Case Report / Olgu Sunumu

Extramedullary plasmacytoma of postnasal space

Arka burun boşluğunda ekstramedüller plazmasitom

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In this article, we present a 75-year-old male with worsened nasal obstruction on the left side. Nasal endoscopy revealed bilateral grade 2 nasal polyps and a large polyp in the left posterior choana. Postnasal space could not be viewed. Urgent computed tomography and magnetic resonance imaging revealed a lesion in the postnasal space. The patient was scheduled for endoscopic excision of the postnasal space lesion and bilateral nasal polypectomy. Histopathological examination of the samples revealed benign nature of polyps and plasmacytoma for postnasal space lesion. The patient is still on radiotherapy. A thorough endoscopic examination of the nasal cavities including the postnasal space is essential in all cases of nasal polyps to avoid postnasal space lesions to be overlooked.

Key Words: Extramedullary; plasmacytoma; prognosis.

Bu yazıda sol tarafta kötüleşen burun tıkanıklığı ile başvuran 75 yaşında erkek bir olgu sunuldu. Yapılan burun endoskopisinde iki taraflı evre 2 nazal polip ve sol arka koanada büyük bir polipe rastlandı. Burnun arka boşluğu görüntülenemedi. Acil bilgisayarlı tomografi ve manyetik rezonans görüntülemede burnun arka boşluğunda bir lezyona rastlandı. Hastaya burnun arka boşluğundaki lezyonun endoskopik eksizyonu ve iki taraflı burun polipektomisi planlandı. Yapılan histopatolojik incelemede poliplerin iyi huylu olduğu ve burnun arka boşluğundaki lezyonun plazmasitom olduğu tespit edildi. Hasta halen radyoterapiye devam etmektedir. Burnun arka boşluğundaki lezyonların gözden kaçırılmaması için tüm burun poliplerinde burnun arka boşluğu dahil burun boşluklarının kapsamlı bir şekilde endoskopik muayenesi şarttır.

Anahtar Sözcükler: Ekstramedüller; plazmasitom; prognoz.

Dalrymple and Bence Jones first described a neoplastic proliferation of plasma cells in 1846. They described a disseminated neoplastic proliferation of plasma cells that was characterized by marked proteinuria and bone pain.[1] Plasmacytoma can be divided into plasmacytoma of bone and extramedullary plasmacytoma (EMP).[2] Extramedullary plasmacytoma represents approximately 3% of all plasma cell neoplasms and 4% of nasal cavity tumors. 80% of these tumors originate in the head and neck region.[3] Extramedullary plasmacytoma mainly occurs between the fourth and seventh decades of life and there is a greater male preponderance.[4] Extramedullary plasmacytoma cases quoted in the literature between 1905 and 1997 were reviewed by

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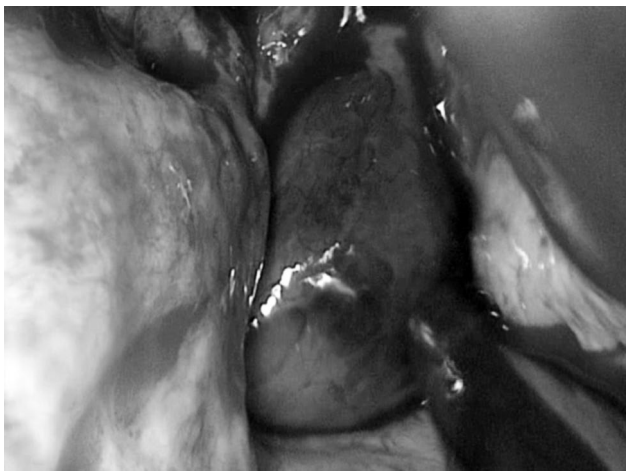

Figure 1. *Intraoperative image showing the lesion occluding the left posterior choana.*

Alexiou et al.[5] from which 82.2% were found in the upper aerodigestive tract. Solitary EMP of the nasopharynx is very rare.[6,7] We report a case of EMP of the postnasal space (PNS).

CASE REPORT

A 75-year-old male presented with nasal obstruction worse on the left side. He also had post-nasal drip and hyposmia but there was no history of headaches, facial pain, asthma or hayfever. He had septal surgery in 1983 for a previous road traffic collision.

Nasendoscopy revealed bilateral grade 2 nasal polyps and a large polyp in the left posterior choana with no view of the PNS (Figure 1). Urgent computed tomography (CT) and magnetic resonance imaging (MRI) scan revealed a focal lesion (27x17x19 mm) arising from the roof of the left nasopharynx with no skull base extension (Figures 2, 3). The patient was listed for endoscopic examination under anesthesia (EUA)/bilateral nasal polypectomy and excision of the polypoidal PNS lesion. The PNS lesion was completely excised and monopolar diathermy was applied to the base of the lesion (Figure 4). The nasal polyps and the PNS mass were sent for histology.

Histology showed the nasal polyps to be benign and the PNS lesion was revealed to have diffuse population of plasma cells with aberrant loss of CD19 consistent with a neoplastic phenotype consistent with plasmacytoma. The patient then had radiological skeletal survey with bone marrow biopsy and serum electrophoresis, under the care of hematologists and multiple myeloma was excluded. At recent ENT review the left side of the PNS appeared healthy with no residual lesion. He is undergoing radiotherapy now under the care of an oncologist.

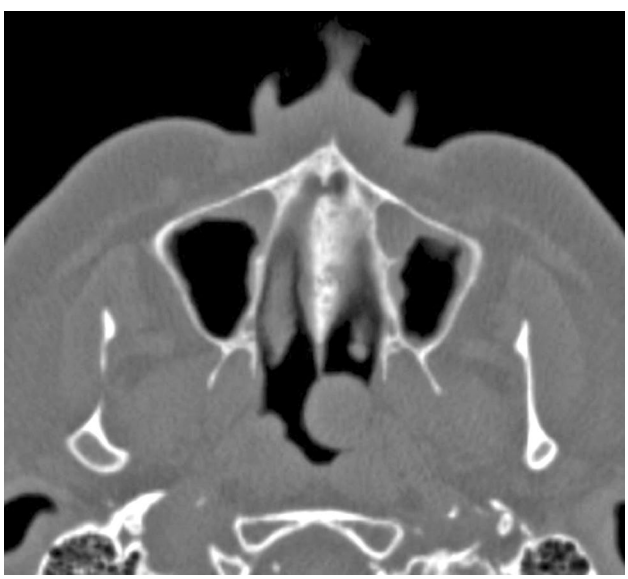

Figure 2. *Axial computed tomography scan showing the lesion in the left side of the postnasal space.*

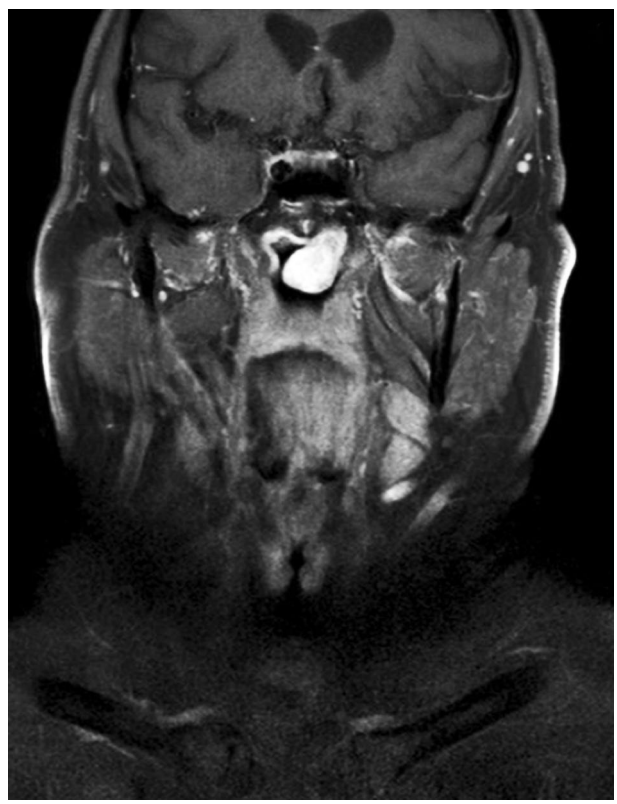

Figure 3. *Coronal magnetic resonance imaging scan, post contrast showing the lesion arising from the superior-lateral wall of the nasopharynx.*

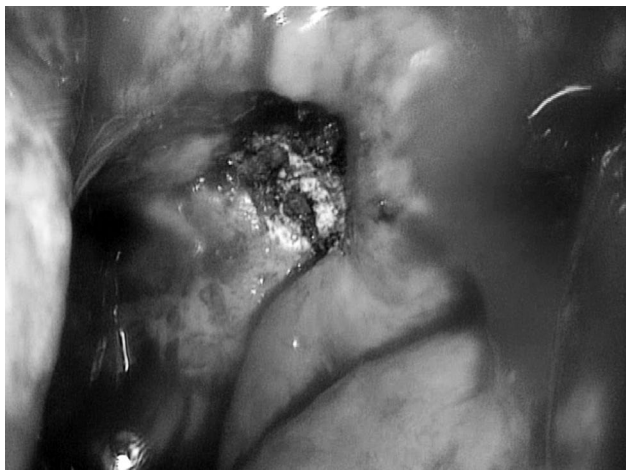

Figure 4. *Image showing the left side of postnasal space post excision.*

DISCUSSION

Neoplasms of plasma cells include three separate entities: multiple myeloma, or disseminated disease, and two localized diseases, namely solitary bone plasmacytoma and solitary plasmacytoma of soft tissue or EMP. Plasma cells are differentiated secretory forms of B lymphocytes, and therefore, these neoplasms are considered as low-grade lymphomas.[8] Extramedullary plasmacytoma is a localized collection of monoclonal plasma cells located in an extra-skeletal site. The age range is 50-70 years with a male: female ratio of 3:1.[4,8] Megat Shiraz et al.[8] and Korolkowa et al.[9] reported that 40% occur in the nasal cavity and paranasal sinus, 20% in the nasopharynx, and 18% in the oropharynx. Approximately 10% of EMPs have multiple sites of involvement.[8]

The diagnosis of plasmacytoma is based on histology, and its specific immunoglobulin secretor type can be determined by immunocytochemistry. The deposition of amyloid in the stroma, although a feature, is not a diagnostic finding. Amyloid is expressed by 15 to 38% of EMP as reported by Sulzner et al.[10] The diagnosis of solitary plasmacytoma must be made after careful exclusion of the simultaneous presence of other plasma cell tumors by a negative bone scan and a normal bone marrow aspiration study. The histopathological diagnostic criterion laid down by Knowling et al.[11] requires an absence of neoplastic plasma cells in the bone marrow biopsy although Corwin and Lindberg[12] and Mendelhall et al.[13] would accept the presence of up to 10% of plasma cells in the bone marrow biopsy. Detection of myeloma protein in serum and Bence Jones protein in urine is uncommon in solitary plasmacytoma. A monoclonal band of serum protein is expressed by approximately 25% of EMPs at an early stage.[14] The commonest immunoglobulin expressed by the tumor cells is immunoglobulin (Ig)G with kappa chain restriction.[8,15]

Plasmacytoma can be graded low (grade 1), intermediate (grade 2) and high (grade 3), based on the cellular atypia. Based on the serum, urine electrophoresis, bone scan, bone marrow examination and radiological assessment, EMP can be staged according to the spread of the disease. Stage I is disease confined to one site. Stage II includes tumors with local extension of lymph node involvement. Stage III has metastatic spread.[8] In our case there was no evidence of infiltration either to the skull base or to the bone marrow and therefore our patient was staged as grade I.

According to the literature, our patient would be put in the category of 20% of cases where EMP occurs in the nasopharynx. His tumor histology did not show IgG with kappa chain restriction as suggested by our literature search but did show plasma cells with aberrant loss of CD19.

The reported conversion rate of EMP to multiple myeloma is 15-20%, and is associated with a poorer prognosis. Dissemination of the tumor takes place in 35-50% of EMP.[8,16]

Excellent control of solitary plasmacytoma can be achieved using radiotherapy with good long-term survival.[17,18] According to the guidelines; the recommended primary treatment for localized EMP is radical radiotherapy. For generalized EMP, chemotherapy is advisable.[8,19,20]

Therefore, radiotherapy was the treatment of choice in this patient who had solitary EMP.

In conclusion, EMP is rare in its own right and EMP of the PNS is even rarer. Moreover this patient's PNS lesion could have easily been missed with just the finding of nasal polyps which would have explained his symptoms. Therefore a thorough endoscopic examination of the nasal cavities including the PNS is essential in all cases of nasal polyps to exclude a second pathology. Although a CT scan would have picked up the lesion, we do not routinely perform imaging in all patients with nasal polyps.

We achieved very good local control with our patient and it is our recommendation for all

patients with solitary EMP to have radiotherapy. As with our patient, all solitary EMP patients will require long term follow-up in partnership with the Hematologists and Oncologists for risk of recurrence or conversion to multiple myeloma. Urinary and serum myeloma should be assessed at regular intervals for the above mentioned risk.

Declaration of conflicting interests

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