Rare Neoplasm of Inferior Turbinate: Extramedullary Plasmacytoma

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Abstract

Plasma cell disorders can be divided into two subgroups: multiple myeloma and plasmacytomas. Extramedullary plasmacytoma is a rare clinical entity found in the head and the neck, frequently located in the nasal/paranasal region. This kind of masses cause to gradually growing symptoms according to their location. It is important to distinguish plasmacytoma from other intranasal masses. In this article, we discussed differential diagnosis, treatment process and follow up of an extramedullary plasmacytoma originated from inferior turbinate with the literature.

Keywords: Inferior turbinate, extramedullary plasmacytoma, endoscopic sinus surgery

INTRODUCTION

Plasma cell disorders are found in the body in two forms: as localized and generalized. The generalized form is multiple myeloma (MM) involving primarily the body and the skull. Localized ones are solitary plasmacytomas involving the bone marrow, and extramedullary plasmacytoma involving the soft tissue (1).

Extramedullary plasmacytoma (EMP) comprises less than 3% of plasma cell disorders. It can arise anywhere throughout the body, majority being present in the head and neck region. It comprises less than 1% of all head and neck malignancies (2, 3) EMP occurs in organs such as larynx, oropharynx, tongue, tonsil, thyroid, and parathyroid throughout the head and neck region, with 75% occurring in the nose and paranasal sinuses (4).

Depending on the EMP's involvement sites in the head and neck region, symptoms including the nasal obstruction, rhinorrhea, otalgia, proptosis, epistaxis, cranial nerve paralysis, dysphonia, dysphagia, hemoptysis, and stridor can be seen (3). To differentiate between these diseases, a radiological examination should be performed, and a final diagnosis established via biopsy.

Considering the localization of the tumor, symptoms, and the patient's status, it should be treated by surgery, radiotherapy, and combined approaches (1).

CASE REPORT

A 52-year-old male patient was admitted to our clinic complaining of increased right-sided nasal congestion that lasted for the past 2 years and occasional epistaxis for the past 3 to 4 months. No finding was detected on the patient's anterior rhinoscopy, except septum deviation. A mass filling the right nasal cavity (Figure 1) was found on the endoscopic examination. No pathology was detected on the ear-nose-throat and systemic examination.

Paranasal computed tomography (CT) showed a lobular contour mass approximately 3.5 cm in size, extending to the median line in the right nasal cavity (Figure 2). Paranasal magnetic resonance imaging (MRI) revealed the mass originating from the inferior turbinate and showing vascularity (Figure 3). All other biochemical and hematological tests of the patient were normal. After obtaining the patient's informed consent, endonasal endoscopic biopsy was conducted under general anesthesia.

During the operation, it was found that the mass originated from the right inferior turbinate and extended to the septum and the middle turbinate (Figure 4). Intraoperative frozen section pathology was performed on the mass. Plasmacytoid cells were detected on the frozen section pathology of the samples, but no differentiation between a benign/malignant tumor could be made. The mass including the inferior turbinate was endoscopically excised along with the safety surgical margins (Figure 5). For convenient postoperative follow-ups, the patients also underwent septoplasty. No pre- and postoperative complications were observed.

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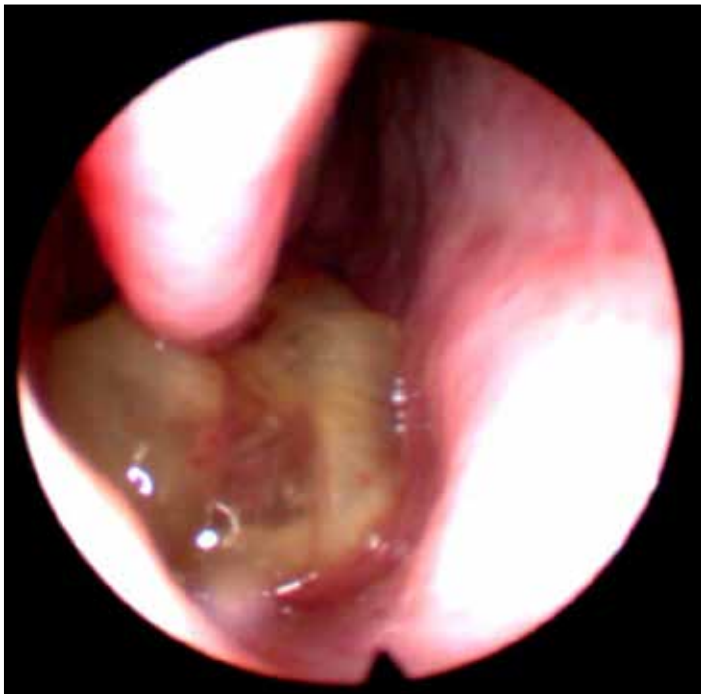

Figure 1. Mass originating from posterior part of the inferior turbinate and extending to the septum in the right nasal cavity

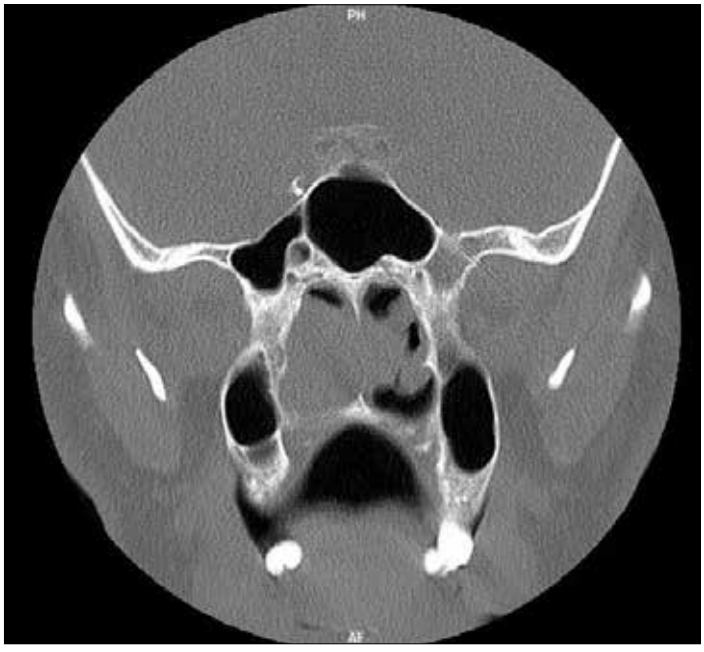

Figure 2. Coronal paranasal sinus CT image of the mass in the right nasal cavity

The patient was referred to the hematology clinic to distinguish between MM and plasmacytoma since the pathology results of the patient were reported as a tissue sample showing lambda monotypic/atypic plasma cell infiltration.

The hematology clinic evaluated of patient's serum immunoglobulin values, Lambda and Kappa protein levels in urine and serum, Beta 2 microglobulin in urine, the light chain protein measurement in urine, and

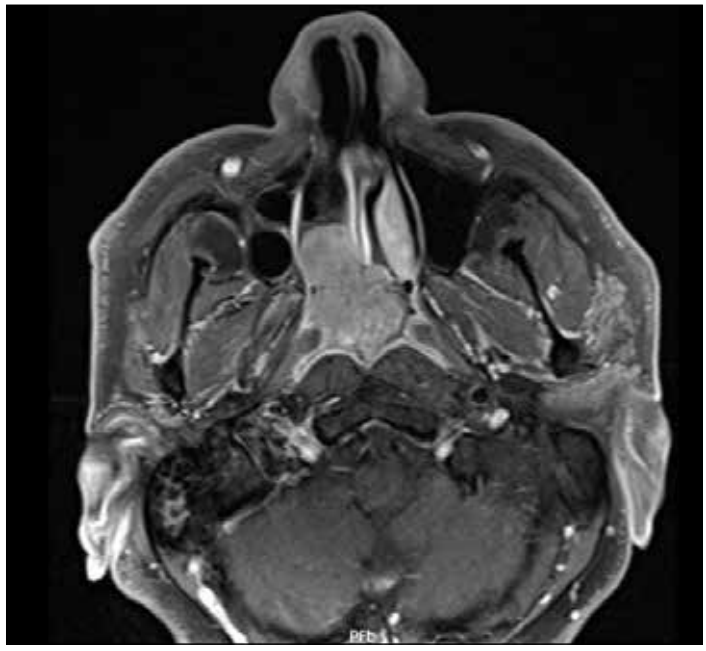

Figure 3. Contrast-enhanced T1 axial paranasal sinus mri image of the mass

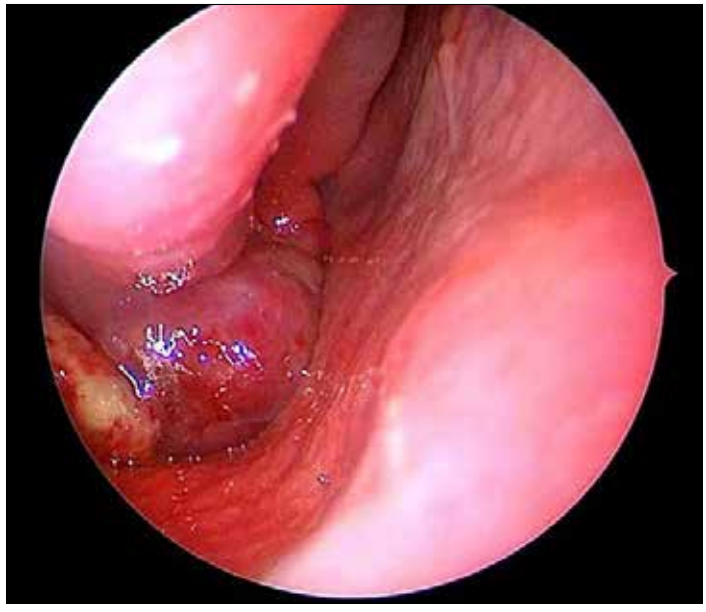

Figure 4. Intraoperative endoscopic image of the mass originating from the inferior turbinate and extending to the choana in the right nasal cavity

urine and serum protein electrophoresis. Positron emission tomography (PET), CT, and bone marrow biopsy were performed as secondary analyses on the patient showing no findings suggesting MM with no monoclonal protein increase in electrophoresis, and a normal Kappa/Lambda ratio. MM was eliminated due to the absence of pathological involvement in the body and no monoclonal cell increase in the bone marrow on PET; the patient was diagnosed with extramedullary plasmacytoma, and no additional treatment was needed. No recurrence was detected on the 18-month follow-up of the patient.

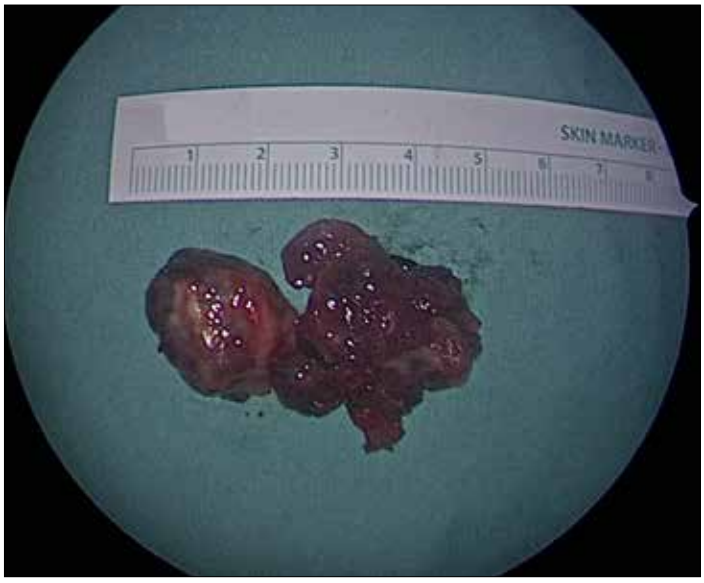

Figure 5. Image of the totally excised mass

DISCUSSION

Plasma cell disorders are malignancies associated with the monoclonal proliferation of plasmocytes originating from B-lymphocytes. They are seen in the body in two forms, as localized and generalized. The generalized form is MM involving primarily the body and the skull.

Solitary plasmacytoma is characterized by the uncontrolled proliferation of plasma cells in the bone marrow at a localized focus, while this uncontrolled proliferation in extramedullary plasmacytoma appears in the soft tissue. The EMP etiologies are not fully clarified. However, their location, which is rich in the mucosa-associated lymphoid tissue, suggest that inhaled allergens, chronic viral agents, and exposure to infection may have a role in the etiology (2, 4)

Extramedullary plasmacytomas (EMPs) are tumors usually occurring in the 6th to 8th decade of life, with an annual incidence of 3 per 100,000 population and are approximately 3 times more common in men than in women. In the literature, it is most commonly detected in the head and neck region (90%). As in our case, it is primarily observed in the nasal cavity, nasopharynx, and paranasal sinuses (75%), and then in the larynx and oropharynx (5-11). Other rarely seen sites include minor salivary glands, tongue, tonsil, thyroid, and parathyroid, colon, and liver. There are reported cases showing involvement in the middle ear as well (12-14).

The symptoms of EMP vary depending on the located site, and the most common symptoms include nasal obstruction, rhinorrhea, epistaxis, dysphagia, dysphonia, otalgia, cranial nerve paralysis, and proptosis. The symptoms are usually similar to symptoms of other diseases. The incidence is very low, and thus the EMP diagnosis is made during the late stage. The most important diagnostic step is biopsy. The fine-needle aspiration is not recommended as a small amount of tissue sample will be taken in the EMP of the head and neck region. These tumors are frequently located in the submucosa, and therefore, deep biopsy is required (2).

The first procedure to be performed on patients with plasma cell tumors is the exclusion of a possible common involvement. In the literature, the presence of near 10% of MM was stated during the diagnosis of EMP (15, 16). The first analyses to be performed for differential diagnosis are the blood count, biochemical examinations, serum protein values, and protein electrophoresis. Imaging methods such as direct graphy, CT, MRI, and PET CT can be used with these analyses in terms of the bone involvement. For the differential diagnosis of localized and generalized forms, it is supposed to be demonstrated that no monoclonal increase is observed in plasma cells by conducting a bone marrow biopsy (1).

As in our case, findings such as bone pain, hypercalcemia, and renal failure suggesting MM are not commonly seen in preoperative period. In laboratory tests, only 25% of patients had elevated levels of protein M in urine or blood. For EMP, which is rarely seen in the head and neck region, although it is not advantageous to perform a protein M analysis on all patients, elevated or reduced levels at the threshold limit value lead us to eliminate only MM (6).

Localized plasmacytomas need a long-term follow-up. Solitary bone plasmacytomas have a worse prognosis compared to extramedullary plasmacytoma. Most frightening situation in the long-term follow-ups of these lesions is the risk of progression to MM. Solitary plasmactyomas of bone progress to MM at a rate of 65%-84% in 10-year follow-ups and at a rate of 100% in 15-year follow-ups (15). It was noted that this risk in extramedullary plasmacytoma was at a rate ranging from 17% to 34% in life-long follow-ups. Five-year survival rates of cases with progression of solitary and extramedullary plasmacytoma to MM were reported as 33% and 100%, respectively (1). No progression to MM was observed in a 12-month follow-up in our case; however, a long-term follow-up of this patient is required.

The treatment choice of plasmacytomas is surgery or radiotherapy. Sometimes combined therapies can be also used. The radiosensitivity of solitary and extramedullary plasmacytomas resulted in using radiotheraphy as the first treatment option (17). Due to surgically easily inaccessible locations and surgical morbidity, radiotherapy is preferred treatment of EMPs of the head and neck region in many cases. In the review conducted by D'Aguillo et al. (16), 89 of 175 patients in the literature were treated with only radiotheraphy.

Surgical treatment has several advantages, including the first treatment option in persistent masses following radiotheraphy, avoiding morbidities caused by radiotheraphy, and total extirpation of tumors in several locations (18). In the study by Gerry et al. (19), EMPs of the head and neck region show a higher rate of disease-free survival and overall survival compared to other EMP localizations. In the review by D'Aguillo et al. (16), surgical treatment was performed at a high rate, as much as a combined therapy of surgery and radiotherapy for the last 3 decades due to increased frequency of MM in long term, following radiotherapy applied to EMPs of the head and neck region, and development of surgical instruments and techniques. It has been shown that surgical treatment and combined therapy with surgery had a better 10-year survival compared to radiotherapy (19).

In the literature review, an inferior turbinate-located similar mass was diagnosed as EMP by biopsy. Following diagnosis, a PET examination performed after the treatment of the patient receiving curative intent radiotherapy for inferior turbinate and neck lymph nodes revealed that the radiotherapy response of the mass was insufficient. The patient underwent the endoscopic endonasal approach to pterygopalatine fossa as a salvage therapy. The patient who showed no recurrence on MRI and endoscopic examination during the 3-month follow-up was referred to the hematology service for systemic involvement (20).

In this case, the origin of the mass detected during the endoscopic examination and its prevalence were investigated with radiologic imaging methods. It was diagnosed by frozen section examination during the endoscopic biopsy. The final diagnosis was established with total resection of the inferior turbinate-originated tumor, and in this case a differentiation between a benign or malignant tumor could not be made on frozen section pathology. The patient was diagnosed with EMP based on the pathology results following hematology consultation. Laboratory analyses, PET, bone marrow biopsy, and molecular research performed for MM differentiation revealed that the patient had a limited EMP. Adjuvant therapy was not recommended for the patient since the tumor was totally resected; thus, the morbidity and cost of the adjuvant therapy were also avoided.

CONCLUSION

As extramedullary plasmacytoma is rarely seen and is most commonly located in the head and neck region, for such cases, a good knowledge about the location of the head and neck lesions, diagnosis, treatment, and follow-up variations is required. The knowledge of local and systemic variations of this disease increases the accuracy of diagnosis and treatment methods. In limited cases, total resection of the tumor especially located in the nasal cavity by endoscopic endonasal surgery alone is a sufficient and distinguished method without any need of adjuvant therapy.

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

Peer-review: Externally peer-reviewed.

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