

Huge Size Intracranial Plasmacytoma Treated with Surgery and Fractionated Stereotactic Radiotherapy

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Surgery and radiotherapy are mainly used for plasma cell neoplasm which constitutes about 1~2% of human malignancy. The authors carried out Fractionated Stereotactic Radiotherapy(FSRT) on the residual tumor after the subtotal removal of intracranial plasmacytoma. A huge mass lesion was observed on MRI (magnetic resonance image) in the left anterior and middle cranial fossa of a 63-year-old man with left exophthalmus which lasted for a month, and was suspected as a meningioma with strong contrast enhancement. Extramedullary plasmacytoma was diagnosed on histopathological examination. After the surgery, FSRT was also carried out on the residual tumor which invaded the skull base. One-year follow up after FSRT showed contrast enhancement only in the left sphenoid bone on MRI, which indicated significant decrease in the size of the tumor without any abnormal neurologic deficits. We treated intracranial plasmacytoma which invaded left anterior and middle cranial fossa and surrounded cavernous sinus without cranial nerve deficit through subtotal tumor removal and FSRT.

KEY WORDS : Intracranial plasmacytoma · FSRT.

Introduction

Extramedullary plasmacytoma(EMP) is a rare plasma cell neoplasm of the soft tissue and constitutes about 4% of all plasma cell tumors^{1,2,7,8}. It can be difficult to distinguish EMP histologically from other malignancies or reactive plasma cell proliferation¹¹. In particular, differentiation from multiple myeloma(MM) can be difficult and depends largely on the definition of the disease. However it seems important to recognize EMP, histopathologically characterized by infiltrates of plasma cells of diverse maturity and by their monoclonal immunoglobulin products, as a separate disease entity because of its distinct pattern of evaluation, treatment and prognosis since the standard treatment for localized EMP is the surgery with or without radiotherapy^{5,8,9}. Together with the solitary plasmacytoma of bone(SPB) and MM, they form a group of plasma cell neoplasms belonging to the category of B-cell neoplasm¹⁰. At our hospital, Fractionated Stereotactic Radiotherapy(FSRT) was carried out on the residual tumor after the surgical removal of intracranial plasmacytoma that invaded left orbit and anterior and middle cranial fossa. It revealed good results which will be reported in this paper with its literature review.

Case Report

A 63-year-old male patient visited our hospital complaining left exophthalmus which lasted for a month without any other neurological symptoms. A huge mass lesion was observed in left anterior and middle cranial fossa on magnetic resonance image(MRI). The iso to high signal intensity was shown in T1WI and T2WI with strong gadolinium enhancement. It was suspected as meningioma. Frontotemporal craniotomy and tumor extirpation was performed. The tumor was originated from left anterior fossa, right sphenoid bone, and ethmoid sinus crossing the midline and invaded left orbit, left temporal fossa, anterior wall of the middle cranial fossa and infratemporal fossa. The tumor was highly vascular. EMP was diagnosed since serum albumin was decreased while gamma globulin fraction was increased showing the M-spike in the gamma globulin region on electrophoresis (Fig. 1) although serum protein was within the normal range on the blood test. The CD79a (B cell marker) was shown positive in most plasma cells on the immunohistochemical study (Fig. 2). The neurological changes were not shown after the surgery and FSRT (total 5,100cGy) was followed on the residual tumor that invaded the skull base on follow-up Brain MRI. One-year follow-up after FSRT sh-

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owed the contrast enhancement only limited in the left sphenoid bone area on MRI, which indicated significant decrease in the size of tumor without any radiological recurrence for 3 year follow-up period (Fig. 3).

Discussion

There are several varieties of plasma cell tumors including MM, solitary myeloma of bone, and EMP^{1,5)}. Neurological complications are common in MM. The mechanical effects of local tumor or displaced bone can compress neural structures such as spinal cord. Metabolic disorders such as uremia or hypercalcemia can produce seizures and confusional states. Peripheral neuropathy has been associated with paraproteinemia and amyloidosis. Direct involvement of the central nervous system independent of skull or spine lesions is unusual in MM. Solitary myeloma of bone refers to an isolated osteolytic plasma cell lesion^{2,4)}. A random sample of bone marrow is normal, although an abnormal protein may be present in the serum or urine. These lesions often disseminate and can evolve into MM. Solitary myeloma of bone presenting as a peripheral neuropathy has been well described⁶⁾.

By contrast, EMP is a plasma cell tumor arising in extrasosseous sites⁴⁾. The characteristics of EMP is the plasmacytoid differentiation of B-type lymphocytes originated in bone marrow which move to the peripheral tissue and further develop to the tumor cell¹⁰⁾. Usually it presents in the nasopharynx, upper respiratory tract, gastrointestinal tract, and lymph nodes. The central nervous system is rarely the site of origin. There are no pathognomonic clinical or radiological features of solitary intracranial plasmacytoma, and diagnosis requires the microscopic examination of a

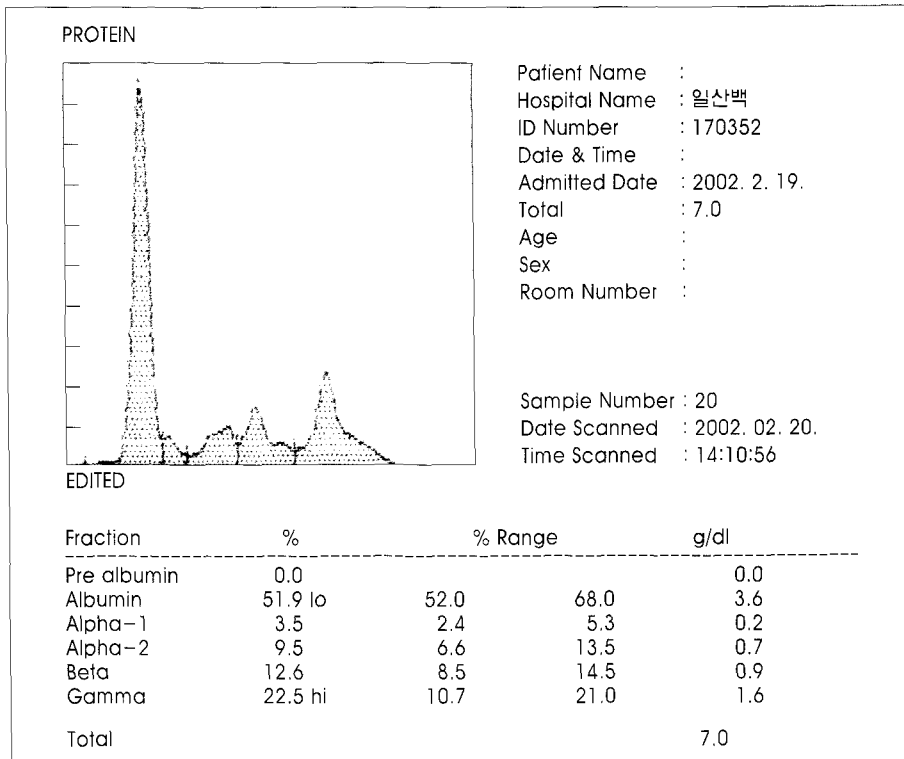


Fig. 1. This electrophoresis report shows that the total serum protein fall under the normal range while the EP pattern indicates the decrease in albumin and the increase in the ratio of gamma globulin fraction. M₁-peak is noted in the gamma globulin region.

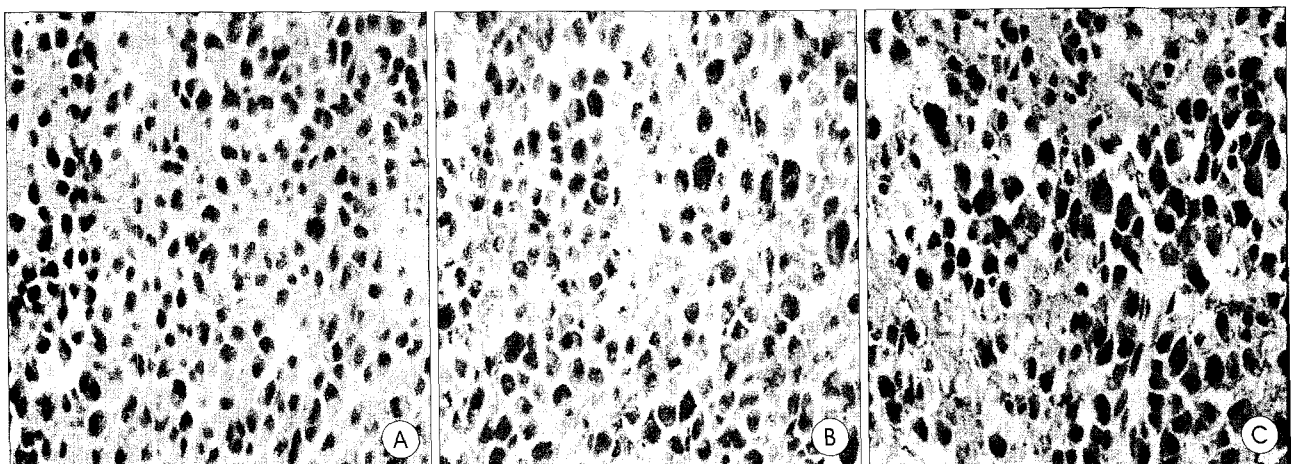


Fig. 2. A : This is light microscopic finding of plasmacytoma. Numerous plasma cells are noted. Some of them have double nuclei (H&E, x 200). B : CD79a (plasma cell marker) is strongly stained in tumor cells immunohistochemically (x200). C : Plasma cells are strongly reactive for Kappa light chain stain (x 200).

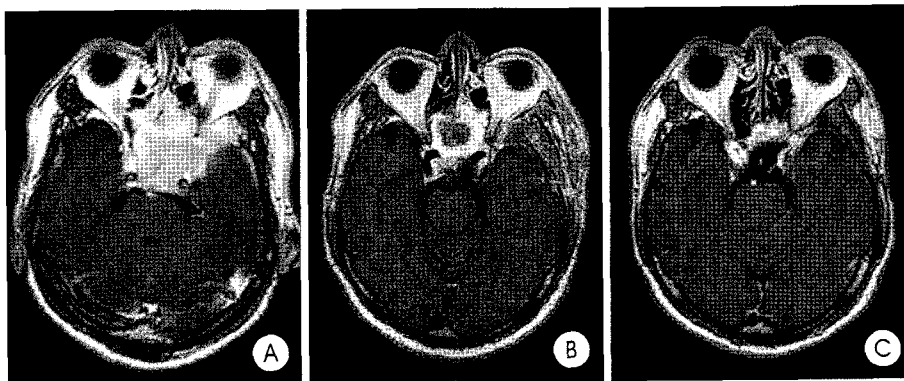


Fig. 3. A : Pre-operative axial T1W gadolinium enhancement magnetic resonance image shows that the part of orbit and sphenoid and estmoid bones are invaded. B : Postoperative axial T1W gadolinium enhancement magnetic resonance image reveals that most of the tumor has been removed, and the residual mass is remained around optic chiasm, estmoid bone, and sphenoid bone. C : The size of mass was more decreased on axial T1W gadolinium enhancement magnetic resonance image after the 3-year follow up since fractionated stereotactic radiotherapy, which also shows no destruction of surrounding structures.

tissue specimen¹⁰. The typical presentation is increased intracranial pressures. The tumors are usually nodular and resemble meningiomas; this is often the preoperative diagnosis. In this case as well, the initial findings based on brain MRI were similar to those of meningioma³. EMP can produce paraproteins locally, but usually insufficient quantities to be demonstrable in serum or urine.

Solitary intracranial plasmacytoma is a rare tumor, and lumbar puncture in the patient with an intracranial mass is risky; routine analysis of CSF immunoglobulins to exclude that lesion is therefore not recommended in the preoperative evaluation of patients with brain tumor. However, serial evaluation of CSF immunoglobulins may be a useful tool in the postoperative management of patients with solitary intracranial plasmacytoma. An elevated or rising monoclonal immunoglobulin fraction of the CSF could suggest the recurrence of tumor.

Plasmacytomas in general are radiosensitive and regress in almost every case after local irradiation³. Thus, unlike MM, solitary EMP is a potentially curable disease. Its treatment has included the surgical removal of the tumor, local radiotherapy, and chemotherapy¹¹. We have treated the solitary intracranial plasmacytoma with surgical excision and radiotherapy. These results suggest that if vital regions of the brain are threatened by tumor, a limited resection or biopsy with subsequent radiotherapy may be successful. Experience with other forms of EMP suggests that local recurrences should be treated vigorously with radiotherapy or excision. Prolonged survival has been documented in patients with locally recurrent EMP⁹. The prognosis in solitary intracranial plasmacytoma is more favorable than in other lymphoreticular tumors of the nervous system. Even in this case, the decrease in the size of tumor was significant after the subtotal removal, but it was more decreased three years later after FSRT (Fig. 2). In addition, considering

that the location of residual mass is around parasella and orbit, good disease course has been shown without any particular neurological disorder. Moreover, it is considered that the absence of a lymphatic system in the brain may limit the potential for extraneural spread⁶.

Conclusion

We could treat intracranial plasmacytoma which invades left anterior and middle cranial fossa and surrounds cavernous sinus without any neurologic deficits through subtotal tumor re-

moval and FSRT and it is still under the examination without radiological regrowth for three years. Therefore, for the treatment of plasmacytoma, it is considered as a good treatment modality to add FSRT after the subtotal resection surgical extirpation the area when it is likely to cause neurological injury with aggressive surgery.

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Commentary

Intracranial plasmacytomas are rare lesions that can arise from the skull, dura, or skull base and exhibit a benign course unless associated with multiple myeloma (MM). MM represents the disseminated form of this disease. Isolated plasmacytomas are classified as either intramedullary or extramedullary on the basis of their association with bone marrow. When plasmacytomas are associated with MM, they show a far worse prognosis. Extracranially, intramedullary plasmacytomas progress to MM more frequently (approximately 50%) than do extramedullary plasmacytomas (approximately 30%). Intracranially, skull and most skull base lesions, being intramedullary, also have greater chance of progressing to MM than do dura-based lesions. The patient with solitary intracranial plasmacytoma should undergo systemic evaluation, including bone

marrow examination, skeletal survey and bone scan to exclude MM. Skull and skull base plasmacytomas are usually highly vascular and have pathological vessels in contrast to extramedullary plasmacytomas that are less vascular. So, I think this case should be a intracranial intramedullary plasmacytoma that have much greater chance to develop MM because this tumor invaded skull base and sinus, and was highly vascular. I think a close follow up should be needed. The ideal treatment is complete surgical resection followed by at least 5000cGy of radiotherapy. However, cure is achievable with conservative subtotal resection and aggressive radiotherapy in this complex skull base lesion because plasmacytomas are very radiosensitive.

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