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Case Report

Solitary Giant Extradural Plasmacytoma

Abstract

Solitary plasmacytoma of the skull is very rare, and only a few cases have been reported in the literature. It remains controversial whether solitary plasmacytoma of the skull is essentially identical with solitary plasmacytoma of bone or not. Solitary plasmacytoma of bone including solitary plasmacytoma of the skull is characterized by a radiologically solitary bone lesion, neoplastic plasma cells in the biopsy specimen, fewer than 5% plasma cells in bone marrow, <2.0 g/dl monoclonal protein in the serum when present and negative urine test for Bence Jones protein (monoclonal light chain). We report one case of a 70-year-old woman who referred to our hospital because of a progressive left parietal swelling. On clinical examination, a painless large soft mass in the right parietal region was observed. Computed tomography revealed an extra-axial mass in the in the left frontoparietal region. The lesion was totally excised despite the bleeding tendency. Histology disclosed the presence of a plasmacytoma. On follow-up examination, 7 months later no tumor recurrence or evidence of multiple myeloma was detected.

Keywords: *Multiple myeloma, plasma cell, plasmacytoma*

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Introduction

Plasmacytes are responsible for the production of antibodies, consisting an important factor of the immune system. Plasmacytomas are referred to benign lesions that may progress to multiple myeloma, a fatal neoplasm.[1] Skull plasmacytomas are unusual tumors accounting for 4% of all plasma cell tumors.[1-6] We report a rare case of a solitary skull plasmacytoma in a 70-year-old patient without neurological deficit, without evidence of multiple myeloma that was successfully treated by surgery alone.

Case Report

A 70-year-old female first noted a painless, soft swelling mass, 10 cm × 10 cm in diameter, in the left parietal [Figure 1] region in January 2014. Neurological examination found no abnormalities. Computed tomography (CT) showed a large extradural mass with heterogeneous enhancement after intravenous administration of contrast material, and bone CT revealed a solitary osteolytic lesion involving the whole layer of the skull [Figure 2]. Laboratory examinations found a red blood cell count of 3.11 × 10 mm^3, hemoglobin 10.3 g/dl, white blood cell count 8100/mm^3, platelets 2.79 × 10^5/mm^3, total serum protein 6.5 g/dl, globulin 2.8 g/dl, gamma-globulin 12.9%, and serum calcium 4.7 mg/dl, which were all within the normal range. Other serum electrolytes were also normal. The immunoelectrophoresis of serum proteins showed immunoglobulins within the normal range. A urine test for Bence Jones protein was negative. Bone marrow aspiration revealed no evidence of systemic myelomatosis. Magnetic resonance imaging (MRI) of the spine detected no additional marrow abnormalities consistent with myeloma. Thus, multiple myeloma and metastatic tumor were denied preoperatively. The tumor extended to the subcutaneous plane and it was extradural was a reddish, soft, and partly elastic mass, highly vascular, and easily separable from the dura matter present in the left parietal region [Figure 3]. The skull defect was sharply demarcated, and there was no osteoplastic reaction along the margin. The marginal bone around the tumor was rongeured out to ensure the complete removal of the tumor. Cranioplasty done using bone cement reinforced with titanium mesh.

Histological diagnosis of the tumor was plasmacytoma. Immunohistochemical staining of neoplastic plasma cells revealed strong and diffuse expression of CD138

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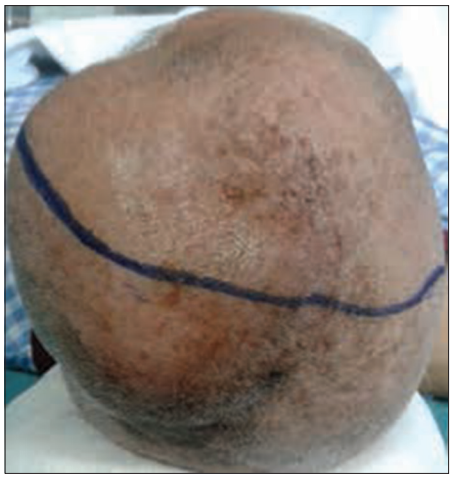

Figure 1: Clinical apperance of skull mass

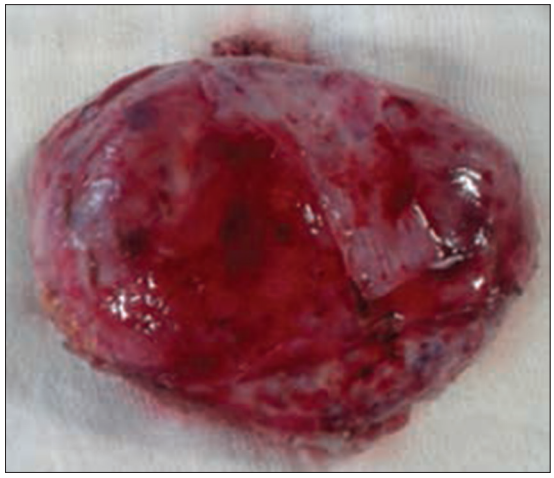

Figure 3: Excised tumor tissue

[Figure 4] and Ig kappa light chain [Figure 5], but not lambda chain. She was discharged without neurological deficit. In the follow-up examination carried out 7 months after the operation [Figure 6], she was doing without development to multiple myeloma.

Discussion

Multiple myeloma has an incidence of 4 cases per 1,00,000/year and constitutes approximately 1% of all malignant neoplasms and 15% of all blood neoplasms.[3] It is characterized by a decrease in the amount of beta lymphocytes and the mean age of the patients is 60 years.[1] Plasmacytoma is a myelomatous mass that may be solitary, in combination with multiple myeloma or may progress to a generalized disease.[7,8] The diagnosis of solitary plasmacytoma can be made only when there is no evidence of multiple myeloma based on bone marrow aspiration, electrophoresis of serum and urine protein, and no other lesion on the complete skeletal survey.[4,9]

Plasmacytoma of the skull is a rare finding.[6] It may involve the cranial vault, skull base, and the orbit. Presenting

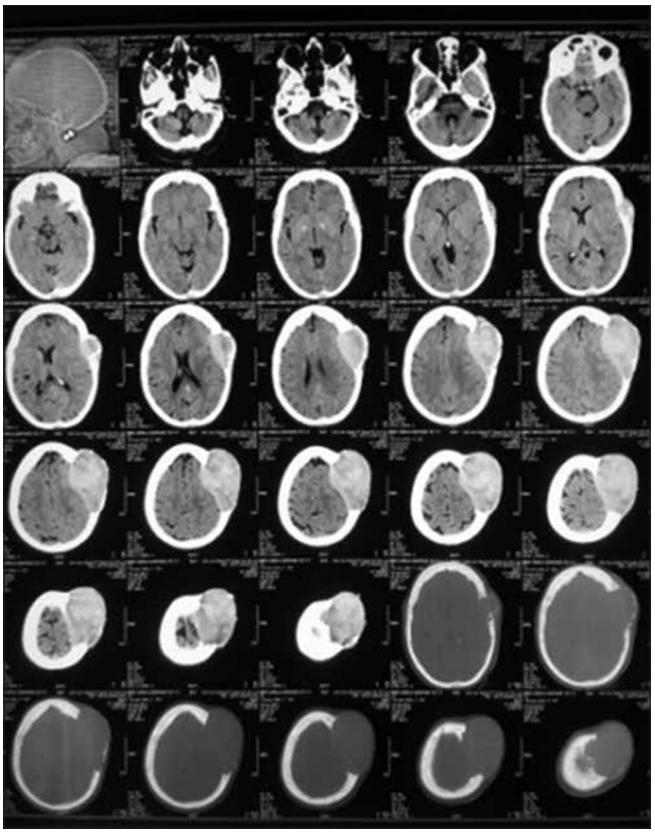

Figure 2: PRE-OP CECT scan brain

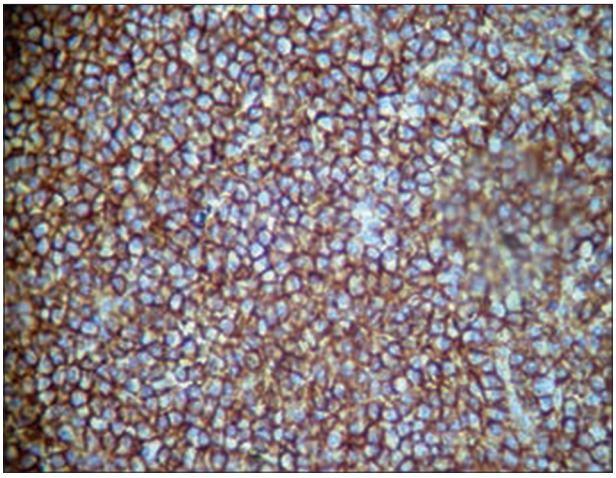

Figure 4: Plasma cells stains diffuse and strongly for CD-138

symptoms and signs are not specific because plasmacytoma lacks neurological symptoms, except of cases of intraparenchymal dissemination or compression of brain and cranial nerves.[8] In that case, symptomatology depends on the lesion's location.[3] Cosmetic skull deformities have been reported to be a usual cause for referring to a specialist.[5] In our case, a problem of cosmetic appearance was evident. On radiological investigation, CT and MRI may provide clues to narrow the differential.[8] Total surgical resection followed by adjunctive radiation therapy has been advocated as an effective treatment in the majority of skull plasmacytomas.[6]

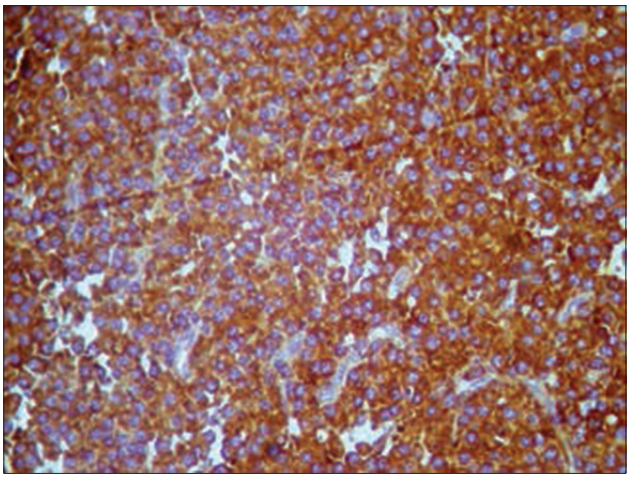

Figure 5: Plasma cell stains diffuse and strongly for kappa Ig chain

Nevertheless, Arienta *et al*. reported that if total resection has been achieved, then radiotherapy should be reserved for the case of tumor recurrence.[2] Furthermore, there are reports of complete cure after biopsy and radiotherapy because plasma cell neoplasms are exquisitely radiosensitive.[10] In our case because of the complete tumoral excision, regular follow-up was preferred reserving radiotherapy for the future. Intraoperative, plasmacytoma may be a highly vascular tumor; therefore, the neurosurgeon should be careful to perform a thorough hemostasis. A case of cardiac arrest from excessive blood loss has been reported.[6] In addition, based on our experience, we recommend craniectomy and cranioplasty because there is a report of high recurrence rate from the remained cells of the tumor to the inner surface of the bone flap.[3] The reconstruction of the cranial vault improved also the cosmetic deformity.

Conclusion

To conclude, plasmacytoma of the cranial vault is a rare clinical entity that demands neurosurgeon's and hematologist's cooperation. Laboratory and imaging evaluation are of paramount importance to exclude systematic involvement. Careful surgical resection if total may be adequate for the disease control. Nevertheless, close follow-up with regular lifelong examinations is important to avoid a generalized incurable disease.

Figure 6: Post of CT scan - bone window

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Conflicts of interest

There are no conflicts of interest.

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