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# Solitary plasmacytoma of the epiglottis: a case report and review of the literature

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**Abstract**
A 59-year-old white male presented with a two-month history of dry cough and shortness of breath. At bronchoscopy, a 1 cm mass on the laryngeal surface of the epiglottis was found. Immunohistochemical stain of the biopsy specimen revealed a monoclonal proliferation of plasma cells. Serological tests revealed normal serum immunoglobulin levels, and a bone marrow aspirate and biopsy was also normal; no abnormalities were found on serum or urine electrophoresis. He received radiotherapy (50 Gy), and was doing well three years following therapy without evidence of disease.

**Key words: Plasmacytoma; Epiglottis**

## Introduction

Plasma cell neoplasms are monoclonal proliferations and accumulations of plasma cells. Localized solitary plasmacytomas are rare accounting for five to 10 per cent of all plasma cell malignancies (Corwin and Linberg, 1979; Knowling *et al.*, 1983; Mayr *et al.*, 1990; Shih *et al.*, 1995). The majority of extra-osseous or extramedullary plasmacytomas are seen in the head and neck region and frequently arise from the submucosa of the nasopharynx, nasal cavity, paranasal sinuses and tonsils (Wiltshaw, 1976; Corwin and Linberg, 1979; Knowling *et al.*, 1983; Mayr *et al.*, 1990; Shih *et al.*, 1995). Solitary plasmacytoma of the epiglottis is extremely rare with only seven cases having been reported in the English literature. In this report, we review the literature and present an additional case that was treated with radiotherapy with a three year follow-up.

## Case report

A 59-year-old white male presented in September 1993 with severe coughing spells and associated shortness of breath. He underwent bronchoscopy and, while no abnormalities were found in the trachea or bronchus, a lesion on the laryngeal surface of the epiglottis was visualized. This appeared smooth in appearance although the mucosa was somewhat thickened. Three months later a further direct laryngoscopy was performed and this revealed a 1.0 cm hard, sessile, spherical, non-mobile, white submucosal lesion on the laryngeal surface of the epiglottis just above the petiolus. Biopsy was performed without complications.

Histological examination revealed a dense infiltration of the respiratory submucosa by sheets of mature plasma cells with mildly atypical nuclei. The submucosal infiltrate was very homogeneous. The neoplastic nature of the plasma cell infiltrate was confirmed by immunohistochemical studies. These studies demonstrated a clonal population of plasma cells showing restricted expression of kappa light chains (Figure 1) and IgG heavy chains. A diagnosis of extramedullary plasmacytoma was made.

A complete blood count (CBC) showed a white blood cell count of 6100/mm$^3$, haemoglobin of 15.7 g/dl and a platelet count of 309,000/mm$^3$. The white cell differential was within normal limits. The beta-2 microglobulin level (normal range 0–2.0 mg/l), total protein and the globulin level were also normal. Serum protein electrophoresis (SPEP) showed normal levels of IgG, IgA and IgM (1070, 264 and 52 mg/dl respectively) and the $\alpha$-1, $\alpha$-2, $\beta$, and $\gamma$ globulins were also within normal limits. Urine total protein was less than 5 mg/dl and no bands of restricted electrophoretic mobility were seen on urine protein electrophoresis (UPEP). Bone marrow aspirate and biopsy showed a normocellular marrow with normal architecture and all haematopoietic elements present in normal ratios. No plasma cell infiltrate was noted. A computed tomography (CT) scan showed no adenopathy in the neck or thorax and no bony abnormalities were identified on a skeletal survey.

It was decided that the patient should have radiotherapy and he received a total dose of 50 Gy in 2 Gy fractions, ending in March 1994. The patient has done well since completion of treatment and was last seen in follow-up in April 1997 when he was symptom free and without any bone pain or tenderness. Fibreoptic examination of the larynx at that time showed no evidence of recurrent disease. Subsequent repetition of his serum and urinary electrophoresis were normal.

## Discussion

Solitary plasmacytoma can be either osseous (solitary plasmacytoma of bone; SPB) or extramedullary (plasma-

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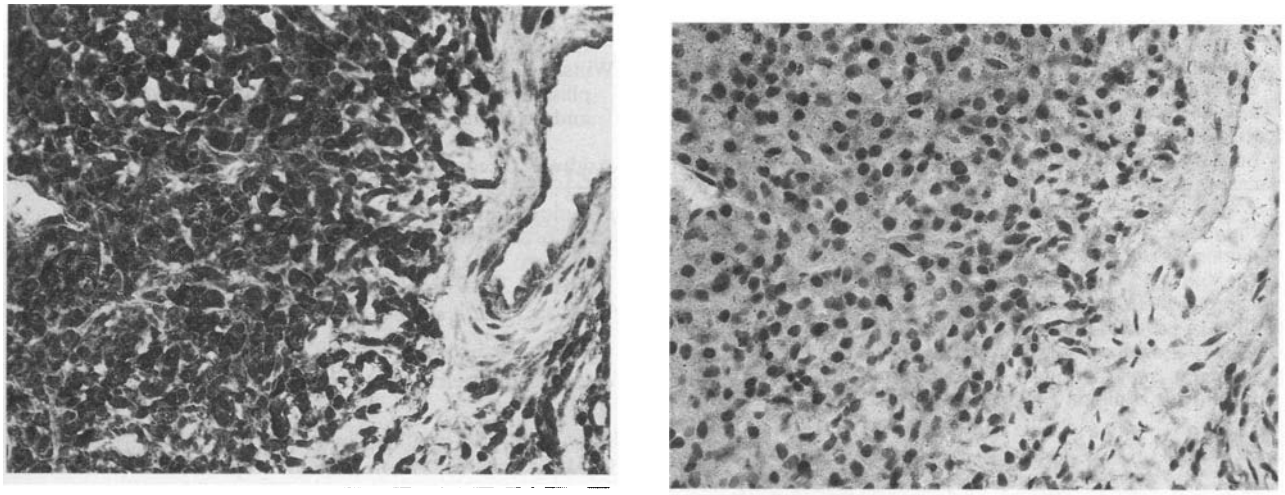

(a) (b)

FIG. 1

Immunohistochemical staining for immunoglobulin chains confirms the monoclonal nature of the plasma cell infiltrate. The plasma cell express kappa light chains (1a, × 400), but they do not express lambda light chains (1b, × 400).

cytomas of soft tissue: EMP). Extramedullary lesions arise submucosally in the head and neck and the most frequent sites of involvement are the nasopharynx, the nasal cavity, the paranasal sinuses and the tonsils (Wiltshaw, 1976; Knowling *et al.*, 1983; Mayr *et al.*, 1990; Shih *et al.*, 1995). Laryngeal extramedullary plasmacytomas are uncommon (Pahor, 1978) and solitary lesions of the epiglottis are rare with only seven cases having been reported previously.

The outcome of patients presenting with epiglottic EMP seems to have been generally favourable although it is difficult to draw firm conclusions from the small numbers that have been reported. The first paper presented two cases (Carson *et al.*, 1955) but it is doubtful that the pathology was correct in one of the patients included and the second, who had a surgical resection, died 13 years later of unknown causes. In a subsequent case, managed with radiotherapy (24 Gy), a patient developed generalized disease eight years after treatment in spite of good local control (Todd, 1965) but a further patient, who presented with a neck mass and a primary lesion on the epiglottis, and who was also treated with radiotherapy (64.15 Gy), was alive with no evidence of disease six years later (Fishkin and Spiegelberg, 1976; Petrovich *et al.*, 1977). Unfortunately a report of a further patient who received surgical treatment gave no details as to the outcome (Gambino, 1988) but in another case, the only female in the literature, the patient survived a supraorbital plasmacytoma seven years previously and presented with an epiglottic lesion which was also successfully treated with radiotherapy (55 Gy) before succumbing to carcinoma of the cervix three years later (Bush *et al.*, 1981). In two subsequent cases one patient enjoyed a 10-year disease-free interval following radiotherapy (45 Gy) and another had a laryngoscopic excision of a mass at the tip of the epiglottis and remained well and disease-free 3 years later (Kost, 1990; Rolins *et al.*, 1995).

When our case is added to the literature there are a total of eight cases of epiglottic EMP (seven male and one female). Of these one was asymptomatic at presentation, one presented with a neck mass and the others had symptoms referable to the upper air and food passages (dysphagia, muffling of the voice, cough or dyspnoea) at diagnosis. Radiotherapy or surgery has appeared to offer satisfactory control in the majority with only one case developing systemic disease. This finding corresponds with the dissemination rate of EMP from other laryngeal primary sites where multiple myeloma has been reported in only four of the 31 cases in the literature (Pahor, 1978). Nonetheless it is clear that the diagnosis of extramedullary plasmacytoma (EMP) should normally provoke investigation for disseminated disease and this should include a skeletal survey, serum and urinary protein electrophoresis, serum immunoglobulins and bone marrow biopsy (Salmon and Cassady, 1997). Immunohistochemical staining for light and heavy immunoglobulin chains is also thought to be helpful in documenting the nature of the plasma cell proliferation and in confirming the diagnosis.

In our case such investigations appeared to confirm that the disease was localized to the epiglottis and the patient has done well with radiotherapy alone. He remains under observation, but at present is symptom free with no evidence of generalized disease.

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