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Extramedullary plasmacytoma of the septum

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Abstract. *Extramedullary plasmacytoma of the septum. Problem*: Extramedullary plasmacytoma of the head and neck is a rare neoplasm characterized by monoclonal proliferation of plasma cells. The nasal cavity and nasal septum are the most common sites of occurrence and the neoplasm can be solitary or multiple. Extramedullary plasmacytoma is associated with the initial appearance of multiple myeloma and may precede overt manifestations of systemic disease by months or years.
Methodology: A seventy-year-old female presented to our clinic with a one-month history of nasal obstruction. We performed a systematic approach to diagnosis using clinical, laboratory, and radiologic investigations in order to exclude systemic involvement.
Results: The patient was diagnosed with extramedullary plasmacytoma and the mass was excised completely via transnasal endoscopy.
Conclusion: Extramedullary plasmacytoma of the nasal cavity is rare and should be considered in the differential diagnosis of nasal cavity masses.

Introduction

Extramedullary plasmacytomas account for 4% of all head and neck malignancies and approximately 80% are localized in the submucosa of the upper respiratory tract.[1] The nasal cavity and nasal septum are the most common locations.[1] Extramedullary plasmacytoma of the upper respiratory and digestive tracts can be solitary or multiple.[1] They are associated with multiple myeloma at its initial appearance may precede the development of systemic disease by months or years.[1]

Case report

A seventy-year-old female presented with a one-month history of nasal obstruction. Her medical history included hypertension. Clinical examination revealed a dark red mass in the right side of the nasal septum that was blocking the nasal cavity (Figure 1). No cervical lymph nodes were palpated. Computed tomography (CT) revealed a 3×1.5-cm mass located in the anterior part of the nasal cavity. The mass had thinned the nasal septum and the perimeter of the mass was not discriminated from middle and inferior concha (Figure 2). Punch biopsy was taken from the mass under local anaesthesia and histopathological analysis indicated the mass to be plasmacytoma. Hematologic and biochemical tests, urine analysis, and radiologic skeletal survey were performed to screen for multiple myeloma. All test results were within normal limits. Multiple myeloma was ruled out and extramedullary plasmacytoma of the nasal septum was confirmed. The intranasal mass was removed completely via transnasal endoscopy. Histopathologic examination of the specimen revealed diffuse atypical plasma cell infiltration that had ulcerated the surface epithelium and destroyed the nasopharyngeal glands (Figure 3). Immunochemical analysis was diffusely positive for CD38 and kappa light chain and negative for lambda light chain (Figure 4). Histopathological and immunohistochemical findings confirmed the diagnosis as extramedullary plasmacytoma. Post-operative CT scan confirmed completed removal of the tumour (Figure 5). The patient exhibited no evidence of recurrence at two months follow-up.

Discussion

Plasmacytomas arising from neoplastic proliferation of antibody-producing plasma cells are classified as multiple myeloma or extramedullary plasmacytoma, according to the site of development and clinical features.[1] They are further classified as either

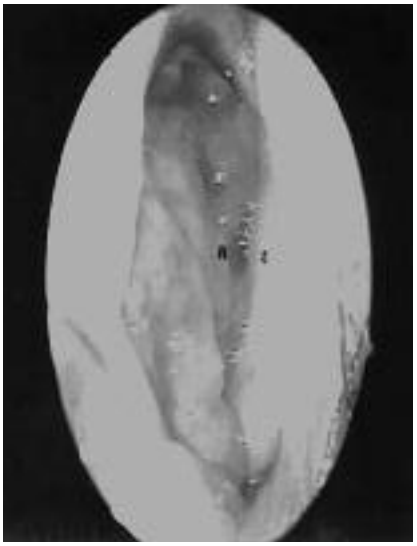

Figure 1
Dark red mass in the anterior part of right nasal cavity (M: Mass; S: Septum).

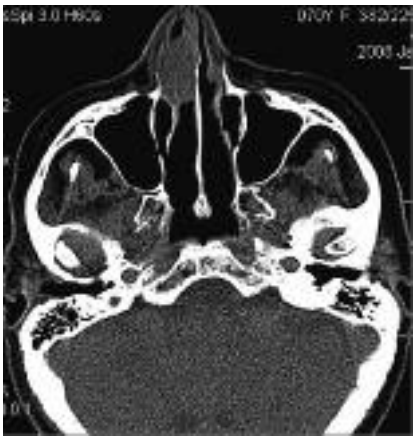

Figure 2
Coronal computed tomography image of extramedullary plasmacytoma in the right nasal cavity. The mass is located in the anterior part of right nasal cavity. The mass has caused thinning of the nasal septum and the perimeter of the mass is not differentiated from the middle and inferior concha.

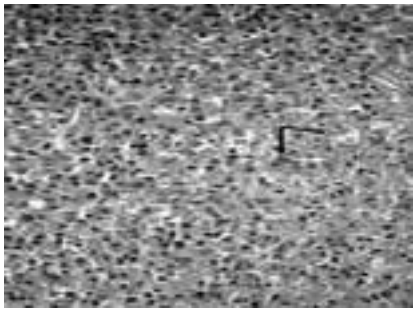

Figure 3
Photomicrograph showing the diffuse atypical plasma cell infiltration and entrapped acini (arrows; hematoxylin and eosin × 200).

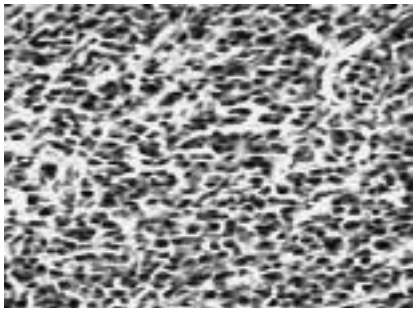

Figure 4
Immunohistochemically diffuse kappa light chain positivity in atypical plasma cells (Kappa, × 400).

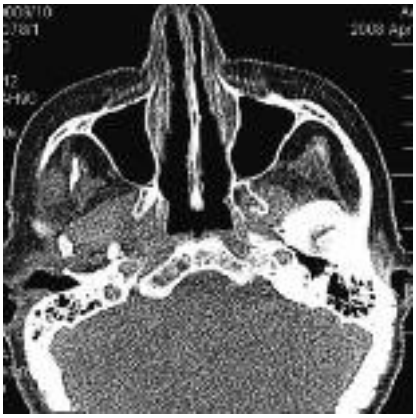

Figure 5
Post-operative computed tomography scan showing complete removal of the tumour.

localized (stage I), localized including local lymph nodes (stage II), or generalized (stage III) according to their clinical manifestation.[2]

Extramedullary plasmacytoma shows a predilection for the head and neck region and the nasal cavity is the most common site of occurrence.[2] Other sites in the head and neck include nasopharynx, maxillary sinus, thyroid gland, soft tissues of anterior cervical region, parotid gland, tonsil, oropharynx, larynx, orbit, choroid and eyelid, sphenoid bone, mastoid, calvaria, skull vault, hyoid bone, temporomandibular joint, maxilla, and mandible.[2]

Extramedullary plasmacytoma of the upper respiratory and digestive tracts can be solitary or multiple.[2] They are associated with multiple myeloma at its initial appearance and may precede overt development by months or years.[2] Therefore a systematic approach to disease staging, including complete blood counts, renal and liver function tests, serum and urinary protein electrophoresis, serum immunoglobulin level evaluation, a skeletal survey, bone marrow examination, and CT of the tumour region must be performed in order to exclude systemic involvement.[3] We performed all of these laboratory and radiological tests for our patient and ruled out multiple myeloma.

Extramedullary plasmacytomas usually manifest in patients between the ages of 50 and 60 years and are more common in men than in women.[3] Our patient was a 70-year-old women. Extramedullary plasmacytoma of the nasal cavity presents with local symptoms of obstruction, discharge, bleeding, and pain, depending on the tumour subsite.[4] Proptosis, cervical lymphadenopathy, and cranial nerve palsy are less common findings.[3] Our patient presented with epistaxis, nasal obstruction, and nasal discharge.

Diagnosis of extramedullary plasmacytoma of the nasal tract requires histologic and immunohistochemical evidence.[1] Galieni established five criteria for diag-

nosing extramedullary plasmacytoma: 1) monoclonal plasma cell histology in tissue biopsy, 2) bone marrow plasma cell infiltration <5% of all nucleated cells, 3) no osteolytic bone lesions or other tissue involvement, 4) absence of hypercalcemia and renal failure, and 5) if present, low serum M-protein concentration.[1] Our patient had four of the five criteria for diagnosis. We did not perform bone marrow biopsy.

Differential diagnosis includes other nasal masses, including epidermoid and non-epidermoid neoplasms of the nose and paranasal sinuses.[5] Benign tumours include osteoma, hemangioma, papilloma, and angiofibroma.[6] Malignant tumours are squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, sarcoma, and malignant melanoma.[6] Concha pyocele may obstruct the osteomeatal complex and mimic an intranasal mass.[7] In addition, angiomatous polyp of the maxillary sinus may extend through the nasal cavity and should therefore be considered in the differential diagnosis.[8]

The treatment for localized extramedullary plasmacytoma is challenging and includes radiotherapy, surgery, or a combination of both.[9] Transmaxillary and transpalatine approaches may cause stress for the patient, whereas the endonasal approach incurs the possibility of incomplete resection.[9] Endoscopic sinus surgery using the KTP/532 laser enables resection of the residual tumour safely and accurately without incising the soft palate.[9] Often, the only curative option for recurrent extramedullary plasmacytomas is surgery.[1] Chemotherapy is advised for generalized extramedullary plasmocytoma.[1] Radiotherapy does not always reduce the size of the tumour because of an abundant deposition of amyloid within the mass.[9] Surgery was preferred for our patient and the mass was excised totally via endoscopic sinus surgery.

The clinical course extramedullary plasmacytoma is unpredictable. Local recurrence may occur in 6-10% of cases.[10] Five-year survival ranges between 53-75%. Development of multiple myeloma occurs in 10-32% of patients within 28 to 36 years; life-time follow-up is mandatory.[1] Ten-year survival is 50%.[1] Intensive staging and long-term follow-up including local treatment and systemic surveillance via blood counts or immunoglobulin measurements is essential for local control and development of multiple myeloma.[3,10]

Conclusion

In conclusion, extramedullary plasmacytoma of the nasal septum is a rare neoplasm that should be considered in the differential diagnosis of nasal masses. Surgical management is an acceptable treatment.

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