Extramedullary Plasmacytoma of the Right Maxillary Sinus- A Case Report

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Introduction

Plasmacytoma's are localized neoplastic proliferations of monoclonal plasma cells. The disease is divided into primary plasmacytoma of the bone and extramedullary plasmacytomas. These are rare, soft tissue tumors arising from malignant proliferations of plasma cells[1]

They primarily occur in the head and neck region usually presenting as soft tissue masses in the nasopharynx and paranasal sinuses[2]

In view of the rarity of the tumor, a case of extramedullary plasmacytoma of the right maxillary sinus is presented, clinical features described and therapeutic options discussed.

Case Report

The patient was a 37 year old Nigerian female who presented with a two year history of persistent nasal obstruction, headache and nasal discharge from the right nasal cavity.

A diagnosis of chronic sinusitis had been made which was suggested by radiographic examination of the paranasal sinuses (figure1) and she was being managed with antibiotics and had 2 episodes of antral washout done. She later presented with a two week history of an ulcerated mass around the nasal region which was associated with a foul smelling discharge three months later.

On examination, minimal patency was found in both nasal cavities worse on right with foul smelling necrotic tissue in the right nasal cavity. There was a dark colored moderate swelling over the right alar region (figure 2) and on probing a defect was found communicating with the right nasal fossa. A nasal clearance biopsy was done and while awaiting histological diagnosis for definitive management, fifth day post operative period patient developed symptoms of peritonitis and aspiration pneumonitis and succumbed to illness while undergoing treatment

Histological examination of the biopsies subsequently confirmed an extramedullary plasmacytoma [EMP].(figure 3)

Discussion

Extramedullary plasmacytoma is an uncommon malignant neoplasm arising outside the bone marrow in patients without clinical evidence of multiple myeloma, with a predilection for the head and neck region[3]. Nearly 80% can be found in the upper respiratory tract or oral cavity[4]. Representing up to 4% of non-epithelial lesions of this region.[5]

Diagnosis from a clinical point of view can

only be suspected and is ultimately made by histological examination.

The tumor arises in the submucosa of the upper respiratory tract or oral cavity. It grows at a variable rate as a polypoid mass or an invasive tumor expanding and destroying any structures in its path, and eventually appearing as a grayish, pink or dark red to purple lobulated mass, varying in consistency in different parts of the tumor from firm to soft and friable. The surface which is lined by intact epithelium or may occasionally be ulcerated, bleeds easily and sometimes profusely on palpation or taking a biopsy.

Small localized lesions may be sessile or pedunculated so as to suggest easy operability. Long standing ones are large and may produce hideous distortions of the features and vast destruction of bony structures.[6]

The symptoms depend on the stage at which the patient presents for treatment, but a common feature is epistaxis[6]. There may be obstruction of the nasal, pharyngeal or laryngeal passages. Pain is usually absent unless there are complications due to infection or pressure on nerves. Other symptoms will depend on the direction of spread into the ethmoids, orbit or cranial cavity[7]

In cases where there is no previous history of plasma cell malignancy, it is important to exclude disseminated disease in the form of multiple myeloma; this should include a radiographic survey of the skeleton and bone marrow biopsy.

As earlier mentioned, diagnosis is made on examination of pathological specimens with immunohistochemical staining techniques. The histological appearance of EMP consists of a monoclonal proliferation of plasma cells set in a very sparse matrix. Cellular and nuclear atypia may be minimal or prominent[8].

Plasmacytomas are very radiosensitive and radiotherapy is the main treatment modality. Surgery can be used effectively for small lesions, or as a salvage procedure. Chemotherapy is reserved for locally advanced recurrent or dissemination disease. Matinson and Pulveitaff noted in their work done on clinical and live cell study of EMP of the upper respiratory tract that in countries like Nigeria where Radiotherapy is not readily available, chemotherapy is of particular interest especially as most cases present for treatment when the lesions is beyond surgical intervention alone. As the progress of the disease is unpredictable, it would be rash to claim full success for any form of treatment, especially in advanced stages of the disease, but the response to chemotherapy in a small group seen by them was encouraging enough to suggest that this form of treatment may be if developed satisfactorily, a useful adjunct to radiotherapy. It is hoped also that multiple lesions will respond to it where total body irradiation may be considered dangerous, even if it were available[9].

Prognosis is fair, as it is a low grade tumour, 5 year survival is usually most favourable amongst the younger age group(less than 60 years) and poorest amongst the older age group(60+ years) decreasing from 76.6% to 53.3% [10]

Conclusion

A rare tumour of the head and neck region, extramedullary plasmacytoma has been presented occurring in the right maxillary antrum with symptoms suggestive of maxillary sinusitis, in view of the urgent need to make a correct diagnosis at the earliest possible time and institute proper management in these patients its clinical features have been highlighted and treatment

modalities discussed.

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