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A case of extramedullary plasmacytoma arising from the nasal septum

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Abstract
A rare case of extramedullary plasmacytoma arising from the nasal septum with localized amyloid deposition is reported. A 75-year-old woman presented with a history of post-nasal discharge and nasal obstruction for several months. Endonasal endoscopic observation revealed the presence of a mass arising from her nasal septum with an extension to the nasopharynx. No cervical lymph nodes were palpable. The biopsy specimen was diagnosed as a plasmacytoma (IgG, lambda-light chain type). Both serum myeloma-protein and urine Bence-Jones protein were negative. Bone marrow biopsy, a chest radiograph, total body skeletal survey and ^{67}Ga- and ^{99m}Tc-scintigrams showed no other systemic lesions. These findings confirmed the diagnosis of extramedullary plasmacytoma in the nasal septum. The patient received irradiation of 40 Gy without clinically detectable reduction of tumour size. The patient eventually underwent complete resection of the tumour by KTP/532 laser under endonasal endoscopic control. Pathologically, the tumour mass was composed mainly of amyloid deposition with a marked reduction of tumour cells. This indicated the radiosensitiveness of tumour cells, which was clinically masked by the increased amyloid deposition. The clinical presentation, pathological features and surgical procedures are described with a review of the literature.

Key words: Plasmacytoma; Amyloid; Nasal septum

Introduction

Plasmacytoma is a rare neoplasmic disorder derived from a lineage of B lymphocytes. Most plasmacytomas present as multiple myeloma, in which the disease can be widespread and incurable at diagnosis. Extramedullary plasmacytoma (EMP) is an uncommon malignant neoplasm arising outside the bone marrow without clinical evidence of multiple myeloma. EMP, which can occur in virtually any part of the body, shows a predilection for the head and neck region[1,2] and comprises four per cent of all non-epithelial tumours of the nasal cavity, nasopharynx, and paranasal sinuses.[3] We describe a case of EMP with localized amyloid deposition arising from the nasal septum, showing some of the clinical and pathological characteristics before and after the combined therapy of radiation and surgery.

Case report

A 75-year-old Japanese female had had a history of post-nasal discharge and nasal obstruction for several months. On endonasal endoscopic observation, a grey-white submucosal mass was seen in the nasal septum, extending to the nasopharynx. No cervical lymph nodes were palpable. Computed tomographic (CT) scan showed that the mass protruded from the posterior part of nasal septum with an involvement of the septal cartilage (Figure 1). Magnetic resonance imaging (MRI) revealed that the tumour had a relatively well-defined margin in the posterior part of the nasal septum with low signal intensity in T2-weighted images. In T1-weighted images, the tumours were isointense or slightly hyperintense compared with surrounding muscles, and exhibited a positive gadolinium-enhancement (Figure 2). Histopathological examination of biopsy specimens revealed a neoplasm consisting of a uniform

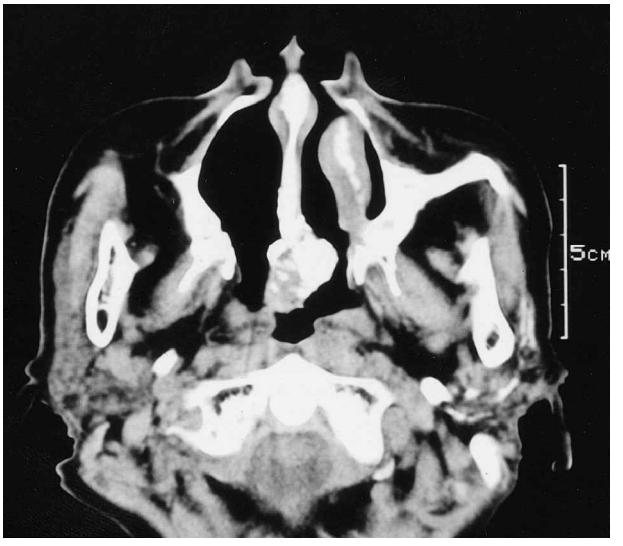

Fig. 1

An axial CT scan showing a mass protruding from the posterior part of the nasal septum with cartilage involvement, extending to the nasopharynx.

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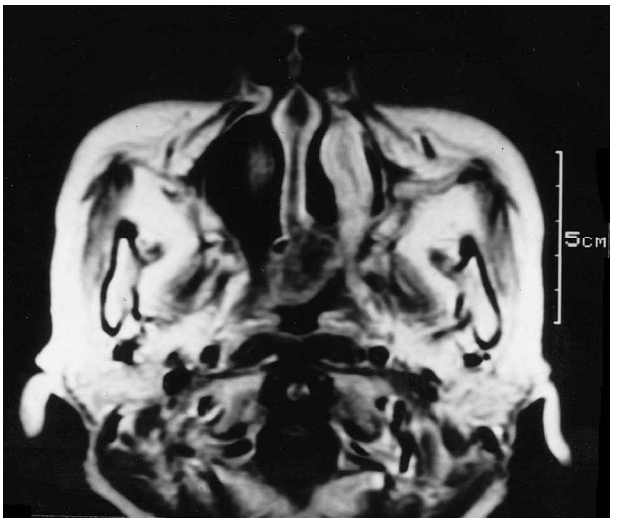

FIG. 2
An axial contrast enhanced T1-weighted MRI showing a mass with well-defined margins in the nasal septum.

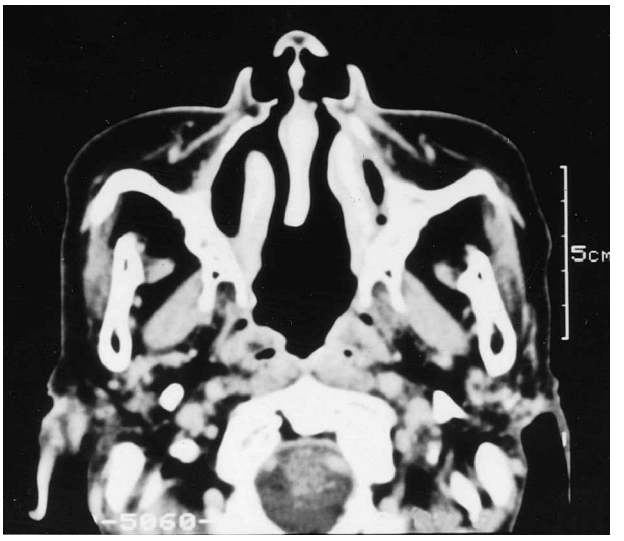

FIG. 4
An axial CT scan revealing complete removal of the tumour.

population of atypical mononuclear cells similar to plasma cells with foci of amyloid deposition (Figure 3). Immunoperoxidase staining showed these atypical cells stained solely positive for IgG and the lambda light chain, but not for the kappa chain. The tumour cells were positive for epithelial membrane antigen and negative for CD20 (pan-B cell marker), CD3 (pan-T cell marker) or cytokeratin, consistent with plasma cell differentiation. Based on these findings, the tumour was diagnosed as plasmacytoma. The amyloid showed red staining and so-called greenish birefringence with Congo red stain, and this staining was resistant to pretreatment with potassium permanganate. These results indicated that this amyloid did not consist of AA amyloid, and was most likely AL amyloid.

Further clinical examinations were undertaken to check the presence of multiple myeloma: bone marrow biopsy, skeletal survey, haematocrit, white blood cell count, white cell differential, urea and electrolytes, creatinine, calcium, quantitative immunoglobulins, immunoelectrophoresis and Bence-Jones protein in the urine. All of these tests were within normal limits. ^{67}Ga- and ^{99m}Tc-scintigrams did not detect other systemic lesions. A multiple myeloma was, accordingly, ruled out, and the final diagnosis of primary extramedullary plasmacytoma of the nasal septum was confirmed.

The patient was treated with radiation of 40 Gy (2 Gy per day) for four consecutive weeks to a field covering the nasal septum and nasopharynx. However, the tumour did not show any reduction in tumour size by both endoscopic and CT scan observations. Thus, the tumour was removed by the transnasal endoscopic approach using a KTP/532 laser. The nasal septum was resected with a tumour margin of 1.5 cm. The operative findings confirmed that the tumour originated from the posterior edge of the nasal septum, extending to, but not adherent to, the nasopharyngeal wall. CT scan after the operation exhibited a complete removal of the tumour (Figure 4). Pathological examination of the surgical specimens (Figure 5) showed an abundant deposition of amyloid, which was resistant to pretreatment with potassium permanganate. Visible tumour cells were only sporadically observed. This suggested that the resistance of tumour to radiation therapy (no tumour mass reduction) could be ascribed to the amyloid deposition, and that most of the tumour cells were in fact radiosensitive. The patient was alive without recurrence or metastasis five months after the operation.

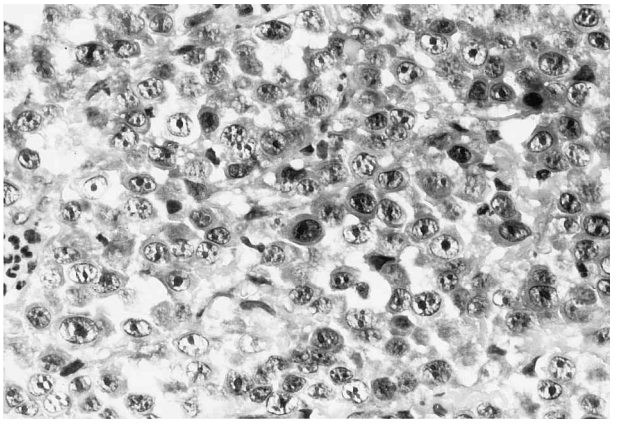

FIG. 3
Biopsy specimen of the mass of the nasal septum showing diffuse proliferation of atypical plasma cells (H&E; ×400).

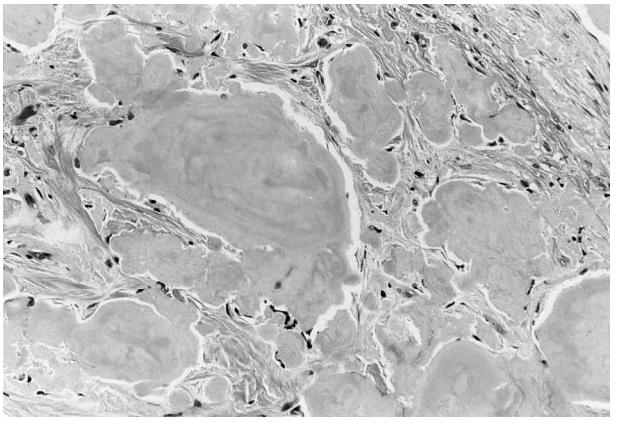

FIG. 5
Stromal amyloid deposition in surgical specimen. Note nearly total disappearance of tumour cells (H&E; ×200).

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Discussion

EMPs are plasma cell tumours involving soft tissue. Nearly 80 per cent of the EMP of the head and neck region can be found in the submucosa of the upper respiratory tract or oral cavity.[1,4,5] Among EMPs in the upper respiratory tract, the nasal cavity is one of the most common sites together with the paranasal sinuses and nasopharynx.[6] Although more than 50 cases of EMP in the nasal cavity have been reported to date,[5-8] tumours arising from the nasal septum are quite rare.[9]

In the present case, a significant shift to IgG lambda chain without detectable kappa chain reaction was demonstrated in the tumour cells by a routine immunoperoxidase method. Gene rearrangement molecular techniques are also useful to investigate the monoclonality. However, such techniques at the present time are time-consuming and very expensive,[8,10] and they were not conducted in this case. Amyloid occurs in 15 to 38 per cent of EMPs, and thus its detection provides a histopathological clue for the diagnosis of EMP.[2] However, no clinical correlation was observed between stromal amyloid deposit and prognosis.[5] In our case, the tumour-associated amyloid deposition showed some interesting features. The biopsy specimen before radiation therapy mainly consisted of neoplastic plasma cells with islands of deposits of amyloid surrounding the cells. In contrast, the surgical specimen after radiation therapy was mainly composed of amyloid substance with only dispersed tumour cells. These histopathological findings suggested that the tumour cells were sensitive to radiation and that the amyloid substance replaced the tumour cells.

Since the majority of patients are elderly and present with low-grade tumours, treatment of EMP is somewhat controversial. Some clinicians favour radiotherapy and others favour surgical management. Radiation therapy dosed between 30 and 50 Gy is often used to manage EMP because the plasma cells are highly radiosensitive.[2,5] The disease is also sometimes managed with surgery followed by radiation therapy.[6,11] Other options for the treatment include high dose radiotherapy, adjuvant alkylating chemotherapy[5] and surgical resection. Since, in our case, the tumour failed to be remarkably reduced by radiation and the patient had radiation-induced stomatitis in her soft palate, minimally invasive surgery was selected. Suzuki *et al.*[10] in Japan, reviewed surgical approaches for eight cases of EMP located in the nasopharynx: (a) four cases by the transpalatine approach, (b) one case by the trans-maxillary approach, (c) one case by the trans-palatine plus trans-maxillary combined approach, and (d) two cases by the endonasal approach. The trans-palatine or trans-maxillary approach caused stress for the patients, whereas the endonasal approach incurred the possibility of incomplete resection.[10] Since 1986, KTP/532 lasers have been used in a variety of medical applications and applied to patients with chronic sinusitis, mucoceles and nasopharyngeal cysts.[12] Its marked advantage is the use of guiding flexible fibres that enables deep lesions to be treated under endoscopic control. By endoscopic sinus surgery using the KTP/532, the residual tumour could be resected safely and accurately without incising the soft palatine. In our case, the tumour seemed to be macroscopically radioresistant because of abundant amyloid deposition; radiation combined with minimally invasive surgery was preferable to high dose radiotherapy or chemotherapy. Therefore, we have to consider extensive amyloidosis as a differential diagnosis when we observe radioresistant EMP.

Local recurrence may occur in six to 10 per cent of cases that have had adequate initial treatment.[1,8,13] Furthermore, dissemination into multiple myeloma has been reported to occur in 10 to 30 per cent of EMP cases within the first two years.[1,4,8,14] A rise in the baseline levels of serum M monoclonal proteins or urinal Bence-Jones protein may signify a recurrence of the primary disease or may indicate the onset of multiple myeloma.[11] Thus, long-term follow-up including not only a local control but also systemic observations via blood counts or immunoglobulin measurement are necessary.

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