

Extramedullary plasmacytoma in Head and Neck

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Abstract Extramedullary plasmacytomas(EMP) are uncommon tumors. We reported two cases of EMP, one in the neck and one in the larynx. Two patients received radiotherapy which dose was 45Gy in one month, followed by four-course M2 chemotherapy. Two patients were followed up 12 years or 9 years respectively, there were not recurrence and no systematical diseases. They are engaging in their own work.

Key words Extramedullary plasmacytoma; larynx; neck;radiotherapy

Extramedullary plasmacytomas (EMPs) are uncommon tumors. The paper is a review of two cases of EMP from 653 malignant tumors in the head and neck from 1979 to 2002, one in the neck and one in the larynx.

CASE REPORT

Case 1

A 42-year-old man was admitted on 29th, May, 1992 because of unintentionally finding a painless mass in the middle of the left neck four days ago. Physical examinations showed a 3cm×3cm soft mass lacking of mobility, it was located under the middle of sternocleidomastoid muscle and the patient felt no pain when it was pressed and its border was unclear. B ultrasound revealed the mass about 4cm×3cm×2cm with low echo outside of the upper of the left thyroid and its border was clear, the inside echo was equal and its shape did not change when it was pressed. The thyroids were normal and no swelled lymph node in the neck. The mass was completely excised including 0.5cm normal tissue around tumor. During the operation, we find about 4cm×3cm×3cm mass was under the middle of sternocleidomastoid muscle and lateral of the left internal jugular vein.

Case 2

A 57-year-old man was admitted on 24th, Feb, 1995 because of hoarseness three months ago. He had a history of smoking with 20 cigarettes per day over 20 years. Examinations under indirect laryngoscope revealed the left laryngeal ventricle was swelling and had a about 0.3cm×0.3cm mass in anterior-middle part of the left vocal cord. Suspension laryngoscope showed a pink smooth mass located in the left laryngeal ventricle obscuring surface of the whole left vocal cord, the border of the mass was smooth. The infraglottic portion were normal. The mass was excised under suspension laryngoscope.

There were no abnormal in digest and respiratory system of two patients. Results of serum and urine electrophoresis tests failed to demonstrate any myeloma component or Bence Jones immune globulin.

The pathological diagnose after surgery: a number of the mature plasma cells of the tumor appeared abundant neoplastic proliferation and diffuse distributions. The results of immunohistochemistry showed the tumor cells produced monoclonal IgG and positive IgGλ, diagnosed as (the left neck or larynx) EMP respectively (fig. 1,2). The diagnosis accorded the EMP criteria ① a bone marrow biopsy showing less than 10% plasma cells. ② normal skeletal survey. According to histological grading criteria devised by Bartl *et al*^[1] for EMP, the plasma cells in our cases belonged to Grade 1.

Two patients received radiotherapy which dose was 45Gy in one month, followed by four-course M2 chemotherapy: in the first day, carmustine, 0.5mg/kg, intravenous injection and cyclophosphamide (CTX), 10mg/kg, intravenous injection; melphalan, 0.25mg/kg,

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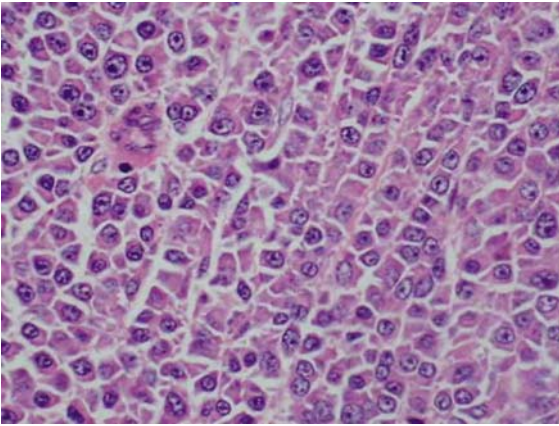


Fig. 1 Extramedullary plasmacytoma in the larynx: a number of mature plasma cells of the tumor appeared abundant neoplastic proliferation and diffuse distributions (HE×200)

p.o, from the first day to fourth day; prednisolone tablets, 1mg/kg, p.o, from the first day to the seventh day, then 0.5mg/kg, from the eighth day to fourteenth day; vincristine (VCR), 0.03mg/kg, intravenous injection, the twenty-first day; five weeks as a chemotherapy course. Two patients were followed up 12 years or 9 years respectively, there were not recurrence and no systematical diseases. They are engaging in their own work.

DISCUSSION

EMP is less than 1% of the head and neck tumors, the most common location of EMP is in nasal cavity, sinuses and nasopharynx^[2,3], and is rare arising in the neck or larynx. The rate of EMP in our finding was 0.3%(2/653) in head and neck malignant tumors. Men are predominantly affected, and these tumors are more commonly seen in the forty to fifty ages. The principal symptom of EMP in the larynx is hoarseness. If the tumor becomes larger gradually, it can occur dyspnoea and/or dysphagia^[4]. The sign of EMP in the neck is usually a painless mass and can grow gradually. Our cases demonstrated hoarseness and painless mass in the neck.

The EMP diagnosis criteria: (1) a pathology-proven after surgery plasma cell tumor involving a single extramedullary site with or without lymph node in the neck involvement; (2) a bone marrow biopsy showing

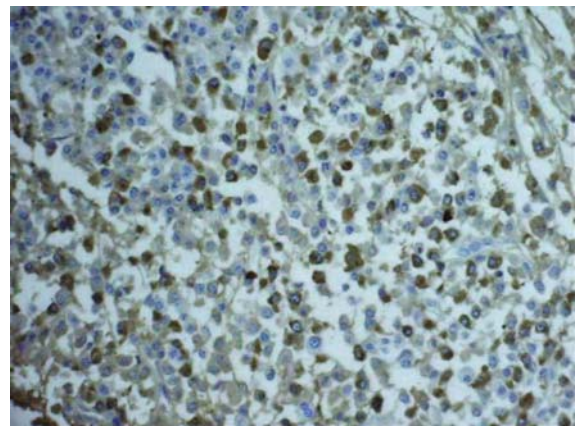


Fig. 2 Extramedullary plasmacytoma in the larynx (immunohistochemical stain): monoclonal IgG λ positive(ABC×100)

less than 10% plasma cells; (3) normal skeletal survey. immunohistochemical stain can differentiate EMP from other tumours or reactive inflammatory infiltrates. Plasmacytomas originating from B lymphocyte are monoclonal proliferation tumors which can produce kappa or lambda light chain restricted-immunoglobulins(IgG κ or IgG λ), however, non-tumor plasmacytes are polyclonal proliferation B lymphocytes. Susnerwala et al found there was a good prognosis when the lower-grade plasmacytes of EMP and/or expression of kappa light chain restricted-immunoglobulins^[3]. The plasma cells in our cases belonged to Grade 1 but the expression of light chain restricted-immunoglobulins was IgG λ . They have survived over 12 years or 9 years.

The treatment for EMP in head and neck depends on the size of tumor and whether the tumor is systemic spread. The most suggested that the best way be combination surgery with radiotherapy^[2-4]. If EMP advanced to MM or recurrence or high malignancy, the chemotherapy should be introduced^[3]. If EMP localizes inside larynx, or it can be removed by surgery even if the tumor spreads outside, radiotherapy followed total laryngectomy should be recommended. However, EMPs easily progress to MM and recur. Susnerwala et al reported a case of EMP in nasopharynx advanced to MM ten years after radiotherapy and chemotherapy. Of EMPs, 38% spread to bony sites, 20% to soft tissue, and 42% to both^[5]. Therefore, all patients with EMP should

be regularly examined serum and urine electrophoresis to test any myeloma component or Bence Jones immune globulin, skeletal X-ray, chest X-ray, esophagram, B-ultrasound in the neck and bone marrow biopsy. We insisted on following up our patients per year in order to find the transformation of EMP to intervene timely.

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Errata for the U.S. Chinese Journal of Lymphology and Oncology, Vol.6.No.1, 2007

Owing to the fault of our job, we did not put the correct volume number and issue number on the top margins of Quanqing Zheng's article on page 31.

We will change the errata on the online fulltext. We apologize for any inconvenience it may cause to Dr.zheng.