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=국문초록=

Solitary Extramedullary Plasmacytoma of the Head and Neck

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Solitary Extramedullary plasmacytoma는 plasma cell neoplasm중 드문 것으로 다른 형태의 plasmacytoma와는 임상적 병리학적으로 완전히 다른 종양으로 알려져 있다. 원발병소는 주로 두경부 특히 상기도로써 주로 국소부위에 발생하며, 국소방사선치료가 근본적 치료로 받아들여지고 있다.

저자들은 1970년 1월부터 1984년 12월까지 두 경부의 solitary Extramedullary Plasmacytoma로 확진되어 연세암센터 방사선 치료실에서 방사선 치료를 받았던 5예를 대상으로 추적 조사하여 그 결과를 보고하는 바이다.

전 5예 모두 진단당시 국소부위에 단일 병소를 갖고 있었고 이중 1예에서 원발병소에 수술을 시행하였으나 수술후 국소재발이 있었고 곧 전신으로 퍼졌으며 진단후 3년 6개월만에 사망하였다. 나머지 4예중 2예에서는 방사선치료만 시행하였고 다른 2예에서는 수술 및 수술후 방사선 치료를 시행하였는데 4예 모두 현재까지 병변없이 생존하고 있다.

ABSTRACT

The details of 5 patients with extramedullary plasmacytoma of the head and neck were reviewed for the period from 1970 to 1984. All patients were presented with localized disease at the time of diagnosis. Out of 5 patients the one treated with surgery alone developed local recurrence and disseminated disease. He died at 3 years and 6 months. Rest of the four were alive with no evidence of the disease. Two patients were treated with radiation therapy alone while other two were treated with surgery and postoperative radiation therapy. Curative radiation therapy is recommanded after the diagnosis is established by biopsy examination.

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INTRODUCTION

Solitary extramedullary plasmacytoma (EMPC) is an unusual form of plasma cell neoplasm and it was recognized as having an entirely different clinicopathological entity from that of other forms of plasmacytoma[2]. Most common site of origin is in the head and neck and especially in the upper

air passage. EMPC tend to remain localized and local irradiation has been accepted as the treatment of choice[2].

The purpose of this review is to detail our experience with 5 patients with EMPC, all of which involved the head and neck. The result of therapy are discussed in the comparison with the other reports in the literature. We have discribed the diagnosis, treatment, and follow-up data.

MATERIALS AND METHODS

The records of all patients with soft tissue plasmacytoma seen at the department of radiation oncology, College of Medicine, Yonsei University, Yonsei Cancer Center from the period January 1970 to December 1984 have been reviewed.

The diagnostic criteria for inclusion were as follows; (1) plasmacytoma arising in extraskeletal site, (2) plasmacytoma proved by biopsy of the lesion, (3) normal serum protein or serum electrophoresis at the time of diagnosis, (4) no Bence-Jones protein in urine, (5) normal marrow biopsy (less than 10% plasma cell), (6) normal skeletal radiographs, aside from bony involvement that was in direct continuity with tumor mass. Six cases met the criteria for this analysis of EMPC. In all 6 patients, the EMPC arose in the head and neck. One patient lost in follow-up was excluded from the analysis, and so 5 patients were evaluated for this report.

The patients age ranged from 35 to 65 years at the time of diagnosis. All patients were males. The involved sites and presenting symptoms were swallowing difficulty and foreign body sensation of the throat in the case of suprahyoid epiglottis (patient 1), nasal obstruction with proptosis when the nasal fossa was involved (patient 2), nasal stuffiness in two cases with maxillary antral mass (patients 3 and 4), and pus discharge and bulging mass on the left side dorsum of nose in the case of nasolacrimal ductal mass (patient 5).

Only one patient demonstrated metastasis on cervical lymph node bilaterally at the time of diagnosis. In the radiographic findings, two patients (patients 2 and 3) revealed bony destruction. Of the four patients who were treated with radiation therapy, two received radiation therapy alone, and the other two patients were treated with surgery and postoperative irradiation. Only one patient was treated by surgery alone. All four patients were treated with radiation using a Co-60 unit. The treatment ports included the tumor bearing area. The treatment was given 5 days a week with the daily tumor dose ranged from 3,400 to 6,600 rads.

RESULTS

Details of the patients under reviewed are summarized in table 1.

There was no evidence of disseminated plasma cell disease at the time of diagnosis in any patient. Performed protein electrophoresis and bone marrow biosy, it was within normal limit and there was no evidence of Bence-Jones protein.

Four patients including the two who received radiation therapy alone and two with surgery and postoperative radiation therapy showed complete regression of disease. One (patient 3) who did not receive radiation therapy and were treated by partial maxillectomy only developed local recurrence and subsequently disseminated disease. The patient died at 3 years and 6 months. In this patient, M protein was not noted initially. But it increased with subsequent dissemination and then disappeared after two course of mephalan and one course of cytoxan.

The patient who had a lesion of suprahyoid epiglottis with bilateral cervical lymph node metastasis showed complete response in primary site and metastatic lymph node lesion following radiation.

Of two patients with bony destruction in the radiographic finding, one (patient 2) was treated successfully with radiation while the other patient (patient 3) failed with surgery alone.

Table 1. Summary of Patient Details

Patient Age/Sex	Symtoms & Signs	Site of primary	Lab. & X-ray Findings	Tx of primary	Follow-up
65/M	Swallowing difficulty & foreign body sensation on throat (D; 5months)	Suprahyoid epiglottis	Unknown	Radiation Tx	NED at 14 years & 6 months
45/M	Nasal obstruction (D; 8months) Proptosis (D; 3months)	Left nasal fossa	Negative/Bony destruction of nasal septum & left ethmoid sinus wall	Radiation Tx	NED at 6 years & 11 months
43/M	Nasal stuffiness & proptosis (D; 3months)	Left maxillary antrum	Negative/Bony destruction of left maxillary antrum extending to the left ethmoid sinus	Partial maxillectomy	Died at 3 years & 6 months
38/M	Nasal stuffiness & posterior nasal dripping(D; 8months)	Left maxillary antrum	Negative/Haziness, no bony destrruction	Caldwell-Luc operation & Radiation Tx	NED at 5 years & 9 months
35/M	Pus discharge & bulging mass on left side dorsum of nose	Left nasolacrimal duct	Negative/Negative	Excision & Radiation Tx	NED at 2 years · 8 months

DISCUSSION

Solitary extramedullary plasmacytoma is rare malignancy[2,3] and has been recognized as an entity distinct from both solitary plasmacytoma of bone and multiple myeloma in the recent years[6]. The majority of EMPC develop in the head and neck[3,17,18,24] and especially in the upper respiratory tract and oral cavity[3,4,8,22] In this area, the nasal cavity and paranasal sinuses are most frequent site of involvement[5,11,12,21]. Despite these observation, they comprised less than 1% of all tumors occuring in this region[16].

The tumor is commonly found in the 6 th and 7 th decades, but may develop at any age. This disease shows a strong male predominance at a ratio of at least 2 : 1[4]. In our series, patients ranged in age from 35 to 65 years and were all males.

Clinical presentation varies according to the involved organ[3,11]. The usual symptoms arise due to pressure or obstruction caused by the tumor initially[3,8,19] and after some time mucous discharge and hemorrhage developed as the tumor enlarged and ulcerated. If neural or bony invasion are presented local pain may be a prominant symptom[8,22]. Symptoms are usually present six months to two years before the diagnosis is made[10].

Cervical lymph node is an infrequent finding in plasmacytoma of the head and neck[5]. Hellwig noted nine of sixty four patients in his review[12]. Lymph node involvement is reportedly of no prognostic significance[17,19]. In our series, only one patient demonstrated metastatic disease in the cervical lymph nodes bilaterally. This lymphadenopathy was noted at the time initial diagnosis. After radiation therapy, the lesion of primary site (suprahyoid epiglottis) and cervical lyph nodes were completely regressed. And the patient was alive with NED at 14 years and 6 months.

The radiographic findings of these lesions varies according to the location and extensiveness. When the tumor is confined to the submucosa and mucosa, it is visible as a soft tissue mass. Tomography is recommended in order to define the degree of bone destruction[16,20]. Bony destruction has been reported to adversely effect the prognosis[2,3,12,18], but the experience at M.D. Anderson Hospital does not conform this impression[6]. Of

eight patients with extramedullary plasmacytoma who are NED beyond 3 years, six presented with bony destruction. In present cases, two patients demonstrated bony destruction in radiographic findings. One patient controlled with radiation.

It is suggest that the good prognosis of plasmacytomas of the head and neck, in contrast to plasmacytomas arising at other site[7], is related to the fact that a high proportion of these tumors are localized at the time of diagnosis and that very large primary tumors are uncommon[1,16,18,23].

Once the diagnosis of EMPC of the head and neck has been established histologically, the appropriate clinical, hematological, biochemical and radiological investigations must be performed to exclude the presence of disseminated disease[24].

Based on our experience and review of the medical literature it may be concluded that these neoplasms are associated with good prognosis following radiotherapy[5,9,13,21,23]. The efficucy of the treatment can be judged in three ways[23]; the initial effect on the tumor, and the subsequent incidence of local recurrence and disseminated disease. Local recurrence occurs most frequently in the first five years after initial treatment, but may occur many years later[5,13,21]. The development of local recurrence sometimes heralds the onset of disseminated disease. Disseminated disease may take the form of either typical myelomatosis or metastasis to soft tissues and bones not classically affected in multiple myeloma[5,13,20].

In presenting series, all patients presented with localized disease in the head and neck at the time of diagnosis. 4 patients were treated successfully by radiotherapy or surgery and postoperative radiation therapy. These patients were alive with NED. One patient (patient 3) who had a lesion at the maxillary antrum were treated with surgery alone. We couldn't perform external radiation therapy to this patient in whom only partial maxillectomy was performed, because he already received radiation therapy and surgery one year ago due to transitional cell carcinoma of the nasal cavity. In this patient, local recurrence occurred at one year after surgery, which was performed by surgery and chemotherapy. But the patient died because of systemic dissemination to the soft tissues and bones.

The tumor can be controlled with local irradiation and surgical resection[14]. Radiation therapy is the primary treatment of choice for these tumors[2,14,23] and the recommanded tumor dose is 3,500~5,000 rads. The treatment ports should include the primary tumor and the regional lymph nodes when there is a clinical suspicion of involvement[15]. Since the introduction of alkylating agents is effective against plasma cell neoplasm, good results have been reported for EMPC and chemctherapy should be used in the management of radioresistant and recurrent tumors[23].

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