



Extramedullary Plasmacytoma in the Nasal Cavity

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Abstract

Plasmacytomas are malignant tumors characterized by abnormal monoclonal proliferation of plasma cells. They originate either in the bone (osseous solitary plasmacytoma) or in soft tissues (extramedullary plasmacytoma) and could be either primary or a part of a systemic process during the course of multiple myeloma.

Keywords: Plasmacytoma, nasal cavity, multiple myeloma

Introduction

Extramedullary plasmacytomas (EMPs) are rare tumors developing as a result of the monoclonal proliferation of plasma cells (1). They constitute less than 1% of all head and neck malignancies, and their incidence is 0.03%. They are more commonly seen in men than in women, and their incidence rises with increasing age. EMP etiology is not completely known. It is mostly in the form of solitary masses but may also be observed as multiple masses. The most common localization site of EMPs is the upper respiratory tract submucosa. The most important reason they are seen in this region is that the submucosa is rich in plasma cells (2). In the large case series, conducted by Alexiou et al. (3), the upper airway tract was detected as the localization site in 714 (82.2%) of 869 EMP patients. The most common localization sites in the upper airway tract are the nasal cavity and paranasal sinuses (43.8%), followed by the nasopharynx (18.3%), oropharynx (17.8%), and larynx (11.1%). The rare occurrence of the tumor, its slow progress, and its nonspecific findings make diagnosis difficult. The tumor has a destructive character and may recur. It can cause lymph node metastasis and can progress to multiple myeloma (MM) (4). The most common clinical symptoms are nasal congestion, epistaxis, and nasal discharge. The differential diagnosis of MMs from solitary medullary plasmacytomas has to be made by examinations such as serum electrophoresis, Bence Jones proteinuria, and bone marrow examination. Radiotherapy is the ideal treatment because it is a radiosensitive tumor. Surgery is ideal to provide local control; however, due to the tumor's localization and greatness, its radical excision is mostly impossible. The effectiveness of chemotherapy is unclear (5).

Case Report

A 53-year-old patient having nasal congestion and nasal discharge complaints for six months was admitted to our clinic. In an endoscopic examination, a 4x5 cm dark, purple, and polypoid mass that originated from the left nasal cavity base, extended into the nasopharynx, and covered the choana by completely filling the nasopharynx was observed. No lymphadenopathy was detected on the neck in the physical examination (Figure 1).

A polypoid mass, having a heterogeneous appearance originating from the left nasal cavity floor posterior, reaching the nasopharynx, and almost completely filling the nasopharynx, was observed. Bone destruction was not observed (Figure 2). Endoscopic excisional biopsy was performed on the mass (Figure 3). Histopathological diagnosis was reported to be EMP. CD138, CD38, CD79a, lambda, and diffuse staining were observed in tumor cells after an immunohistochemical study was conducted. Partly staining was observed with CD117 and LCA. There was no staining with kappa, CD20, CD99, S-100, Melan A, HMB45, synaptophysin, chromogranin, MyoD1, and myogenin. The Ki67 proliferation index was evaluated to be 10–15% in the densest place.

The diagnosis of MM was eliminated, and that of sinonasal EMP was made definite. No radiotherapy was given to the patient after the operation. No symptom was encountered for recurrence to be considered in the 1-year follow-ups of the patient (Figure 4). Written informed consent was taken from the patient.

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Figure 1. The mass originating from the left nasal cavity base and extending to the nasopharynx



Figure 4. The endoscopic image on the 1st postoperative month

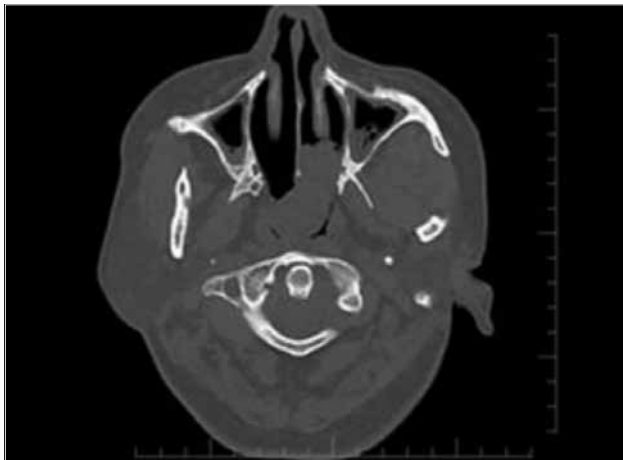


Figure 2. The image of the mass filling the nasopharynx on axial paranasal CT
CT: computerized tomography



Figure 3. The macroscopic image of the postoperative mass

Discussion

Extramedullary plasmacytomas may occur in the head and neck region as a single mass at the rate of 80% and as multiple masses at the rate of 10–20%. They are tumors having atypical symp-

toms, and they slow clinical course. Clinical complaints generally occur because of a mass formed in the region where the tumor developed. The most common involvement occurs in the sinonasal region and nasopharynx. It was reported in literature that there may be rare involvement in the tonsils, minor salivary glands, posterior pharyngeal wall, thyroid gland, parathyroid gland, middle ear, colon, and liver (6). The most common complaints in sinonasal involvement are nasal congestion, nasal discharge, and nosebleeds (7). In the EMP series of Kapadia et al. (4) involving 20 patients having head and neck localization, the most common complaint was the mass complaint observed in 80% of the patients. Furthermore, airway obstruction was observed in 35% of the patients, epistaxis in 35%, localized pain in 20%, proptosis in 15%, regional lymphadenopathy in 10%, and abducens palsy in 5%. Our patient had a single mass localized in the nasal cavity, and he applied to our clinic late because he only had complaints of nasal congestion and nasal discharge. If there was the complaint of nosebleeds, the tumor could have been diagnosed before.

In the physical examination, the tumors are generally observed to be purple–gray, pedunculated, or sessile polypoid masses in the submucosa. They easily bleed with simple manipulations. The mucosa is generally healthy, but ulcerations and necrosis can be observed in advanced stage cases. During the surgery, intact mucosa on the mass and any ulceration and/or necrotic region were not observed. A radiological diagnosis is generally difficult because the findings of computerized tomography (CT) and magnetic resonance imaging (MRI) are non-specific (8). No tumor-specific image was observed in CT and MRI examinations.

Extramedullary plasmacytomas diagnosis is made by immunohistochemical staining. Generally, the tumor formed by mature plasma cells is observed in the histopathological examination. When immunohistochemical staining is conducted, plasma cells associated with monoclonal immunoglobulin are stained. Congo stain is involved in the presence of amyloid (9).

Immunohistochemical staining also provides information about the tumor structure by indicating the monoclonal structure of plasma cells. Additionally, the differential diagnosis of undiffer-

entiated tumors, melanomas, and esthesioneuroblastomas can be made by immunohistochemical staining (7).

Solitary plasmacytomas are radiosensitive tumors. The standard use of radiotherapy in solitary plasmacytoma is an accepted view. There is a view that surgical excision is insufficient and that radiotherapy is recommended after excision even if wide excision is conducted. Surgical excision is an acceptable treatment method for EMPs, and radiotherapy is recommended only in case of surgical margin positivity (10). Sometimes, tumor sizes are so large that surgery may not be possible (11). In our patient, because the tumor was small and appropriate for resection, surgical treatment was performed. Moreover, as it did not have surgical margin positivity, radiotherapy was not given after surgery and no relapse was detected in the 1-year follow-ups.

The mean survival rate is between 4 and 10 years, and local recurrence is 8–30%. Another important point is that EMP MMs can progress. In a study conducted, it was reported that 8–36% of the patients progressed to MMs in 3–61 months (12). For this reason, patients have to be followed-up throughout their life. MM was excluded in our patient, and no MM progression was observed in the 1-year follow-ups. CT scan, serum immunoglobulin, and urinary Bence Jones protein can be used in indicating recurrence and MM progression (13).

Conclusion

Extramedullary plasmacytomas are rare masses seen in the nasal cavity. They must be kept in mind in the differential diagnosis of intranasal masses. A multidisciplinary approach is important in the diagnosis and treatment. Systemic involvement must be eliminated.

Informed Consent: Written informed consent was obtained from the patients.

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