Recurrent Extramedullary Plasmacytoma of the Nasopharynx: A Case Report and Review of the Literature

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Citation

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Abstract

Introduction: Extramedullary Plasmacytoma (EMP) is a rare neoplasm belonging to the category of non - Hodgkin Lymphomas. EMP makes up approximately 4% of plasma cell tumours which usually arise in the soft tissues of the upper airways. The incidence of neck involvement in EMP of the upper aerodigestive tract is 10-20%

Case Report:
The authors highlight the clinical presentation and management of a 59 year old patient with recurrent extramedullary plasmacytoma of the nasopharynx. The patient remains free of disease 2 years post treatment. A review of the relevant literature to date is also presented.

INTRODUCTION

Extramedullary Plasmacytoma (EMP) is a rare neoplasm belonging to the category of non â Hodgkin Lymphomas. EMP makes up approximately 4% of plasma cell tumours which usually arise in the soft tissues of the upper airways. [1]

A neoplastic proliferation of plasma cells was first described by Dalrymple and Bence Jones in 1846. They described a disseminated neoplastic proliferation of plasma cells that was characterised by marked proteinuria and bone pain.[2] Currently, plasma cell tumours are classified into one of three categories; multiple myeloma, medullary plasmacytoma and extramedullary plasmacytoma. [3]

80 % of these tumours originate in the head and neck region. EMP mainly occurs between the fourth and seventh decades of life. Alexiou et al reviewed all cases of extramedullary plasmacytoma quoted in the literature between 1905 and 1997. 82.2% were found in the upper aerodigestive tract. [1] The incidence of neck involvement in EMP of the upper aerodigestive tract is 10-20 %. [4] In the absence of known or suspected risk factors the pathogenesis of plasma cell tumours remains enigmatic. Solitary extramedullary plasmacytoma involving the nasopharynx is very rare. [5] We report a case of EMP arising in the Nasopharynx.

CASE REPORT

A 59 year old male engineer presented to the head and neck clinic with symptoms of throat discomfort, sense of a lump in the throat and bleeding per mouth. He was otherwise fit and well and had stopped smoking 30 years ago. Nasendoscopy revealed prominent vessels over the soft palate and an irregular ulcerated lesion on the posterior surface of the uvula. He underwent an urgent examination of the Nose and postnasal space under general anaesthesia. Multiple biopsies revealed an extramedullary plasmacytoma of the dorsal surface of the soft palate. The histology showed a dense infiltrate of plasma cells which show nuclear atypia. Fungal stains were negative (Figures 1 & 2). Light chain staining with vs38c antibody suggested a plasmacytoma. (Figure 3) There was no clinical or radiological evidence of neck disease. Subsequent bone marrow biopsies were clear of tumour. The patient underwent radical radiotherapy treatment of the nasopharyngeal field with a dose of 45Gy in 20 fractions.

Figure 1

Figure 1: Low power view of lesion demonstrating plasma cells

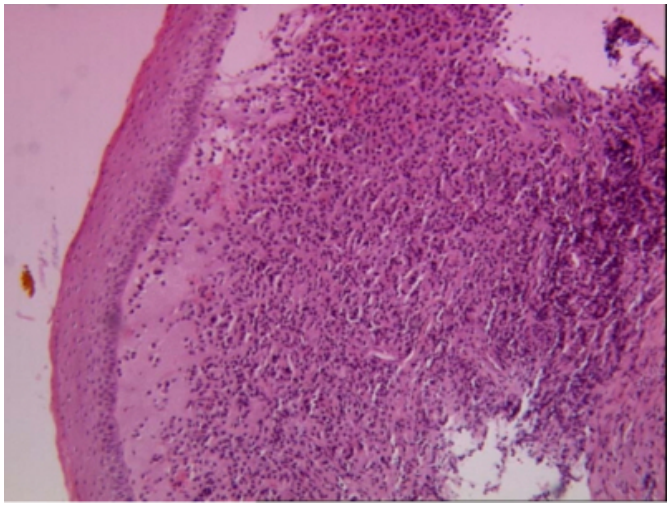


Figure 2

Figure 2: High power view demonstrating typical plasma cell cytoplasm and clock face chromatin. Some pleomorphic atypical forms also present

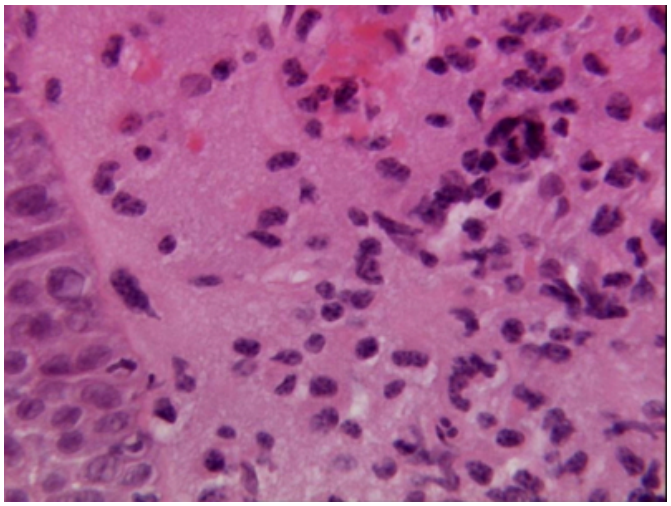
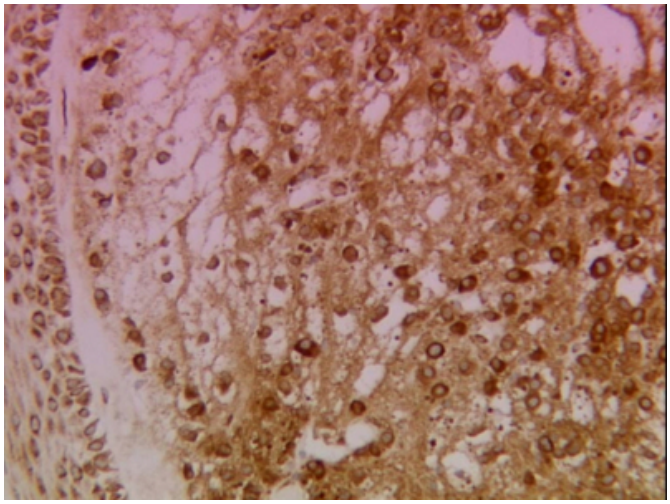


Figure 3

Figure 3: Immune-histochemistry demonstrating staining of plasma cells with vs38c antibody



The patient subsequently developed a recurrence of the extramedullary plasmacytoma in the left side of the nasopharynx four months following radiotherapy. He underwent KTP ablation of the post nasal space. A further examination under anaesthetics of the nasopharynx revealed bony sequestra which was negative for tumour tissue. (Figures 4& 5) The patient is now 2 years post laser surgery. His serum paraproteins and repeat MRI scans show no evidence of recurrence during this period.

Figure 4

Figure 4: Bony sequestra protruding through the posterior nasopharyngeal wall

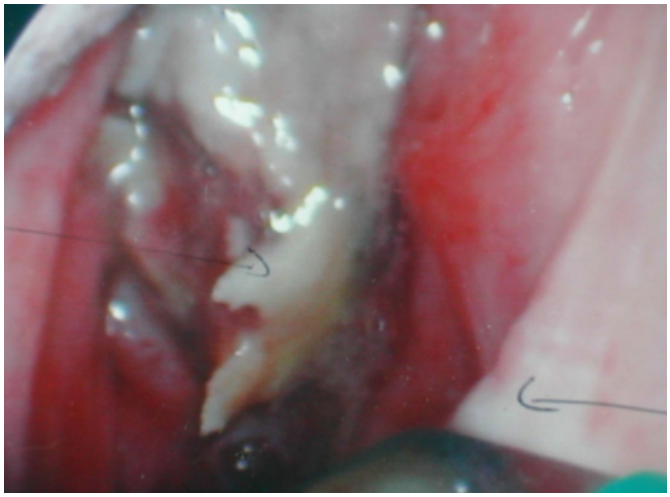
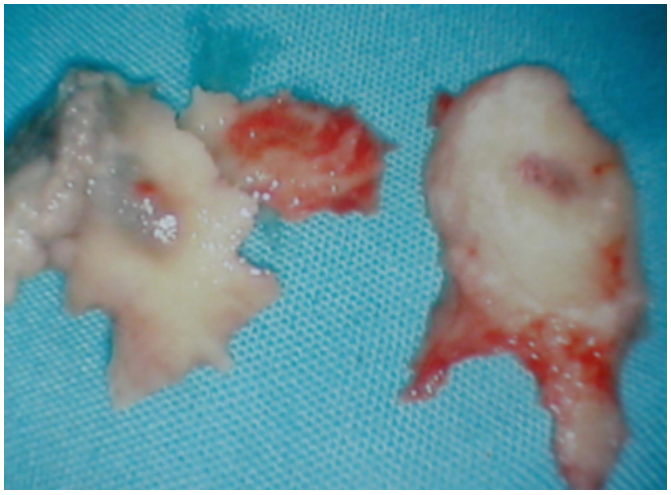


Figure 5

Figure 5: Multiple bony sequestra removed from the nasopharynx



DISCUSSION

There exists considerable controversy in the literature over whether plasmacytomas represent an early localised stage of multiple myeloma or are distinct entities. Unlike medullary plasmacytomas, in which dissemination or progression to multiple myeloma occurs in up to 85% of patients, extramedullary plasmacytomas usually remain well localised. Only 15% to 20% of cases progress to multiple myeloma or develop regional disease. Local recurrence may occur in 6% to 10% of cases that have had adequate initial treatment. [3] Wiltshaw in a study of the natural history of extramedullary plasmacytoma suggested that bony lesions developed in 81% of patients with disseminated extramedullary plasmacytomas. However, these lesions differed in character and distribution from those found in patients with multiple myeloma. [6]

Overall, in 61.1% of the cases with treatment for extramedullary plasmacytoma in the upper aerodigestive tract, no multiple myeloma or recurrence occurred; in 22% there was a recurrence, and in 16.1%, there was a conversion to multiple myeloma. 0.8% with extramedullary plasmacytoma developed solitary plasmacytoma of the bone. [1]

Differential diagnosis of this lesion exhibiting small round cells includes undifferentiated carcinoma, malignant melanoma, olfactory neuroblastoma and malignant lymphoma. With the use of Immuno-cytochemical staining techniques and electron microscopy these entities can usually be separated.

Alexiou et al suggested that surgery alone gave the best results for EMP of the upper aero-digestive tract when respectability is good. However, if complete surgical resection was not possible or doubtful, and/or lymph nodes were affected, then combined therapy (surgery and radiation) was recommended. [1]

There are very few reports of local recurrence of plasmacytomas treated primarily by surgery. [6,8] Surgery is usually recommended if there is local failure after radiotherapy in a respectable tumour as was the case for our patient.

The KTP laser was used in our case for precise excision of the recurrent lesion in the nasopharynx and to minimise bleeding. We intend to follow our patient in the head and neck clinic for life to detect any further recurrence.

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