Extramedullary Plasmacytoma Of The Nasal Cavity
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Citation

DOI: 10.5580/1827

Abstract
Extramedullary Plasmacytomas (EMP) are rare tumors accounting for 0.4% of all head and neck malignancies and 4% of all nonepithelial tumors of nasal cavity. The absence of multiple myeloma must be confirmed with bone marrow examination and skeletal surveys, before arriving at a diagnosis of EMP. We report a case of extramedullary plasmacytoma of the nasal cavity in a 60 year old female, who was a chronic tobacco sniffer. The case demonstrates the multidisciplinary approach required for the optimal diagnosis and management of such tumors and proposes chronic irritation of the nasal mucosa as one of the etiological factors for EMP.

INTRODUCTION
Extramedullary Plasmacytomas (EMP) are rare tumors accounting for 0.4% of all head and neck malignancies and 4% of all nonepithelial tumors of nasal cavity. About 80% of the extramedullary plasmacytomas are localized in the submucosa of the upper respiratory tract, with a large proportion occurring in the sinonasal/nasopharyngeal area. The absence of multiple myeloma must be confirmed with bone marrow examination and skeletal surveys, before arriving at a diagnosis of EMP. The therapy consists of combined surgery and radiotherapy. The main prognostic indicator for EMP is progression to multiple myeloma. 10-32% of all patients develop multiple myeloma which reduces the mean survival time from 8.3 years to 20 months. We report a case of EMP of the nasal cavity in a 60 year old female who was a chronic tobacco sniffer. The patient was managed by endoscopic removal of the tumor followed by radiotherapy. A 6 month follow up of the patient revealed neither recurrence of the tumor nor a progression to multiple myeloma.

CASE REPORT
A 60 year old female, with unremarkable past medical history, presented with right sided nasal obstruction, mucoid nasal discharge and intermittent epistaxis for the past 5 months. There was history of occasional headaches. The patient had been sniffing tobacco for the past 35 years. The general physical examination did not reveal any abnormality, nor was there any evidence of lymphadenopathy. Anterior rhinoscopy revealed a pinkish, fleshy mass in the right nasal cavity which was firm in consistency, bled easily and was insensitive to probing. Endoscopic examination confirmed the presence of a dark red sub mucosal lesion with a smooth surface. The CT scan of the patient revealed a hypodense mass localized to the right nasal cavity showing contrast enhancement with no bony erosion (figure 1). A nasal biopsy was subsequently taken which revealed a dense infiltrate of plasma cells (figure 2). A histological diagnosis of plasmacytoma was made. Routine blood tests and serum biochemistry including calcium, phosphorus, blood urea nitrogen, urea, uric acid and creatinine were all within reference range. Both serum myeloma proteins and urine Bence- Jones proteins were negative. Serum and urine electrophoresis did not reveal M component. Bone marrow biopsy revealed a plasma cell infiltrate of less than 3% of all nucleated cells. Chest radiograph, total body skeletal survey and Tc99 scintigram, showed no systemic lesion. The finding confirmed a diagnosis of stage 1 extramedullary plasmacytoma. The patient underwent complete surgical removal of the tumor endoscopically. This was followed by radiotherapy of 5000 cGy, delivered over a period of 6 weeks. A 6 month follow- up of the patient did not reveal any recurrence.
Extramedullary Plasmacytoma Of The Nasal Cavity

Figure 1
FIGURE 1: CONTRAST ENHANCED CORONAL CT SCAN PNS SHOWING HYPODENSE MASS IN THE RIGHT NASAL CAVITY.

Extramedullary Plasmacytoma is a localized collection of monoclonal plasma cells, arising within soft tissues in an extraskeletal site. Extramedullary Plasmacytomases can either be – Primary (true) Plasmacytoma (without evidence of systemic disease) or Extramedullary manifestation during the course of multiple myeloma.

The first case of EMP was reported by Schridde in 1905. Alexiou et al in 1999, reviewed all previous reports of EMP and found 869 cases out of which 714 (82.2%) had occurred in the upper aero digestive tract. The most commonly affected area in this region were the nasal cavity/Para nasal sinuses (43.8%) followed by nasopharynx (18.3%), the oropharynx (17.8%) and larynx (11.1%). EMP affects males 3-4 times more than females with a median age of 55 years (range 50-65 years) which is 10 years younger than multiple myeloma. Most of the EMP’s are solitary lesions, however 10-20% manifest as multiple lesions. The etiology of EMP is still unknown. Because of its presentation in the mucosa of the upper aero digestive tract (78%), chronic stimulation by inhaled irritants or viral infection has been implicated as causative factors. The clinical presentation varies according to the site of involvement. As most lesions are located in the sinonasal/nasopharyngeal area, the presenting symptoms include soft tissue mass or swelling (80%), airway obstruction (35%), epistaxis (35%), pain (20%), proptosis (15%), nasal discharge (10%), regional lymphadenopathy (10%), or cranial nerve palsy (5%) with the median duration of symptoms being 4-5 months. The physical examination essentially reveals a gray to red, soft or firm, sessile or pedunculated, submucosal mass that bleeds easily and is usually nontender and smooth without obvious ulceration of the mucosa. Osseous destruction is uncommon in extramedullary plasmacytomas.

Histopathologically, the tumor is characterized by a dense homogenous infiltrate of plasma cells with varying degree of dysplasia. The plasma cells have round eccentric nuclei with dense chromatin clumps arranged along the nuclear membrane in a cartwheel fashion. Plasmacytic, plasmablastic and anaplastic cell types have been described. Local amyloid deposits are found in 11-38% of cases; systemic amyloid being very rare. No amyloid was detected in our patient. Histopathology cannot distinguish multiple myeloma from extramedullary plasmacytoma. Confirmation of diagnosis can only be made after any systemic

DISCUSSION
Plasma cell neoplasm are a group of clinical disorders characterized by monoclonal expansion of plasma cells that elaborate a single homogenous immunoglobulin molecule or fragment. Depending on their site of development and clinical features, plasma cell neoplasm are divided into: a) Solitary extramedullary plasmacytoma (b) Solitary bone plasmacytoma (c) Multiple myeloma (d) Multifocal form of multiple myeloma (e) Plasmablastic sarcoma.

Figure 2
FIGURE 2: MICROPIC'TOGRAPH SHOWING SHEETS OF CLOSELY PACKED PLASMA CELLS
involved has been excluded. A complete blood count
with WBC count and platelet count, serum biochemistry
including calcium, BUN, creatinine, uric acid, serum
proteins, serum and urine electrophoresis, bone marrow
biopsy and a complete skeletal survey are recommended to
rule out multiple myeloma.10

The diagnostic criterion for EMP include: (a) Biopsy of
tissue showing monoclonal plasma cell histology. (b) Bone
marrow plasma cell infiltrates not exceeding 5% of all
nucleated cells. (c) Absence of osteolytic bone lesions or
other tissue involvement (no evidence of myeloma
elsewhere). (d) Absence of hypercalcemia or renal failure.
(e) Low serum M protein concentration, if present.11

EMP can be staged as follows: Stage 1-Limited to
extramedullary site. Stage 2: Local extension or involvement
of regional lymph nodes. Stage 3: Disseminated disease.2

According to Batsakis, the natural history of extramedullary
plasmacytoma may be characterized as: (a) localized,
solitary controlled by surgery, radiotherapy, or both, does
not recur or become disseminated.(b) locally recurrent
controlled by additional therapy.(c)aggressive, persistent or
recurrent disease, producing death through uncontrollable
local extensions.(d) local disease with “metastatic”
involution of regional lymph nodes without evidence of
distant spread.(e) local disease, recurrent or otherwise
followed by dissemination and development of multiple
plasma cell neoplasm and/or multiple myeloma.12

The treatment of Extramedullary Plasmacytoma is still
controversial. Some clinicians favour radiotherapy2 , others
prefer surgical excision, while some advocate combined
approach.13 .Alexiou et al in their review of 714 cases of
upper aero digestive tract extramedullary plasmacytomas
reported between 1905-1997,found that the median overall
survival or recurrence free survival was longer than 300
months for patients who underwent combined surgery and
radiotherapy as compared to a median survival rate of 144
months for patients who underwent radiotherapy and 156
months for surgically managed patients.3 Most clinicians
recommend a combined approach (surgery and radiotherapy
) for the management of upper aero digestive EMP.2,13 . The
optimal dose for local control is 5000 cGy, with a fraction
size of 200 cGy delivered over 4-6 weeks.5 The patient in
our case had localized disease of the nasal cavity which was
resected surgically and subsequently subjected to
radiotherapy.

The median survival of patients varies from 4-10 years.
Local recurrences have been reported in 22% cases of
adequately treated EMP in the upper aero digestive tract
with 16% of EMP evolving subsequently into multiple
myeloma.5 Because of the tendency of EMP to progress into
disseminated multiple myeloma, a lifelong follow-up of
these patients is recommended.

Our case clearly demonstrates that a multidisciplinary
approach is required for the optimal diagnosis and
management of extramedullary plasmacytoma. It is essential
to exclude any systemic involvement before arriving at a
diagnosis of solitary extramedullary plasmacytoma. The
present case being a chronic tobacco sniffer, further supports
Witshaw’s view that chronic stimulation of the mucosa of
the upper aero digestive tract by inhaled irritants may be one
of the etiological factors for EMP.2

References
1. Fu YS, Perzin KH. Non epithelial tumors of nasal cavity, paranasal sinuses and nasopharynx. Cancer 1978; 42:
2399-2406.
2. Wiltshaw E. The natural history of extramedullary plasmacytoma and its relation to solitary myeloma of bone
:47-51.
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