Maxillary sinus plasmacytoma

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Plasma cell neoplasms constitute a group of disorders characterized by monoclonal proliferation of plasma cells and the presence of monoclonal immunoglobulins (M-component) in the serum. Batsakis [1] classified PCNs occurring in the head and neck region as three disorders: extramedullary plasmacytoma, solitary plasmacytoma of bone, and manifestation of multiple myeloma. These disorders essentially represent distinct manifestations of a disease continuum. The clinical findings are critical to diagnosis, and distinguishing one disorder from the other has significant implications for treatment and survival.

Plasmacytomas represent a localized proliferation of plasma cells. Their symptoms, such as localized areas of bony pain, swelling or, rarely, soft tissue masses obstructing the upper respiratory tract or the oral cavity, are caused by the expanding plasma cell mass. We present a rare case of a maxillary sinus plasmacytoma.

PCN = plasma cell neoplasm

Patient Description

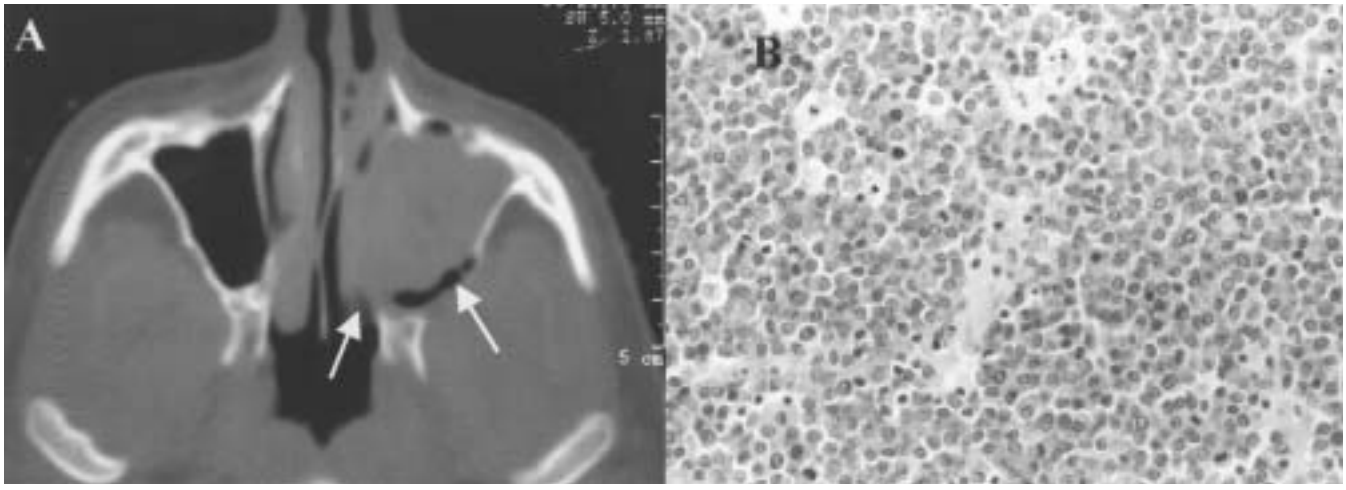
A 34 year old previously healthy patient presented at our institution with a 4 month history of nasal discharge, left nasal obstruction, pain in the left maxillary region, and difficulty opening his left eye. Before his admission he was treated with an oral antibiotic, an oral antihistamine and local nasal decongestant with no improvement. This treatment was initiated by his family physician after an X-ray film of the sinuses revealed opacification of the left maxillary sinus.

On physical examination, mild proptosis of the left eye was noted. The nasal septum was deviated to the right with bilateral enlargement of the inferior turbinates. The medial wall of the left maxillary sinus had expanded into the nasal cavity and was smooth. The rest of the physical examination was unremarkable.

A computerized tomography scan of the sinuses [Figure A] demonstrated a large mass within the left maxillary sinus with destruction of the medial and posterior walls of the sinus. The mass extended into the left nasal cavity and eroded the floor of the left orbit. Endoscopic rhinoscopy revealed a mass filling the left middle meatus and a biopsy was taken. Histopathologic examination showed fibrotic tissue infiltrated by plasma cells with varying degrees of maturity and atypia [Figure B]. These cells were stained by kappa light chain monoclonal antibodies.

A whole-body bone scan and systemic skeletal radiographic survey were normal. Bone marrow biopsy revealed a normocellular bone marrow with normal presentation of all three hematopoietic lines. Urinalysis results were negative for albumin and Bence-Jones protein. Immunoelectrophoresis did not show paraproteinemia. Blood β2-macroglobulin was normal. Based on the above, sinonasal extramedullary plasmacytoma was diagnosed.

Irradiation, 5,040 cGy, was administered to the left maxillary and nasal regions. The mass disappeared after completion of



[A] Bone window axial CT scan of the paranasal sinuses demonstrating a mass within the left maxillary sinus. The mass destroys the medial and posterior walls of the sinus (arrows) and extends into the left nasal cavity.

[B] Light microscopy photomicrograph of a maxillary plasmacytoma demonstrating plasma cells with varying degrees of maturity and atypia. (Hematoxylin & eosin, original magnification x 40).

therapy. The patient then underwent a left dacrocystorhinostomy due to narrowing of the nasolacrimal duct. Today, 4 years later, he is alive and with no evidence of recurrence.

Comment

Plasma cell neoplasms are uncommon in the head and neck region and occur most frequently in elderly patients. These malignant tumors can present as either a solitary tumor or part of a multifocal process. Localized forms of plasmacytoma include extramedullary plasmacytoma and solitary plasmacytoma of bone. Each disorder comprises about 3% of PCNs [2].

Extramedullary plasmacytomas comprise less than 1% of head and neck tumors [3]. These are soft tissue plasma cell tumors found in patients with no evidence of bone marrow disease and with no findings on total body skeletal survey. However, bone erosions adjacent to the plasmacytoma may occur. Men are affected 3 to 5 times more frequently than women and the median age at diagnosis is 55–60 years. Approximately 80% of head and neck EMPs arise submucosally, the most common site being the sinonasal region. Other sites are nasopharynx, salivary glands, thyroid gland, tonsils, cervical lymph nodes, larynx, and skin. The presenting symptoms and signs of EMPs are nonspecific and usually reflect the mass effect of the tumor. The most common findings are soft tissue mass or swelling, or airway obstruction. The presenting symptoms in the sinonasal region are nasal obstruction, nasal mass, nasal discharge, epistaxis, and facial pain mimicking recurrent sinusitis. Diffuse infiltration of neighboring structures such as orbit, hard or soft palate, skin, or skull base can also occur. Radiographic imaging is important to evaluate the location and size of the lesion. CT scan delineates bone destruction and magnetic resonance imaging defines the extension of the soft tissue mass. EMPs have a lower rate of conversion to disseminated MM compared with SBP. In 20–30% of EMPs, progression to MM may occur. Survival at 10 years is around 70% [2].

In contrast to EMP, SPB is a lytic bony lesion in a patient with otherwise normal findings on total body skeletal survey and with a normal bone marrow biopsy. It rarely involves the head and neck region and usually affects the long bones. The median age at diagnosis is 50 years and men and women are equally affected. SPB is considered to be an early manifestation of MM; 58% of SPBs progress to MM and survival rate at 10 years is only 16% [2].

It is important to distinguish MM from EMP or SPB because the treatment and prognosis are different. Evaluation for PCNs includes total body skeletal survey using plain radiographs, serum and urine electrophoresis or immunoelectrophoresis, complete blood count, calcium level and bone marrow biopsy. The diagnosis of EMP or SPB is made by three negative findings (no clinical, histologic or radiologic evidence of MM), in the presence of a single plasmacytoma in soft tissue or bone respectively.

Fine needle aspiration of a plasmacytoma may be non-diagnostic because of the limited tissue available for special staining and for complete histologic examination. Therefore, incisional or excisional biopsy, depending on the size and location of the mass, is necessary. Histologically, plasmacytomas are characterized by a diffuse or sheet-like proliferation of plasma cells with varying degrees of maturity and atypia. The nuclei are oval to round and eccentrically located, with a dispersed ("clock-face") nuclear chromatin pattern and a clear or halo area. Plasmacytomas may be confused with other benign and malignant conditions such as benign reactive plasmacytosis, undifferentiated carcinoma, non-Hodgkin's lymphoma, malignant melanoma, or esthesioneuroblastoma. Therefore, immunohistochemical staining assists in typing the neoplastic and the monoclonal nature of the cells.

PCNs are highly radiosensitive. Radiation therapy is currently the treatment of choice for EMPs and SPBs [4]. Plasmacytomas respond to radiation at doses of 4,500–6,000 cGy, delivered as daily fractions of 125–200 cGY 5 days a week. Wax et al. [5] demonstrated excellent locoregional control using radiation therapy as the primary modality of therapy. Surgical excision may be used in limited disease, such as in the parotid or thyroid glands or cervical lymph nodes, with radiation therapy reserved for treatment failures. However, while these tumors may present as aggressive locally destructive lesions, their management should be organ sparing because of the excellent control that can be achieved in the majority of cases by irradiation. Long-term follow-up by an otolaryngologist and a hematologist is necessary to identify patients whose disease converts to disseminated MM.

EMP = extramedullary plasmacytoma
MM = manifestation of multiple myeloma
SBP = solitary plasmacytoma of bone

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Women like silent men. They think they're listening.

Anonymous