CASE REPORT
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Plasma Cell Neoplasms of Paranasal Sinuses: Two Case Reports

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ABSTRACT

Sinonasal system malignancies are rare, constituting only 1% of all malignancies and less than 5% of head and neck malignancies. The common form of plasma cell dyscrasias is called multiple myeloma (MM), and the localized forms are called solitary bone plasmacytoma and extramedullary plasmacytoma (EMP). EMP constitutes 3-4% of malignant plasma cell dyscrasias and 1% of all head and neck tumors. 90% of EMP cases occur in the head and neck region, because the nasal and paranasal sinus regions are rich in plasma cells at around 75%. This study aims to present two different cases of plasma cell tumors presenting in the paranasal region. Based on the imaging studies, the first case is seen to have been diagnosed as MM due to bone marrow involvement and to have received systemic chemotherapy treatment, whereas the second case is seen to have undergone endoscopic total excision of the lesion due to the absence of systemic involvement. In the first case, MM was considered to have gone into remission in the sixth month after chemotherapy; however, the patient died one year after diagnosis. In the second case, no recurrence was detected during the fifth year postoperative follow-up. As a result, patients who show sinonasal symptoms should be examined in detail.

Keywords: Extramedullary plasmacytoma, paranasal sinuses surgery, multiple myeloma

INTRODUCTION

Sinonasal system malignancies are rare, constituting only 1% of all malignancies and less than 5% of head and neck malignancies (1). The common form of plasma cell dyscrasias is known as multiple myeloma (MM), with the localized forms being solitary bone plasmacytoma and extramedullary plasmacytoma (EMP). EMP constitutes 3-4% of malignant plasma cell dyscrasias and 1% of all head and neck tumors (2, 3). The median age at diagnosis is 55-60 years, with approximately two-thirds of patients being male (4). 90% of EMP cases occur in the head and neck region (5), due to the nasal and paranasal sinus regions being rich in plasma cells at around 75% (6, 7). The study aims to present two different cases of plasma cell tumors presenting in the paranasal region that required different treatment and had different prognoses.

CASE PRESENTATIONS

Case 1

A 72-year-old female patient presented to the clinic with a complaint of a headache that had been occurring behind her left eye for the last 2 months. The patient had a history of diabetes and hypertension. On physical examination, limitation was seen to be present in the left eye movements, as well as diplopia. Upon endoscopic examination, a mass lesion was observed in the left nasal passage filling the middle meatus. The patient's complete blood count, biochemical parameters, kidney function tests and coagulation test values were normal. Paranasal computed tomography (CT) revealed a mass filling the left nasal passage, eroding the anterior wall of the sphenoid sinus and the anterior skull base (Figure 1). In magnetic resonance imaging (MRI), a solid mass lesion measuring

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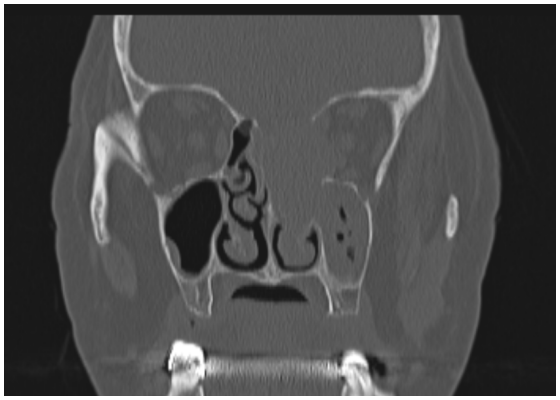

Figure 1: In the preoperative paranasal CT scan along the coronal plane without contrast, soft tissue is seen in the superior part of the nasal cavity filling the anterior part of the sphenoid sinus and extending superiorly anteriorly to the base of the cranial fossa, largely filling the nasal cavity and extending into the intra-orbital space on the left.tympanic segment of the facial canal

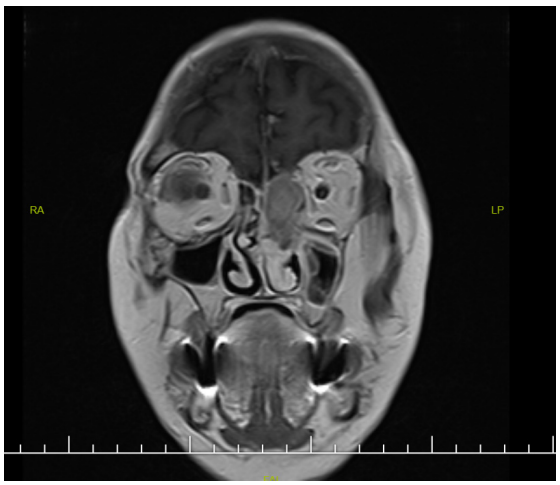

Figure 2: This orbital T1-weighted MR imaging scan of the coronal view shows a solid mass lesion measuring approximately 50x33x28 mm, with minimal compression in the medial rectus muscle, narrowing the optic canal; it is indistinguishable from the medial and inferior parts of the bilateral cavernous sinus

50x33x28 mm was observed, compressing the optic nerve and appearing hyperintense in the contrast-enhanced series (Figure 2). A written informed consent form was obtained from the patient.

Multiple biopsies were taken under general anesthesia from the polypoid mass extending to the anterior and posterior ethmoid cells. The pathology result was reported as EMP. In the positron emission tomography, fludeoxyglucose (FDG) uptake was detected in the medullary region of the left iliac wing, in addition to the mass in the paranasal region. The patient was referred to the hematology department for diagnosis. A bone marrow biopsy was taken and reported as plasma cell myeloma, after which chemotherapy treatment was planned. The orbital MRI and CT performed at six months after treatment showed the lesion to have regressed (Figures 3 and 4).

The patient presented with poor general condition 12 months after the end of her treatment. A brain MRI showed widespread parenchymal and leptomeningeal contrast enhancement and edema in the bilateral cerebellar and cerebral hemispheres. This was thought to possibly be a result of leptomeningeal involvement due to MM. Chemotherapy treatment could not be given to the patient due to her poor overall health. Despite all interventions, the patient died due to cardiopulmonary arrest while following up in the intensive care unit.

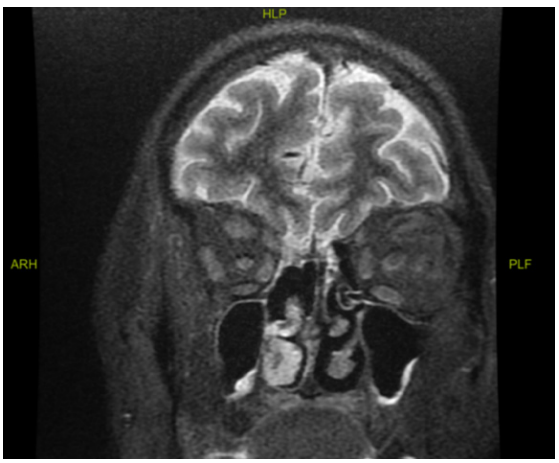

Figure 3: Regression of the existing lesion from the previous imaging studies in the orbital T1-weighted MR imaging scan from the coronal view, six months post-chemotherapy

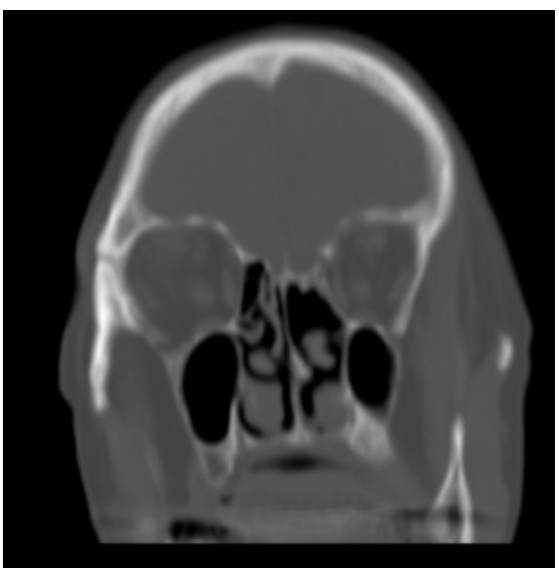

Figure 4: A paranasal CT of the coronal view in the sixth month after chemotherapy showing regression of the existing lesion in the previous imaging studies

Case 2

A 35-year-old male patient was admitted to the clinic with complaints of nasal congestion and nosebleeds for the last 3 months. In the endoscopic examination, a polypoid mass was observed starting from the nasal vestibule and extending posteriorly. He had no history of any chronic diseases. The patient's complete blood count, biochemical parameters, kidney function tests, and coagulation values were normal. In the paranasal CT, a mass was observed in the nasal cavity that had destroyed the anterior wall of the sphenoid sinus, ethmoid cells, skull base, and left orbital medial wall. In the MRI, a 41X33 mm mass with a mild to moderately contrast-enhancing was observed in the left nasal cavity expanding toward the left maxillary sinus in the medial nasal cavity, inferomedial of the orbital and anterior skull base (Figures 5 and 6). Written informed consent form was obtained from the patient.

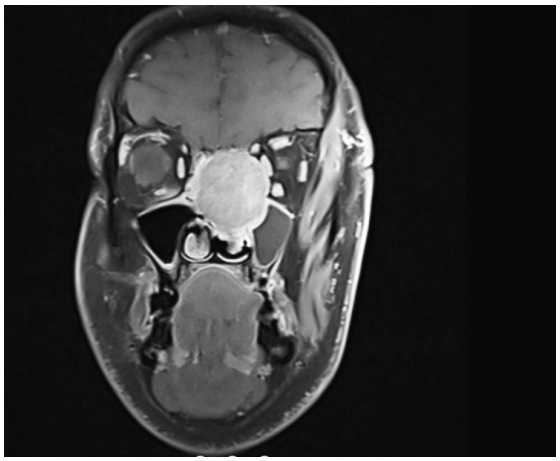

Figure 5: This paranasal T1-weighted MR imaging scan from the coronal view shows a contrasting lesion 41X33 mm in size in the left nasal cavity expanding towards the left maxillary sinus , inferomedial of the orbital and anterior skull base

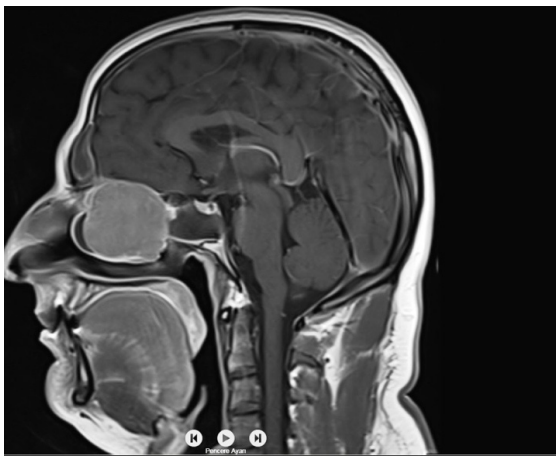

Figure 6: A paranasal T1-weighted MR imaging scan of the sagittal view showing a contrasting lesion 41X33 mm in size in the left nasal cavity expanding toward the anterior skull base

The patient underwent endoscopic excision of the mass under general anesthesia. The frozen section obtained during the operation was reported as a plasma cell tumor, and the lesion was completely excised endoscopically with blunt and sharp dissection from the surrounding structures. A defect was observed at the skull base, but no finding was present

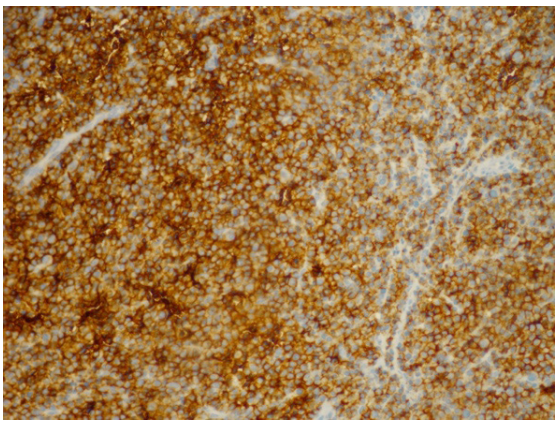

Figure 7: All cells were stained with CD38 (Immunohistochemistry; x20 obj)

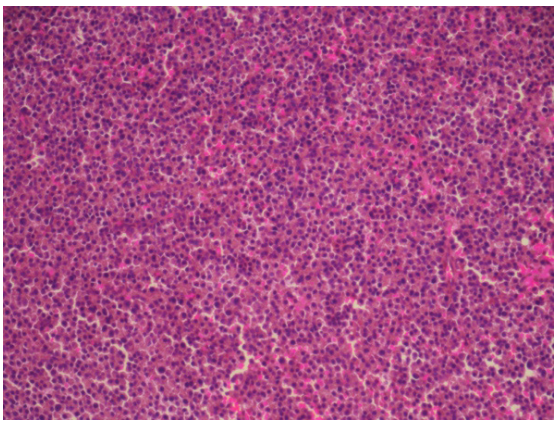

Figure 8: Neoplastic plasma cells (H&E; x20 obj)

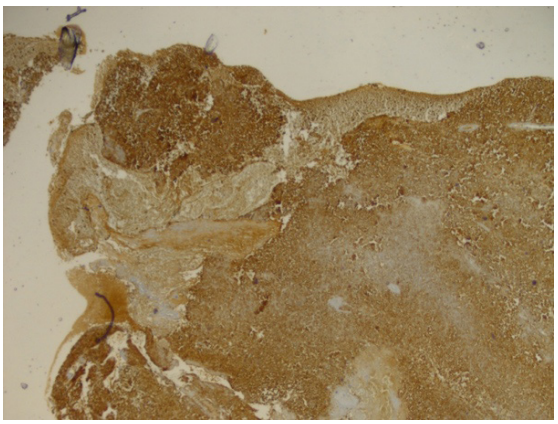

Figure 9: Positive staining with lambda (Immunohistochemistry; x4 obj)

suggestive of dural invasion, and the operation was terminated without repair due to the diameter of the defect being less than 1 cm. The pathology result was reported as a completely resected plasma cell neoplasia showing lambda clonality (Figures 7-9).

In the lumbar spine MRI, no lesion was detected, and the bone marrow biopsy results showed hypercellular bone marrow, thus ruling out any systemic involvement. No recurrence was detected in the endoscopic examination, blood tests, or MRI in five years of follow-ups.

DISCUSSION

EMP can remain localized or develop into systemic MM after a latent period. EMP has a better prognosis than MM and can be treated surgically (8). EMP progresses as a locally invasive tumor, but 5-20% of cases may present with cervical lymph node metastasis at diagnosis (9). The symptoms due to tumoral mass include nasal congestion, facial swelling, pain, and nose bleeding (10). The diagnosis of EMP is made by lesion biopsy and pathological examination of clonal plasma cell populations, with a lack of clonal plasma cells in the bone marrow and involvement in the spine and pelvis (11). The overall 10-year survival rate in EMP cases has been reported to be 70% (12), with 11-30% of cases able to progress to MM within 10 years (13).

Various surgical methods are found in the treatment of EMP; however, radiotherapy is considered as a preferred treatment method due to the tumor's radiosensitivity. İn recent studies, the combination of radiation therapy (RT) and surgical excision has shown better results for the long-term follow-up periods. In a case study examining seven cases, the authors performed RT in the first postoperative month. At the time of analyzing these data 5 patients were alive and two have died of their disease. A single patient, presenting local relapse at 6 months, one patient progressed to multiple myeloma (14). Sasaki et al. showed patients who receive any combination of RT and surgery to have significantly improved survival rates compared to patients receiving RT or surgery alone (15). The present study's first case had been referred to systemic treatment because she had been diagnosed with MM. In The second case, only surgical treatment was applied after the presence of a systemic disease had been excluded. Due to no tumor being found within the surgical margins, radiotherapy treatment was not given afterwards.

CONCLUSION

Because the symptoms in the early stages of paranasal sinus tumors are subtle and the findings may mimic chronic sinusitis, proper diagnosis may be delayed. All patients with symptoms of sinonasal neoplasia should undergo detailed endoscopic examination.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer Review: Externally peer-reviewed.

Author Contributions: Conception/Design of Study - H.A., D.M.M.; Data Acquisition- M.M.; Data Analysis/Interpretation- G.A., D.Ö.; Drafting Manuscript- H.A., D.M.M., G.A.; Critical Revision of Manuscript- M.M., D.Ö.; Final Approval and Accountability- H.A., D.Ö., M.M., G.A., D.M.M.

Conflict of Interest: The authors have no conflict of interest to declare.

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REFERENCES

1. Harvey RJ, Dalgorf DM. Chapter 10: Sinonasal malignancies. Am J Rhinol Allergy 2013;27(Suppl 1):S35-8.
2. Nowak-Sadzikowska J, Weiss M. Extramedullary plasmacytoma of the larynx. Analysis of 5 cases. Eur J Cancer 1998;34(9):1468.
3. Nofsinger YC, Mirza N, Rowan PT, Lanza D, Weinstein G. Head and neck manifestations of plasma cell neoplasms. Laryngoscope 1997;107(6):741-6.
4. Frassica DA, Frassica FJ, Schray MF, Sim FH, Kyle RA. Solitary plasmacytoma of bone: Mayo Clinic experience. Int J Radiat Oncol Biol Phys 1989;16:43-8.
5. Çömez G, Pehlivan Y, Sevinç A, Kervancıoğlu S, Sarıca MA, Tutar E, et al. Extramedullary plasmacytoma with maxillary sinus involvement. UHOD 2008;2(18):112-5.
6. Susnerwala SS, Shanks JH, Banerjee SS, Scarffe JH, Farrington WT, Slevin NJ. Extramedullary plasmacytoma of the head and neck region: Clinicopathological correlation in 25 cases. Br J Cancer 1997;75(6):921-7.
7. Holland J, Trenker DA, Wasserman TH, Fineberg B. Plasmacytoma: treatment results and conversion to myeloma. Cancer 1992;69(6):1513-7.
8. Ayan N, Yılmaz B, Çınar F, Çevikbaş U, Gürsel AO. Extramedullary plasmacytoma. KBB ve Baş Boyun Cerrahisi Dergisi 1995;3:73-5.
9. 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.
10. Patel TD, Vázquez A, Choudhary MM, Kam D, Baredes S, Eloy JA. Sinonasal extramedullary plasmacytoma: a population-based incidence and survival analysis. Int Forum Allergy Rhinol 2015;5(9):862-9.
11. Ramadan O. Outcomes of patients with nasopharyngeal plasmacytoma: A systematic review. Otolaryngol Open J 2017;3(2):30-40.
12. 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;144:86-94.
13. Straetmans J, Stokroos R. Extramedullary plasmacytomas in the head and neck region. Eur Arch Otorhinolaryngol 2008;265(11):1417-23.
14. Vlad D, Trombitas V, Albu S. Extramedullary plasmacytoma of the paranasal sinuses: Combining surgery with external radiotherapy. Indian J Otolaryngol Head Neck Surg 2016;68(1):34-8.

15. Sasaki R, Yasuda K, Abe E, Uchida N, Kawashima M, Uno T, et al. Multi-ınstitutional analysis of solitary extramed- ullary plasmacytoma of the head and neck treated with cura- tive radiotherapy. Int J Radiat Oncol Biol Phys 2012;82(2):626-34.