surgical intervention alone or combined with radiotherapy may be considered [31].

The prognosis of extramedullary plasmacytoma is generally good. Nonetheless, some patients might recover fully or suffers from multiple myeloma with a conversion rate between 11% to 33% over 10 years [32]. Maheshawi et al. reported 5 patients were treated with surgical resection followed by radiotherapy in which 3 (60%) patients are alive and disease-free, 1 (20%) patient is alive with multiple myeloma and 1 (20%) patient died of multiple myeloma in a median follow up period of 24 (range 18-40 months) with no cases of loco-regional recurrence [33]. Hence, long term close follow up is mandatory to detect locoregional recurrent or conversion to multiple myeloma.

In the present case, the patient is still undergoing oncological treatment, nevertheless, surgery is still the next available treatment to be considered if the tumour is radioresistant or locoregional recurrent.

4. CONCLUSION

Solitary extramedullary plasmacytoma in head and neck region treatment remain controversial. However, early diagnosis is warranted for early management for a better outcome. It is crucial not to neglect ear symptoms to rule out nasopharynx pathology.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Straetmans Jos, Robert Stokroos. Extramedullary plasmacytomas in the head and neck region. *European Archives of*

- Oto-Rhino-Laryngology. 2008;265(11): 1417-1423.
2. Windfuhr JP, Ott G. Extramedullary plasmacytoma manifesting as a palpable mass in the nasal cavity. *Ear Nose Throat J.* 2002;81(2):110-14.
 3. Susnerwala SS, et al. Extramedullary plasmacytoma of the head and neck region: Clinicopathological correlation in 25 cases. *British Journal of Cancer.* 1997; 75(6):921-927.
 4. Shahrizal, Tengku A, et al. Isolated extramedullary plasmacytoma of the middle turbinate. *Ear, Nose & Throat Journal.* 2009;88(2):786-789.
 5. Sodhi, Kushaljit Singh, et al. Solitary extramedullary plasmacytoma of the nasal tract: An unusual cause of epistaxis. *Ear, Nose & Throat Journal.* 2013;92(6):E51-E54.
 6. Lomeo, Paul E, John E. McDonald, Judith Finneman. Extramedullary plasmacytoma of the nasal sinus cavities. *American Journal of Otolaryngology.* 2007;28(1):50-51.
 7. Chang, Yen-Liang, Po-Yueh Chen, Shih-Han Hung. Extramedullary plasmacytoma of the nasopharynx: A case report and review of the literature. *Oncology Letters.* 2014;7(2):458-460.
 8. Corvo, Marco Antonio dos Anjos, et al. Extramedullary nasal plasmacytoma: Literature review and a rare case report. *International Archives of Otorhinolaryngology.* 2013;17(2):213-217.
 9. Belić, Branislav, et al. Nasal septum extramedullary plasmacytoma. *Vojnosanitetski Pregled.* 2013;70(2):221-224.
 10. Hazarika, Produl, et al. Solitary extramedullary plasmacytoma of the sinonasal region. *Indian Journal of Otolaryngology and Head & Neck Surgery.* 2011;63(1):33-35.
 11. Tandon S, Meher R, Chauhan A. extramedullary plasmacytoma of nasal cavity. *Clinical Rhinology.* 2016;9.1:50-52.
 12. Ishtiaq, Rizwan, et al. Nasal plasmacytoma: A rare cause of persistent epistaxis. *Postgraduate Medicine.* 2018; 130(6):507-510.
 13. Gökmen, Muhammet Fatih, Hazan Başak, Süha Beton. Rare neoplasm of inferior turbinate: Extramedullary plasmacytoma. *Allergy.* 2018;1:50-3.
 14. Manganaris A, et al. Uncommon presentation of nasopharyngeal extramedullary plasmacytoma: A case report and literature review. *B-ENT.* 2010; 6(2):143-6.
 15. Ashraf, Mohamad Javad, et al. Extramedullary plasmacytoma of the nasal cavity report of three cases with review of the literature. *Iranian Red Crescent Medical Journal.* 2013;15(4):363.
 16. Adoga, Adeyi A, Olugbenga A. Silas, John P. Yaro. Extramedullary plasmacytoma of the nasal cavity and ethmoidal sinus in human immunodeficiency virus-positive patient. *Journal of Cancer Research and Therapeutics.* 2020;16(1):157.
 17. Di Stadio, Arianna, et al. Septal Nasal extramedullary plasmacytoma: A rare tumor in an Unusual Area. *Ear, Nose & Throat Journal.* 2020;0145561320911735.
 18. Anil, Sukumaran. Solitary plasmacytoma of the maxilla--a case report and review of the literature. *General Dentistry.* 2007; 55(1):39-43.
 19. Jaswal, Abhishek, et al. Extramedullary plasmacytoma of maxillary sinus in 10-year-old girl: A rare presentation. *International Journal of Pediatric Otorhinolaryngology Extra.* 2008;3(3):143-146.
 20. Özdemir, Süleyman, et al. A case of extramedullary plasmacytoma in the sphenoid sinus with unilateral loss of vision. *Journal of Cranio-Maxillofacial Surgery.* 2013;41.2:140-143.
 21. Krishnasamy, Gopiraj, Ganapathy Dhanasekar. Extramedullary plasmacytoma of postnasal space. *Kulak burun bogaz ihtisas dergisi: KBB= Journal of Ear, Nose, and Throat.* 2013;23(3):183-186.
 22. Grover, Nancy, et al. Extra medullary plasmacytoma of the nasal cavity: Treatment perspective in a developing nation. *Ear, Nose & Throat Journal.* 2006;85(7):434-436.
 23. Micozkadioglu SD, Erkan AN, Kocer NE. Extramedullary plasmacytoma of the septum. *Acta oto-rhino-laryngologica Belgica.* 2009;5(3):169.
 24. Shreif, Jamal A, et al. Extramedullary plasmacytoma of the nasal cavity. *Otolaryngology-Head and Neck Surgery.* 2001;124(1):119-120.
 25. Selvarajoo J, Ying JKP, Tang IP. Nasal extramedullary plasmacytoma - A rare case. *Ann Clin Lab Res.* 2019;7(1):285.

26. Baek, Byoung J, et al. Extramedullary plasmacytoma arising from the nasal septum. *Ear, Nose & Throat Journal*. 2005; 84(11):720-722.
27. Hidaka, Hiroshi, et al. A case of extramedullary plasmacytoma arising from the nasal septum. *The Journal of Laryngology & Otology*. 2000;114(1):53-55.
28. Sasitharan, Paskaramoorthy, Nor Idayu Mohd Yusof, Kugan Varatha Raju. Extramedullary plasmacytoma of the nasal cavity: A case report. *Romanian Journal of Rhinology*. 2018;8(32):233-234.
29. Chao MW, et al. Radiotherapy in the management of solitary extramedullary plasmacytoma. *Internal Medicine Journal*. 2005;35(4):211-215.
30. Grammatico S, Scalzulli E, Petrucci MT. Solitary plasmacytoma. *mediterr. J Hematol Infect Dis*. 2017;9(1):e2017052.
31. Vento, Seija Inkeri, et al. Clinical findings in 25 patients with sinonasal or nasopharyngeal extramedullary plasmacytoma in a four-decade single-centre series. *Acta Oto-Laryngologica*. 2017;137(9):975-980.
32. Hu, Kenneth, Joachim Yahalom. Radiotherapy in the management of plasma cell tumors. *Oncology (Williston Park, NY)*. 2000;14(1):101.
33. Maheshwari A, Panigrahi R, Mahajan S. Solitary extramedullary plasmacytoma of nasal cavity: an emerging differential diagnosis of nasal masses. *International Journal of Scientific Study*. 2016;3(11): 178-181.

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