CASE REPORT

Extramedullary Plasmacytoma of Maxillary Antrum

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Abstarct

Extramedullary plasmacytoma (EMP) represents approximately 3% of all plasma cell neoplasms.With an idea to heighlight its diagnosis,treatment modalties, we report a case of extramedullary plasmacytoma in a 52 years old male patient.

Key Words

EMP, Maxilary antrum, malignancy

Introduction

Extramedullary plasmacytoma (EMP) represents approximately 3% of all plasma cell neoplasms. More than 80% EMP arise in upper aerodigestive tract, mostly in nose, sinuses, nasopharynx and oral mucosa. This is probably related to long-term stimulation by inhaled irritants or viral infection. EMP occur approximately three times more often in men than in women, usually seen in age group 50-70 years (1). Cases of primary growth of EMP in salivary glands, the orbit, the lacrimal glands, the trachea, the thyroid gland and the larynx have been described (2). Regional lymphnodes are invaded in less than 10% of EMP and 16% of patients progress to multiple myeloma. The patients usually present with localized bone pain, but presentation may be with painless swelling or pressure effect (3). These tumors are usually sessile in the nose and nasopharynx but pedunculated in the larynx and pharynx. These are firm in consistency in larynx and pharynx but soft in maxillary antrum (3). The color varies from yellowish grey to dark red and are generally smooth without ulceration (4). With an idea to heighlight dignosis and treatment modalties, in this presentation, we report a case of extramedullary plasmacytoma in a 52-year old Muslim male.

Case Report

A 52 year old male presented with progressive swelling of the left cheek and left nasal obstruction for the last 8 months with occasional pain and epistaxis on the same side.The swelling was diffused on left side of cheek, approximately 5 cm^2 in size, firm, non-tender with free overlying skin. The nasolabial furrow was obliterated. The left upper gingivolabial sulcus was swollen. The patient was having proptosis on the left side associated with epiphora. No lymphadenopathy was detected clinically.On anterior rhinoscopy, no growth was seen on left side and the cavity was grossly reduced as the lateral wall of the nose was pushed medially. Posterior rhinoscopy did not reveal any abnormality.Right nasal cavity was normal on examination. The patient was not having any trismus at the time of admission but he developed it in the hospital in due course of investigations. Examination of oropharynx and palate was normal.

The remainder of the ENT examination was unremarkable with absence of any neck nodes. His chest X-ray, full blood count, liver function tets, urea and electrolytes were within normal limits. Bence-Jones proteins were not present on urinalysis and the bone marrow examination detected no abnormality.CT scan *(Fig-I a & b)* with enhancement showed an expanding mass in left maxillary antrum with central necrosis. Superiorly, the mass was extending into the left orbital fat planes, inferiorly into the infra-temporal fat planes and medially extending into the left nasal cavity with destruction of all the bony walls of the maxillary antrum and the left nasal turbinates. Other sinuses were normal. Biopsy of the lesion confirmed the diagnosis as plasmacytoma. Taking ito consideration the patient's willingness who insisted for surgery because of tumor mass and destructive pathology of the tumor, the patient

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was taken for surgery and total maxillectomy was performed. During surgery, the mass was friable with extensive bleeding. This time also the histopathology report came as plasmacytoma *(Fig-2 a & b)* showing sheets of immature plasma cells with eccentric nucleus and increased nucleus-cytoplasm ratio.

A diagnosis of extramedullary plasmacytoma of left maxillary antrum was made and the patient was operated with the idea of debulking the tumor, although the tumor is very much radio sensitive. The second reason to operate was that patient himself insisted for surgery despite being explained the treatment protocols.Treated with post-operative radical radiotherapy as the tumor is radiosensitive and there were chances of non-clearance of the tumor tissues in toto due to massive bleeding. The trismus of the patient improved and the patient was doing well upto 6 months in the postoperative follow-up.

Discussion

Neoplasms of the plasma cell region have been classified as solitary plasmacytomas, multiple or diffuse myeloma and plasma leukemia. Solitary plasmacytoma are further characterised as intramedullary and extramedullary. The extramedullary plasmacytoma is a malignant plasma cell dyscrasia which arises from soft tissues. This is a rare tumor which can be seen in any organ (2). Extramedullary plasmacytomas arise primarily in the head and neck region and more than 80% of EMPs arise in upper aerodigestive tract, mostly in nose, sinuses, nasopharynx and oral mucosa (3). Outside the head and neck area, the EMP is very uncommon but primary tumors in the lung, pleura, stomach, small bowel, colon, ovary, uterus, testes, kidneys, skin and breast have been reported (5).

Classification of individual cases should therefore be tampered by time, as it is possible for apparent solitary EMP to be the first presenting sign of a generalized disease. The treatment and prognosis of primary EMP are different from that of extraskeletal spreads in MM. It is, therefore, important to look for systemic skeletal involvement in all patient with EMP prior to the institution of therapy (2).The diagnosis of EMP can be made only on morphologic examination of the tumor. In some instances, it may be difficult to determine whether the plasma cell proliferation is reactive or neoplastic. EMP should be distinguished from non-neoplastic conditions such as reactive plasmacytic hyperplasia, plasma cell granuloma (PCG), and pseudo-lymphoma (PL). The malignancies those can be mistaken for EMP include

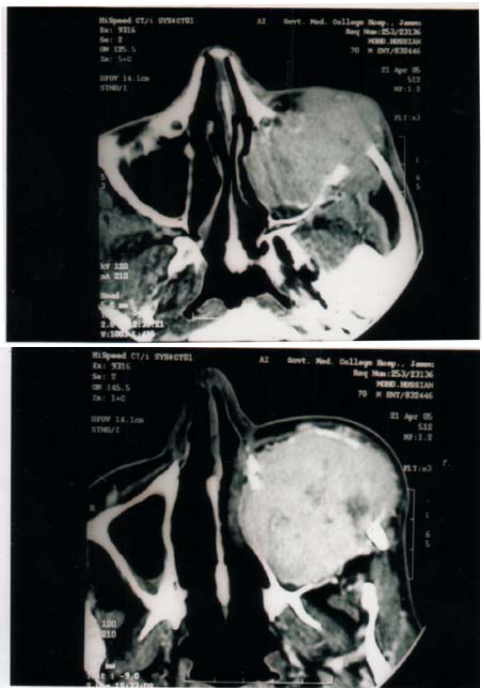
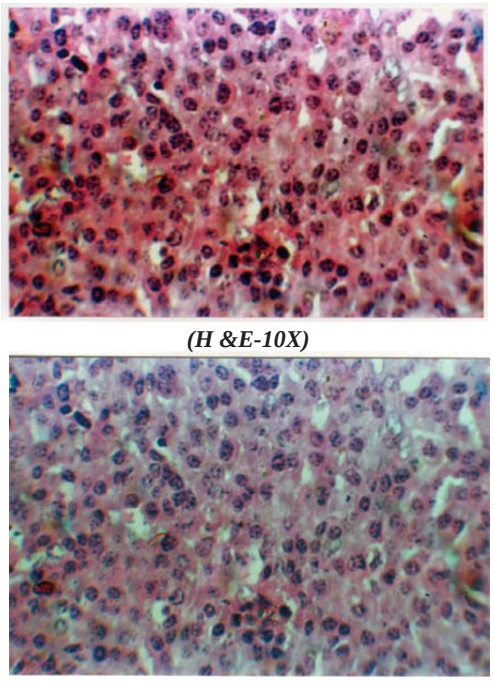


Fig 1 a & b: CT Scan Showing Mass in Left Maxillary Antrum With Central Necrosis & Extension into Left Orbital Fat Planes, Left Infratemporal Fossa & Left Nasal Cavity With Destruction of All Bony Walls of Maxillary Antrum & Left-Sided Turbinates



(H &E-10X)

(H &E-40X)

Fig 2 a & b:Photomicrograph Showing of Mature Plasma Cells and Occasional Binucleate Plasma Cells

other haematopoietic neoplasms, malignant melanoma, olfactory neuroblastoma, anaplastic carcinoma and metastases (2). On histological examination, closely packed masses of immature or atypical plasma cells of varying differentiation and a scant stroma are seen with replacement of normal tissue. The cytoplasm of plasma cells is strongly pyroninophilic. A characteristic concentrically arranged lamellar rough endoplasmic reticulum and prominent Golgi apparatus are seen on electromicroscopy. Identification of a single type of Ig using the immunoperoxidase method is considered evidence of the neoplastic nature of the plasma cell proliferation. The cells have cart-wheel nucleus with basophilic cytoplasm and occasionally multi-nucleated gaint cells are also seen (6).

Laboratory tests recommended by Kyle 1997 for diagnosing EMP are (3) :-Complete blood cell count, Serum calcium and creatinine, Serum electrophoresis / immunofixation / quantitative serum immunoglobulins, Electrophoresis of aliquot from 24 hours urine specimen followed by immunoelectrophoresis, b2 microglobulin, CRP and LDH estimation, Skeletal survey including humerus and femur, Bone marrow aspiration and biopsy and the plasma cell labelling index, if available, Peripheral blood plasma cell count and labelling index, CT and MRI (optional).Extramedullary plasmacytoma is associated with 'M' proteins in less than 30% of cases and usually disappear after radiotherapy (3). Monoclonal Ig is found in 97% of patients with MM, when immunoelectrophoresis is performed on serum and urine and an abnormal skeletal survey in 7% of cases (2). In our patient, we could reach the diagnosis only on CT scan and biopsy. None of the other tests were showing any abnormal findings pointing towards the diagnosis of plasmacytoma.

The radiographic manifestations of EMP depend upon the size, location and extent of the tumor. When confined to submucosa/ mucosa, it is visible as a soft tissue mass which may block sinus orifices. Multiple sites of simultaneous tumor development may be apparent. Calcification, within primary tumors has not been observed and metastatic deposits outside the liver, spleen and lymphnodes have not been reported (2). In our patient, which is presented here, the microscopic and radiographic evaluation was consistent with other authors.

EMP are radiosensitive and radiotherapy (RT) to the local lesion is the mainstay of treatment. The majority of mucosal and small EMP can be controlled by 3000 and 4000 rads. For large EMP, one should attempt to irradicate the local tumor and final radiation dose should be determined by the size of the tumor, the extension to the involved bone, muscle and other deep structrues and the completeness of the regression following a certain dose of radiation. Some authors have recommended a uniform dose of upto 5000-6000 rads in the treatment of these tumors. Radical surgery is not usually indicated. Treatment with chemotherapy and steroids is indicated in paients with secondary EMP and when dissemination occurs in the course of primary EMP (2). In our ptient, the surgery was done to debulk the tumor mass and also the patient was very particular for the surgery. Radiotherapy was given postoperatively.

The prognosis is more favourable in patients with primary EMP not associated with MM. Although a small proportion of these patients have a rapidly progressive course with dissemination and death in a few months, about 33-75% survive five years. Dissemination eventually occurs in 5-50% of patients with primary EMP months to years later, at times as late as several decades after diagnosis (7). Bone pain, anemia, renal impairment, bronchopneumonia and other infections may herald the onset or complicate the course of MM. Once MM occurs, infection and renal failure are the commonest cause of death (2).

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