Intracranial tumor behavior of plasma cell neoplasms. Report of two cases and literature review

Ramiro López-Elizalde, Yazmín Lemus-Rodríguez, Marisol Godínez-Rubí, Arturo Madrigal-Saray, José Antonio Muñoz-Serrano, Héctor Velásquez-Santana

Abstract

Background: Multiple myeloma is a plasma cell neoplasm characterized by skeletal destruction, renal failure, anemia and hypercalcemia. Skull plasmacytomas represent <1% of the head and neck tumors. They may be the primary lesion or occur as a secondary manifestation of multiple myeloma in 20-30% of patients or can even manifest several years later after the diagnosis of plasmacytoma. Although some lesions may be surgically accessible, the aggressive natural behavior will complicate the evolution of the patients. We present two cases of Mexican females with intracranial plasmacytomas, one of them associated with multiple myeloma.

Clinical cases: The first case was a 24-year-old female diagnosed with multiple myeloma with plasmacytic-plasmablastic bone infiltration; 90% of disease was surgically removed. The patient presented a local recurrence that required a second intervention for removal. The second case was a 62-year-old female with a malignant intracranial plasmacytoma that was totally resected. Both patients received adjuvant treatment with chemo- and radiotherapy with favorable results. The patients died at 5 and 1.5 years, respectively, due to renal failure secondary to systemic disease.

Conclusions: Chemo- and radiotherapy should be an essential component of the treatment for this condition because the aggressive behavior can complicate the evolution despite being surgically accessible.

Key words: Multiple myeloma, intracranial plasmacytoma, metastasis, skull neoplasm.

Introduction

Intracranial plasmacytoma is a plasma cell tumor described as a solitary myeloma affecting the cranium, meninges and brain. It is a benign lesion that may progress to multiple myeloma with fatal results. Although some plasmacytomas do not meet the clinical criteria for multiple myeloma due to their rapid growth, they are extremely aggressive and change the natural history of the disease. These neoplasms originate from plasma cells that include: a) multiple myeloma and plasmacytoma; b) Waldenstrom macroglobulinemia and monoclonal gammapathies; c) chronic lymphocytic leukemia. The incidence of multiple myeloma is 4/100,000 of the population. It is more common in males >60 years of age with associated genetic racial and geographical factors. Deaths are due to recurrent infections and renal failure, with a survival of 2 to 3 years. Diagnosis is made according to the following criteria: 1) histological evidence of plasmacytoma or bone marrow infiltration; 2) arthralgia, anemia, or renal failure; 3) serum gammapathy or osteolytic lesions; 4) anemia, which is found in 85% of the patients with multiple myeloma. Plasmacytomas represent 5-10% of plasma cell neoplasms. They are classified according to solitary bone plasmacytoma and extramedullary plasmacytoma. In the first case, long bones are most commonly affected, whereas in the latter they are located in the mucosas, head and neck in 80-90% of cases. Most cases of solitary extramedullary plasmacytoma are located in the upper respiratory tract, lamina of the GI tract and soft tissue with free air. Intracranial plasmacytomas are rare; however, in some cases of intrasellar plasmacytoma and the clivus have been reported in the literature.

Solitary plasmacytosis of the cranium must be differentiated from myelomatosis. It can manifest as an unusual sign...
in patients with multiple myeloma as reported in a case by Nassar. Solitary plasmacytoma of the cranium generally has a good prognosis when complete surgical removal is accomplished, in which case radiotherapy may not be required.

The frequent evolution of plasmacytoma to multiple myeloma occurs in 58% of solitary bone plasmacytoma and from 20 to 30% for extramedullary plasmacytoma. Ten-year follow-up shows a survival of 16 and 70%, respectively.

This report demonstrates our experience in the treatment of two patients with intracranial bone lesions with histopathological diagnosis of plasma cell lesion.

**Clinical Cases**

**Case 1**

We present the case of a 24-year-old female with a pregnancy of 33 weeks gestation. Two weeks prior to her pregnancy, the patient noticed a small right frontal tumor, which gradually increased in size. During the last 2 months of her pregnancy she reported frontal headaches accompanied by vomiting and tonic-clonic seizures. For this reason, brain computed tomography was carried out. Two tumors were found: one 3-cm in diameter in the right frontal region with bone destruction and the other of 5 x 3 cm in diameter in the left frontotemporal region (Figure 1). According to laboratory studies, Bence-Jonce proteins were negative, negative serum protein electrophoresis, and serum calcium concentrations and blood count were both normal. The pregnancy terminated with a cesarean section without complications. Two months later, the patient underwent resection of 90% of the frontal tumor, and bone and meninges infiltrations were found. Following this, a cerebrospinal fluid leak appeared and was treated conservatively 1 week later with a satisfactory resolution. Because she had a local frontal recurrence (Figure 2), she was operated again to remove the lesion.

Histopathological results reported multiple myeloma with bone infiltration of the plasmacytic-plasmoblastic type (Figure 3). The patient underwent bone marrow aspirate, which reported the absence of plasma cell infiltration. The patient was not operated for the second initial cranial bone lesion and only received additional radiotherapy. Recurrence of the lesion was demonstrated at 45 days. For this reason the patient underwent an additional surgical intervention and received adjuvant therapy consisting of chemo- and radiotherapy. She remained disease-free for 5 years. However, kidney failure then developed, causing her death.

**Case 2**

We report the case of a 62-year-old female who was admitted because of a left frontoorbital lesion with 6 months evo-
Intracranial tumor behavior of plasma cell neoplasms

Figure 3. Case 1. Plasma cell neoplasm of predominantly plasmacytic type with polygonal cells with abundant cytoplasm and eosinophilia, eccentric nucleus, round or ovoid hyperchromatic (H&E: 10x).

Figure 4. Case 2. Brain magnetic resonance in sagittal section with left frontoorbital tumor with mainly solid appearance and containing cystic areas. The tumor affected the roof of the left orbit, with compression and displacement of the ipsilateral frontal lobe.

Figure 5. Case 2. Angiography showing the nutrition of the predominant tumor through branches of the superficial temporal artery, with venous drainage to the facial vein and the superior sagittal sinus.

Evolution, affecting bone and intra- and extracranial structures, which remained asymptomatic. Laboratory studies showed hypercalcemia and imaging revealed a large frontal tumor that destroyed the calvarium with a mostly solid aspect and contained cystic areas (Figure 4). The tumor also affected the roof of the left orbit with compression and displacement of the ipsilateral frontal lobe. The lesion was highly vascularized and affected the intra- and extracranial structures. Angiographic study showed in detail the great vascularity of the lesion, predominantly nourishing itself with branches of the superficial temporal artery with venous drainage to the facial vein and the superior longitudinal sinus. The anterior and middle left cerebral arteries were observed to be clearly thinned, perhaps due to the steal phenomenon through the external carotid artery (Figure 5). Bone scan revealed increased radioactive uptake in the left fronto-parieto-temporal region, in addition to the sternum, ribs and thoracolumbar spine.

The patient underwent surgery and was operated for left superficial temporal artery ligation, with partial resection of the lesion. The procedure was discontinued due to bleeding; therefore, the surgery was repeated 1 week later to complete resection of the lesion, which was achieved successfully from a macroscopic point of view. The patient was discharged without complications 2 weeks postoperatively. Histopathological results reported a neoplastic plasma cell tumor (Figure 6). The patient underwent adjuvant chemotherotherapy. After an 18-month follow-up, there was no evidence of recurrence of a cranial lesion. However, the patient died as a result of kidney complications due to systemic disease.
Discussion

From a histopathological point of view, it is very important to determine whether the parent cell line is attributed to plasmacytic or plasmoblastic cells because of the marked differences related to prognosis and treatment. The tendency to be more aggressive with the dose of radiotherapy in the plasmablastic cell pattern was reported by Schwartz et al.\textsuperscript{11} who suggested a dose of at least 5000 cGy after surgical resection.

Anatomopathological findings in both patients were consistent with multiple myeloma and bone infiltration; therefore, adjuvant treatment with chemo- and radiotherapy was carried out. The postoperative course in each of the cases was different despite the similar pattern of marrow involvement, with important tumor-like lesions with intra- and extracranial extension and a rapid growth associated with clinical evidence of intracranial hypertension.

Complete surgical treatment of lesions with adjuvant chemo- and radiotherapy resulted in being a good choice in both patients because there was no evidence of disease recurrence. This is justified by comparing the first conservative treatment with radiotherapy alone in Case 1, which was not optimal compared with the second intervention with radio- and chemotherapy where there was no recurrence. In Case 2 where the patient was already diagnosed with multiple myeloma and despite combined treatment, we failed to increase survival as in Case 1 because the natural evolution of multiple myeloma continued with systemic failure and death.

In conclusion, the natural history of multiple myeloma is progressive deterioration with damage to various organs, ultimately leading to death. This intracranial disease is rare but extremely quick and aggressive, prompting the neurosurgeon to act quickly and to conduct an oncological resection. All patients should be given adjuvant treatment with chemo- and radiotherapy. This combination was shown to be effective for intracranial lesions in both cases; therefore, comprehensive treatment for the disease is required. For these reasons we consider it to be important to report both cases, as well as the patients’ response to treatment.

References