Extramedullary plasmacytoma in the maxillary sinus

Megat Shiraz M A R, Jong Y H, Primuharsa Putra S H A

ABSTRACT

Extramedullary plasmacytoma is a rare malignant plasma cell tumour. We report an extremely aggressive case of extramedullary plasmacytoma of the right maxillary sinus, which had metastasised to the brain and rib. A 56-year-old man presented with recurrent epistaxis and acute anaemia. Nasendoscopy revealed a medialised medial wall of the right maxilla and a mass occupying the whole nasopharynx. Magnetic resonance imaging revealed a right maxillary tumour with extension to the ipsilateral nasal cavity, nasopharynx, right sphenoid and ethmoidal sinuses. There was an extra-axial brain metastasis. There were metastases to the right parietal region and left eighth rib. Histopathology examination of the maxillary mass revealed abundant plasma cells with kappa-chain restriction. He was planned for four cycles of chemotherapy. Unfortunately, in view of the advanced stage of disease, he succumbed to his disease during the first cycle of chemotherapy.

Keywords: extramedullary plasmacytoma, maxillary sinus, plasma cell tumour, sinus neoplasm

Singapore Med J 2008;49(11):e310-e311

INTRODUCTION

Extramedullary plasmacytoma is a localised collection of monoclonal plasma cells in an extraskeletal site. The tumours arise predominantly in the submucosal tissues of the nasal cavity, paranasal sinus, nasopharynx, oropharynx and larynx. The incidence in males is three times that of females. The most affected age group is 50–70 years. It is usually solitary or localised, but 10% of the patient have multiple site involvement. We report a patient who had extramedullary plasmacytoma of the right maxillary sinus with metastases to the brain and rib.

CASE REPORT

A 56-year-old male retired police officer complained of recurrent right-sided epistaxis for three months, associated with bleeding from the mouth. The epistaxis stopped with external and cold compression at home after an hour. His last episode was two months ago, when he also presented with symptoms of anaemia. He also had right nasal blockage with mild facial swelling which was associated with dull pain. He also had odynophagia and progressive dysphagia to solid food. He experienced a loss of appetite and loss of weight, approximately 5 kg in the past two months. In addition, he had a right parietal swelling for the past two years. It was slow-growing and associated with an occasional throbbing headache.

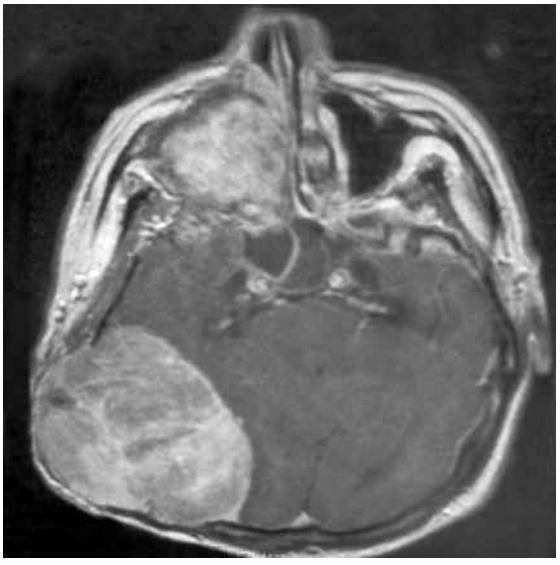

Fig. 1 Contrast-enhanced axial T1-W MR image shows a right maxillary sinus tumour with a large extra-axial brain metastasis.

Physical examination revealed a pale and cachexic man with tachycardia. A diagnostic nasoendoscopy revealed a medialised medial wall of the right maxilla and a mass occupying the whole nasopharynx with blood clots. Intraorally, the right hard palate was pushed inferiorly but the mucosa remained intact. There was a palpable firm and fixed mass in the right parietal region, measuring 7 cm × 6 cm. Blood investigations revealed severe anaemia with hypoalbuminaemia. He was resuscitated with colloid solution and 2 pints of packed cells. Computed tomography showed a right maxillary mass, and a right temporal-parietal mass eroding the skull to scalp. Magnetic resonance (MR) imaging (Fig. 1) showed a right maxillary sinus tumour with extension to the ipsilateral nasal cavity, nasopharynx, right sphenoid and ethmoidal sinuses. The proximal portion of the right optic nerve at the orbital apex appeared to be encased. There was extra-axial brain metastasis. Bone scintiscan showed heterogeneous uptake involving the right parietal bone associated with an overlying increased soft tissue uptake, right maxilla and left eighth rib.

Examination under anaesthesia revealed a friable and bloody tumour mass at the right middle meatus, arising

Department of Otorhinolaryngology, Head & Neck Surgery, Faculty of Medicine, Hospital Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Cheras, Kuala Lumpur 56000, Malaysia

Megat Shiraz MAR, MD, MSurg
Specialist

Jong YH, MD
Registrar

Ear, Nose, Throat - Head & Neck Consultant Clinic, Seremban Specialist Hospital, Jalan Toman 1, Kemayan Square, Seremban 70200, Malaysia

Primuharsa Putra SHA, MD, MSurg
Consultant

Correspondence to:
Dr Doris Evelyn Jong Yah Hui,
Tel: (60) 3 9170 2415
Fax: (60) 3 9173 7840
Email: evejong76@yahoo.com

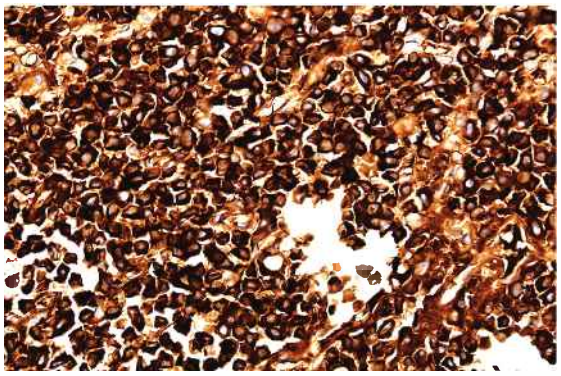

Fig. 2 Immunohistochemical studies with kappa-lambda antibody stain showed immunopositivity towards kappa light chain restriction (× 200).

from the antrum, and extending posteriorly to the postnasal space and left posterior choana. Biopsy was taken for histopathological examination. The histopathological examination revealed the presence of plasma cells with kappa light chain restriction (Fig. 2) and immunopositivity towards LCA. The impression of plasma cell tumour was made. The clinical features and histopathological findings were consistent with extramedullary plasmacytoma stage III. Bone marrow examination was normal. In view of the extensive disease, he was planned for chemotherapy. However, he succumbed to the disease during the first cycle of chemotherapy.

DISCUSSION

Extramedullary plasmacytoma arising from the maxillary sinus with metastasis is extremely rare. It is more common to encounter squamous cell carcinoma. Therefore, our initial provisional diagnosis was maxillary carcinoma with metastasis. However, the histopathology revealed extramedullary plasmacytoma, a rare entity. Extramedullary plasmacytoma is a localised collection of monoclonal plasma cells located in an extraskeletal site. The median age is 50–70 years with a male:female ratio of 3:1. Korolkowa et al reported that 40% occur in the nasal cavity and paranasal sinus, 20% in the nasopharynx, and 18% in the oropharynx. Approximately 10% of extramedullary plasmacytomas have multiple sites of involvement.[1] Murthy et al reported a case of solitary extramedullary plasmacytoma of the premaxilla which was treated with surgical and postoperative radiotherapy.[2] Webb et al reported a case of extramedullary plasmacytoma of the tongue base.[3] Windfuhr and Ott reported a case of extramedullary plasmacytoma in the nasal cavity in a 60-year-old man who was managed by interdisciplinary teams for optimal therapy.[4]

The clinical presentations of extramedullary plasmacytoma and carcinoma of maxillary sinus are almost similar. The diagnosis of plasmacytoma is based on and confirmed with histology and immunohistochemistry. There is dense, homogeneous infiltrate of plasma cells. Amyloid deposition may be seen in 15%–38% of extramedullary plasmacytoma. The commonest immunoglobulin expressed by the tumour cells is IgG with kappa chain restriction. Plasmacytoma can be graded low (grade 1), intermediate (grade 2) and high grades (grade 3), based on the cellular atypia. Based on the serum, urine electrophoresis, bone scan, bone marrow examination and radiological assessment, extramedullary plasmacytoma can be staged according to the spread of the disease. Stage I is disease confined to one site. Stage II includes tumours with local extension of lymph node involvement. Stage III has metastatic spread. Therefore, this patient was diagnosed to have extramedullary plasmacytoma with the brain and bone metastasis, i.e. stage III. There was no evidence of infiltration to the bone marrow or multiple myeloma. The reported conversion rate of extramedullary plasmacytoma to multiple myeloma is 15%–20%, and is associated with a poorer prognosis. Dissemination of the tumour takes place in 35%–50% of extramedullary plasmacytomas.[5]

Localised extramedullary plasmacytoma is highly radiosensitive. According to the guidelines published in British Journal of Haematology,[6] the recommended primary treatment for localised extramedullary plasmacytoma is radical radiotherapy. For generalised extramedullary plasmacytoma, chemotherapy is advisable. Therefore, chemotherapy was the treatment of choice in this patient who had extramedullary plasmacytoma with multiple metastasis. However, due the age, poor haemodynamic status, poor nutrition and extensive disease as well as the intolerance to side effects of chemotherapy, the patient succumbed to the disease during the early stage of treatment. Even though extramedullary plasmacytoma of the maxillary sinus is rare, it should be one of the differential diagnosis for maxillary tumours. The cooperation among the otorhinolaryngolists, pathologists and haematologists are required to manage extramedullary plasmacytoma patients effectively in order to provide optimal treatment.

REFERENCES

1. Korolkowa O, Osuch-Wójcikiewicz E, Depta a A, Suleiman W. [Extramedullary plasmacytoma of the head and neck]. Otolaryngol Pol 2004; 58:1009-12. Polish.
2. Murthy V, Agarwal JP, Jalali R, Chinoy RF. Solitary extramedullary plasmacytoma of the premaxilla. Australas Radiol 2003; 47:202-4.
3. Webb CJ, Makura ZGG, Jackson SR, Helliwell T. Primary extramedullary plasmacytoma of the tongue base. Case report and review of the literature. ORL J Otorhinolaryngol Relat Spec 2002; 64:278-80.
4. Windfuhr JP, Ott G. Extramedullary plasmacytoma manifesting as a palpable mass in the nasal cavity. Ear, Nose Throat J 2002; 81:110-4.
5. Wanebo H, Geller W, Gerold F. Extramedullary plasmacytoma of the upper respiratory tract. Recurrence after latency of thirty-six years. N Y State J Med 1966; 66:1110-3.
6. Soutar R, Lucraft H, Jackson G, et al. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. Br J Haematol 2004; 124:717-26.