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Case Report

Hidden intruder: Plasmacytoma causing uncontrolled epistaxis in an elderly patient ☆

Omar Younis, MD[a], *Anas Odeh, MD*[a], *Motaz Saifi, MD*[a], *Yazan Dumaidi, MD*[a], *Mazen Kazlak, MD*[a,b,*]

[a] *Department of Medicine, Faculty of Medicine and Health Sciences, An-Najah National University, Nablus, Palestine*
[b] *Department of Otorhinolaryngology, An-Najah National University Hospital, Nablus, Palestine*

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ABSTRACT

Extramedullary plasmacytoma (EMP) is an uncommon tumor marked by the monoclonal growth of plasma cells without the characteristics of multiple myeloma. EMP represents 3% of all plasma cell tumors. An 89-year-old male patient with hypertension was admitted to our tertiary care hospital with uncontrolled unilateral epistaxis. After a year and a half of recurring epistaxis, the patient's bleeding became more frequent and could no longer be controlled with nasal packing. Angiofibroma was suspected as the initial differential diagnosis, and angiofibroma embolization was performed. The patient experienced difficulty swallowing and slurred speech and was diagnosed with an ischemic stroke. However, antiplatelet and tranexamic acid medications were contraindicated due to bleeding risks. The patient underwent functional endoscopic sinus surgery, and unexpectedly, histology results revealed a plasmacytoma. After surgery, the patient remained stable and was discharged for further management.

Introduction

Plasmacytoma is an uncommon plasma cell dyscrasia that presents a unique clinical challenge because it shows up as one or more masses in the axial skeleton or soft tissues without systemic disease. According to the International Myeloma Working Group [1], it can be labeled into 2 primary sorts: solitary plasmacytomas and multiple solitary plasmacytomas (MSP). Solitary plasmacytoma similarly divides into solitary plasmacytoma of bone (SPB), originating from bone marrow plasma cells, and extramedullary plasmacytoma (EMP), springing up from mucosal surfaces.

Notably, SPB is more prevalent, constituting 2% to 5% of all plasma cell malignancies, while EMP accounts for 4% of all such malignancies [1]. It is crucial for otolaryngologists to be familiar with this condition, as 80% to 90% of extramedullary plasmacytoma cases are located in the head and neck, allowing for local treatment options [2]. These tumors primarily manifest in the respiratory tract, particularly in the sub-

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* Corresponding author.
E-mail address: mazen.kazlak@najah.edu (M. Kazlak).

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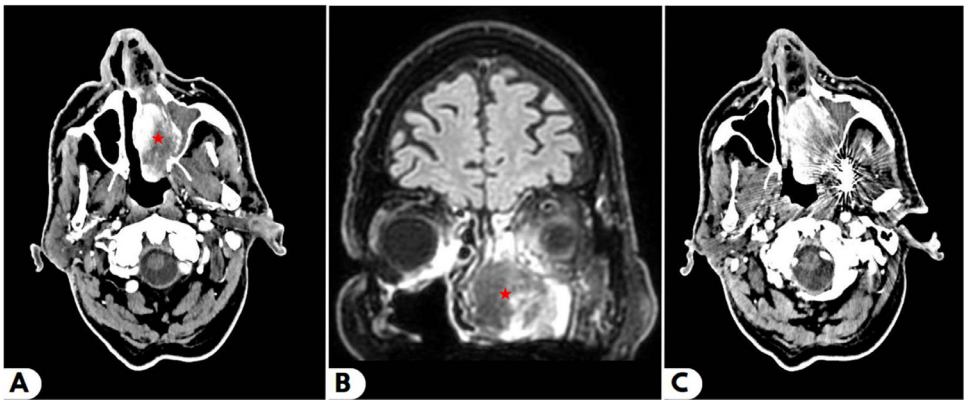

Fig. 1 – (A) Axial sinus CT showing an intensely enhancing highly vascular soft tissue mass lesion measuring 5.5 x 3.2 cm noted in the region of the left posterior choana and nasopharynx (asterisk), extending into the left pterygopalatine fossa with anterior bowing and destruction of the posterolateral wall of the left maxillary sinus. The left maxillary sinus is almost obliterated and opacified. Anteriorly, the lesion is extending to the left nasal cavity with expansion and remodeling of the bony nasal septum. (B) Coronal Sinus MRI showing a heterogeneous mass in the left nasal cavity extending to the nasopharynx with bowing and destruction of the medial wall of the left maxillary sinus and nasal septum. (C) Post-embolization axial CT.

mucosa of the nasal cavity, paranasal sinuses, nasopharynx, oropharynx, and larynx. Treatment modalities for plasmacytoma encompass surgery, radiotherapy, and chemotherapy, as warranted.

Given the rarity of this condition, clinical diagnosis, and treatment pose significant challenges. In this context, we present a case of EMP, confirmed through pathological analysis following surgical resection under nasal endoscopy. This report seeks to share our clinical experience in diagnosing and treating EMP, especially within a high-surgical-risk patient population.

Case presentation

An 89-year-old male patient with a history of hypertension was admitted to our tertiary care hospital with uncontrolled unilateral epistaxis. For the past year and a half, the patient has complained of recurrent epistaxis; frequent nasal packings have alleviated the condition. However, 20 days before admission, the bleeding became more frequent, uncontrollable even with nasal packing, and associated with lightheadedness. The patient denied having ever had surgery before, as well as any history of trauma, bleeding disorders, or digital manipulation. Moreover, there was no family history of head and neck cancers in his past.

Upon admission, the vital signs assessment showed a blood pressure of 136/76 mm Hg, a heart rate of 98 beats/min, a respiratory rate of 19 breaths/min, and an oxygen saturation of 95% on room air. The physical examination revealed a conscious, alert, and oriented patient who appeared pale but was not in pain, had no facial swelling, or was experiencing respiratory distress. However, there was nasal bleeding noted on the left nostril. In addition, the laboratory studies included a complete blood count (CBC), which revealed a hemoglobin level drop from his baseline (13 g/dL) to 9 g/dL and a platelet count of 416 × 10^3/μL. The coagulation profile showed a prothrombin time (PT) of 14.3 seconds, a partial thromboplastin time (PTT) of 25.5 seconds, and an international normalized ratio (INR) of 1.1. These numbers are within the normal range, which suggests that the blood clotting process is working properly.

Computerized tomography (CT) angiography and magnetic resonance imaging (MRI) revealed a 5.5 × 3.2 cm highly vascular mass occupying the region of the left posterior choana and nasopharynx, with destruction of the posterolateral wall of the left maxillary sinus (Figs. 1A and B). Due to the need for urgent intervention to stop the bleeding and the suspicion of angiofibroma, the patient underwent immediate embolization (Fig. 1C). Following the embolization, the patient experienced difficulty swallowing and slurred speech. The brain MRI with contrast showed more damage, including several small strokes above and below the ventricles on both sides, as well as the mass occupying the left nasal cavity (Fig. 2) mostly attributed to chronic small vessel ischemia due to bilateral periventricular white matter involvement.

Despite the embolization, the patient still had epistaxis, and due to the substantial bleeding risk associated with angiofibroma, antiplatelet, and tranexamic acid medications were contraindicated. Blood urea nitrogen was 25 mg/dL and creatinine were 1.6 mg/dL. These results were in line with pre-renal azotemia from blood loss and dehydration as well as contrast-induced nephropathy from the earlier procedure.

Considering these challenges, a multidisciplinary team decided to proceed with mass excision due to nonresolving bleeding. After being made aware of the risks associated with the procedure, the patient and their family signed a high-risk consent form. The patient had functional endoscopic sinus surgery (FESS) with left nasopharyngeal excision of the mass while under general anesthesia. There was not much intraoperative bleeding, and the patient recovered from the procedure a little later than anticipated. After receiving sugam-

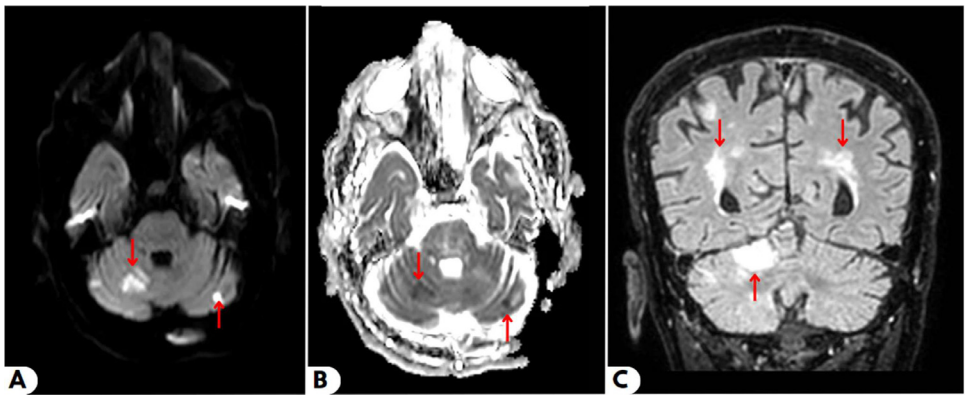

Fig. 2 – (A, B, and C) Brain MRI with axial DWI and ADC sequences, and coronal FLAIR sequence respectively, show an area of diffusion restriction in the right cerebellar hemisphere measuring 1.8 x 1.5 cm, representing acute infraction. There are multiple bilateral supra- and infratentorial acute lacunar infarctions.

madex and naloxone, the patient was eventually able to be extubated and moved to the Surgical Intensive Care Unit (SICU) for postoperative care.

The patient had an intravenous stat dose of Labetalol (5 mg), a Glasgow Coma Scale (GCS) score of 8/15 (E2M4V2), and a blood pressure reading of 200/100 mmHg when they arrived in the SICU. The patient was only dimly conscious at the moment. However, an hour later, the patient's responses were worsening, with a desaturation below 80% and a GCS score of 3/15.

As a result, an emergent crash intubation was performed. A few hours later, the patient showed some response, such as opening eyes and moving limbs, and was sedated with fentanyl injections. After 1 day, the patient demonstrated progressive improvement, was extubated, and was subsequently transferred to the general ward.

Histopathology results, on the other hand, revealed that the removed mass was a plasmacytoma (Fig. 3). The incidence of nasal EMPs in men and women has been shown to be 3:1 in a previous study, and the onset age is usually between 40 and 70 years of age [3]. After establishing the diagnosis, the patient remained stable and did not experience subsequent episodes of epistaxis. He was subsequently discharged from the hospital and referred to another hospital oncology department for further treatment of the plasmacytoma.

Discussion

Solitary extramedullary plasmacytomas (SEP) in the nasal tract occur in three cases out of every 100,000 people each year [4]. The male-to-female ratio is higher than 3:1, and it typically affects people in their 60s to 80s. About 80% of all extramedullary plasmacytomas are characterized by solitary lesions [5].

The etiology of plasmacytoma remains unidentified [6,7], however, there is a theory indicating that EMP might be triggered by the inhalation of chemicals, excessive exposure to radiation, viral infections, and genetic abnormalities in the reticuloendothelial system [8].

SEPs are slowly expanding masses, and individuals affected typically display symptoms of localized disease with a subtle and gradual clinical progression. The predominant clinical manifestations are related to the mass effect, with nasal obstruction and swelling of the soft tissues being the most frequently observed symptoms. In the majority of cases, the initial symptoms consist of nasal obstruction and soft tissue swelling. Less commonly, nasal SEP may present with epistaxis, nasal discharge, cervical lymphadenopathy, and stridor [9]. The disease's nonspecific clinical symptoms, combined with its rarity, contribute to a low level of suspicion, often leading to a delayed diagnosis.

When the patient is examined physically, the tumors are usually found as purple-gray, pedunculated, or sessile polypoid masses in the submucosa. They are prone to easy bleeding with simple manipulations. While the mucosa is generally healthy, advanced-stage cases may exhibit ulcerations and necrosis. Radiological diagnosis is often challenging as CT and MRI findings are non-specific, lacking tumor-specific images [10].

The diagnosis of extramedullary plasmacytomas relies on immunohistochemical staining. Histopathological examination typically reveals a tumor composed of mature plasma cells. Immunohistochemical staining specifically highlights plasma cells associated with monoclonal immunoglobulin, while Congo stain is utilized to detect the presence of amyloid [11]. Our patient's age and persistent bleeding demanded us to act surgically before further confirming the diagnosis by biopsy which led us to an unexpected diagnosis.

Because plasma cells are highly radiosensitive, localized primary radiotherapy has become the standard of care for SEP, especially for larger lesions. Following appropriate radiation therapy, local recurrence rates have been reported to be less than 7% [5]. When it is possible to obtain clear margins for a small tumor with minimal surgical morbidity, surgery is used as the main treatment. Additionally, addressing residual material after radiation therapy becomes a compelling reason for surgical intervention, especially in cases where patients develop mechanical obstruction due to delayed radiation side effects appearing months later. Usually, a combination of radiation and surgery is used for larger tumors.

In summary, the primary manifestation of plasmacytoma is the blockage of the respiratory tract. Early detection is chal-

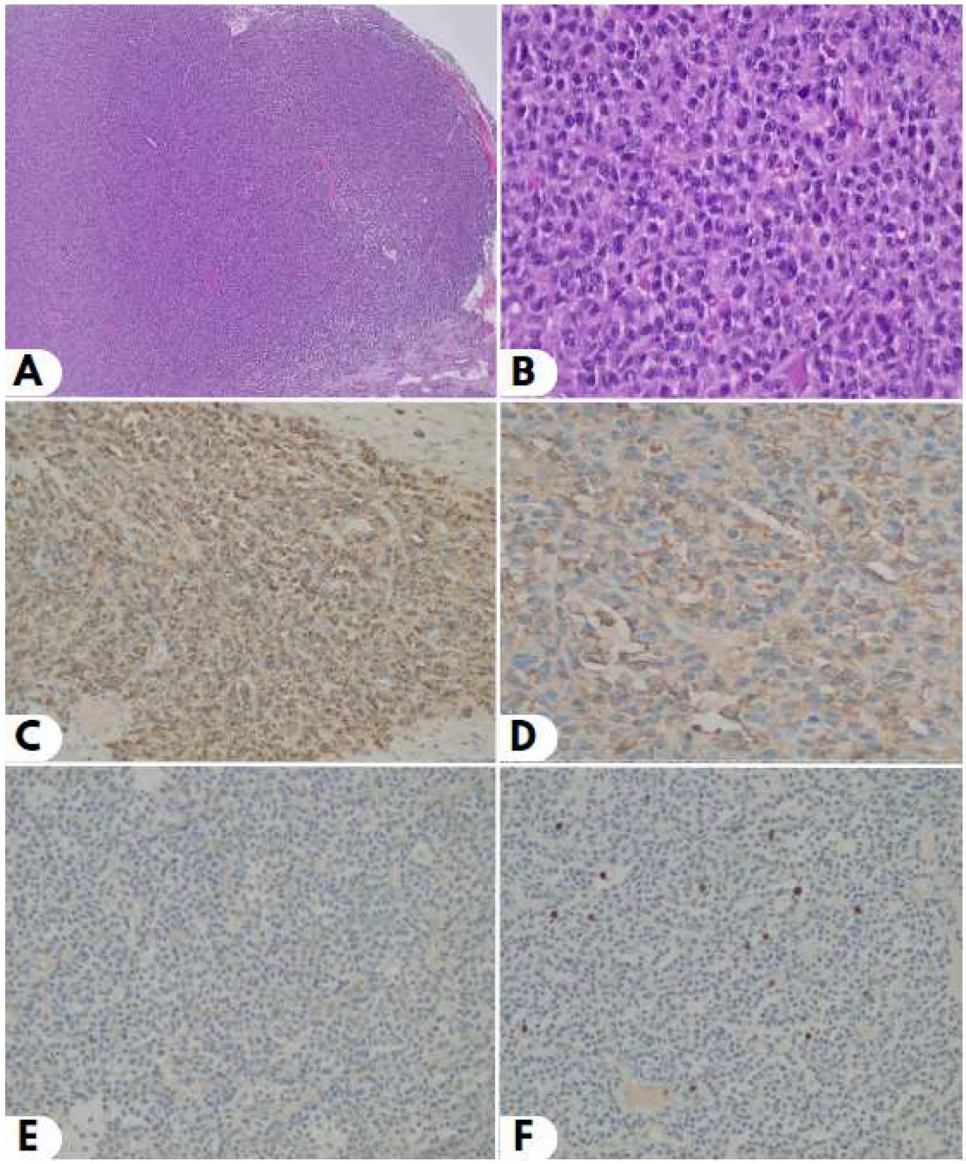

Fig. 3 – (A) Hematoxylin and eosin (H&E) staining of the mass (x 40). Microscopic examination of the mass showed diffuse infiltration by monomorphic cells. (B) H&E staining shows the plasmacytoid morphology of the infiltrate (x 400). The infiltrate is composed of cells with plasmacytoid morphology, including eosinophilic cytoplasm and nuclear cartwheel appearance, and they show mild nuclear atypia. (C) CD138 immunostain shows membranous expression in tumor cells (x 200). (D) CD138 immunostain (x 400). (E) PAX5 immunostain is negative in tumor cells. (F) CD3 immunostain highlights scattered T lymphocytes in the background.

lenging, especially when present with less common manifestations like epistaxis, making it susceptible to misdiagnosis or oversight. Complete removal of the tumor through surgical intervention not only alleviates symptoms but also facilitates an accurate pathology diagnosis for effective treatment.

Patient consent

In accordance with ethical and legal requirements, written informed consent for the publication of this case has been obtained from the patient.

REFERENCES

[1] Iqbal QUA, Majid HJ. Plasmacytoma. StatPearls [Internet], Treasure Island (FL): StatPearls Publishing; 2023. [cited 2024 Jan 1]. Available from http://www.ncbi.nlm.nih.gov/books/NBK573076/.

[2] Corvo MA dos A, Granato L, Ikeda F, de Próspero JD. Extramedullary nasal plasmacytoma: Literature review and a rare case report. Int Arch Otorhinolaryngol 2013;17(2):213–17.

[3] Hu H, Hu X, Hu G, Li D, Cai J. Diagnosis and management of extramedullary plasmacytoma in nasal cavity: clinical experience and literature review. Medicine (Baltimore) 2023;102(2):e32647.

[4] Windfuhr JP, Ott G. Extramedullary plasmacytoma manifesting as a palpable mass in the nasal cavity. Ear Nose Throat J 2002;81(2):110–14.
[5] Sulzner SE, Amdur RJ, Weider DJ. Extramedullary plasmacytoma of the head and neck. Am J Otolaryngol 1998;19(3):203–8.
[6] Lombardo EM, Maito FLDM, Heitz C. Solitary plasmacytoma of the jaws: therapeutical considerations and prognosis based on a case reports systematic survey. Braz J Otorhinolaryngol 2018;84(6):790–8.
[7] Lalla F, Vinciguerra A, Lissoni A, Arrigoni G, Lira Luce F, Abati S. Solitary extramedullary plasmacytoma presenting as asymptomatic palatal erythroplakia: report of a case. Int J Environ Res Public Health 2021;18(7):3762.
[8] Gholizadeh N, Mehdipour M, Rohani B, Esmaeili V. Extramedullary plasmacytoma of the oral cavity in a young man: a case report. J Dent 2016;17(2):155–8.
[9] Wax MK, Yun KJ, Omar RA. Extramedullary plasmacytomas of the head and neck. Otolaryngol–Head Neck Surg Off J Am Acad Otolaryngol-Head Neck Surg 1993;109(5):877–85.
[10] Ching ASC, Khoo JBK, Chong VFH. CT and MR imaging of solitary extramedullary plasmacytoma of the nasal tract. AJNR Am J Neuroradiol 2002;23(10):1632–6.
[11] Atan D, Kilic M, Ozcan KM, Ikinciogulları A, Ensari S, Dere H. Extramedullary plasmacytoma in the nasal cavity. Istanb Med J 2015;16(3):122–4.