INTRACRANIAL SOLITARY PLASMACYTOMA: CASE REPORT

Plasmocitoma solitario intracraneal: Reporte de caso

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ABSTRACT

Introduction: Solitary intracranial plasmacytoma is a rare plasma cell tumor that affects the skull, meninges, and brain. Unlike multiple myeloma, it does not have systemic manifestations. Its symptoms are characterized by a progressively growing tumor that generates local pain. It does not have a pathognomonic image and can be confused with other lesions, being its diagnosis histological. The prognosis is good and the main treatment modalities are surgery and/or radiation therapy.
Clinical Case: 64-year-old female patient with a clinical picture characterized by loss of strength in lower limbs and thermal rise. On examination: Patient awake, Glasgow 15 points, paraparesis 4/5, photoreactive and isochoric pupils. Imaging examinations show an extensive contrast-enhancing lesion involving cerebral meninges at the frontoparietal level bilaterally, with a moderate mass effect. A subtotal resection of the lesion is performed, with the patient presenting a good clinical evolution. The histological result was a plasma cell tumor. In the 6-month follow-up, no residual lesion was observed, maintaining outpatient control by an outpatient clinic. This case shows a rare pathology that is sometimes confused with a meningioma.
Conclusion: Solitary intracranial plasmacytoma is a rare tumor that can easily be confused with other more common lesions. Its diagnosis is only made with a histological study. It has a good prognosis and can be treated by surgical resection and/or radiotherapy.

Keywords: *Plasmacytoma, Brain, Meninges, Neoplasms, Plasma Cell (Source: MeSH NLM)*

RESUMEN

Introducción: El plasmocitoma solitario intracraneal es un tumor raro de células plasmáticas que afecta el cráneo, las meninges y el cerebro. A diferencia del mieloma múltiple no tiene manifestaciones sistémicas. Sus síntomas se caracterizan por una tumoración de crecimiento progresivo que genera dolor local. No tiene una imagen patognomónica pudiendo confundirse con otras lesiones siendo su diagnóstico, histológico. El pronóstico es bueno y las modalidades principales de tratamiento son la cirugía y/o radioterapia.
Caso Clínico: Paciente mujer de 64 años con cuadro clínico caracterizado por perdida de fuerza en miembros inferiores y alza térmica. Al examen: Paciente despierta, Glasgow 15 puntos, paraparesia 4/5, pupilas fotorreactivas e isocóricas. Los exámenes de imágenes muestran una lesión extensa captadora de contraste que compromete meninges cerebrales a nivel fronto-parietal de forma bilateral, con moderado efecto de masa. Se realiza una resección subtotal de la lesión presentando el paciente una buena evolución clínica. El resultado histológico fue tumor de células plasmáticas. En el seguimiento a los 6 meses no se observa lesión residual, manteniéndose el control ambulatorio por consultorio externo. Este caso muestra una patología poco frecuente que en ocasiones es confundido con un meningioma.
Conclusión: El plasmocitoma solitario intracraneal es un tumor infrecuente que puede ser confundido fácilmente con otras lesiones más comunes. Su diagnóstico solo se realiza con estudio histológico. Tiene buen pronóstico, pudiendo ser tratado mediante resección quirúrgica y/o radioterapia.

Palabras clave: Plasmocitoma, Encéfalo, Meninges, Neoplasia de Células Plasmáticas *(Fuente: DeCS Bireme)*

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Solitary intracranial plasmacytoma is a rare plasma cell tumor that affects the skull, meninges, and brain. It represents between 5-10% of the neoplasms originating from these cells and unlike the Multiple myeloma develops in isolation without systemic manifestations.1,2,3 affecting mainly males with a 4: 1 ratio, between 50 and 60 years.4 The incidence is 3 per 100,000 people per year and it is not known a predisposing factor.5

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These tumors can appear in any place where there are formations of the reticuloendothelial system, being the rarest locations at the level of the head and neck. Solitary plasmacytomas of cranial and cervical locations are characterized by clinical manifestations such as local pain and a progressively growing tumor.[1,3,6]

The radiographic appearance of solitary cranioencephalic plasmacytoma can mimic that of the more common neoplasms. In the computed tomography the appearance is of a rounded or lobulated, expansive, lytic lesion, with diploic, homogeneous, hyperdense participation that reinforces the contrast. [3,7-9] 10-20% of all plasmacytomas are multicentric, the homogeneous characteristics of punch lesions differentiate them from metastases.[6]

The diagnosis of solitary plasmacytoma is histological. The differential diagnosis should be made, mainly, excluding the presence of multiple myeloma, infiltrating plasma cell leukemia, and other lymphoproliferative processes; However, at the radiological level, other lesions that present osteolytic reaction such as meningioma, osteosarcoma, hemangioma, and metastasis should be excluded.[7]

The prognosis is good considering a benign lesion, but it can progress to multiple myeloma, a malignant and often fatal neoplasm.[1] This progression occurs in 50% of patients diagnosed with solitary plasmacytomas between three and five years after diagnosis. For this reason, these patients, despite having had a good response to treatment with surgery and/or radiotherapy, must have a close follow-up for a long time since relapses are known up to 28 years after treatment.[10,11,12] We present the case of a patient with cranial plasmacytoma operated in our Hospital.

CLINICAL CASE

History and examination: 64-year-old female patient with a history of frontal intracranial tumor diagnosed as meningioma. A month before she was admitted to the emergency room, she presented a loss of strength in the lower limbs (LL) that increased, making it difficult for her to walk. She was admitted to the emergency room due to a feverish condition added to the motor deficit of LL.

On physical examination: Patient awake, disoriented, Glasgow Coma Scale of 14 points, paraparesis of 4- / 5, photoreactive and isochoric pupils, Karnofsky scale of 70. A soft tumor was palpated in the frontal region. Imaging examinations (CT and MRI) showed a bilateral tumor at the level of the frontal and parietal convexity that occupies the superior longitudinal sinus, with a mass effect, which captures contrast in a homogeneous way and presents perilesional edema that collapses both lateral ventricles. The diagnosis of atypical meningioma was raised. An osteolytic reaction was also observed in the bifrontal skullcap. *(Fig. 1)*

Treatment: A cerebral angiography was performed which demonstrated complete closure of the superior longitudinal sinus. During her hospitalization, she was diagnosed with diabetes mellitus which was controlled with insulin, and urinary infection due to Proteus mirabilis which was successfully treated with Ertapenem.

He was scheduled for elective surgery with a frontoparietal craniotomy. During surgery, infiltration of the cranial shell was found, which was bulging and translucent. Due to bone involvement, it was decided to perform a frontoparietal craniectomy. The lesion found was brown in color with a mamelon appearance with hemorrhagic and necrotic areas that compromised the meninges, with the largest volume of the lesion being in the epicranial space. A subtotal resection of the lesion was performed, leaving a small fragment that was closely adhered to the brain parenchyma and a cortical vein. Infiltration was observed in the superior longitudinal sinus that extended to the brain parenchyma, which was carefully dissected. There were no complications, and a Duroplasty with galea and a cranioplasty with acrylic cement was finally performed. *(Fig. 2)*

Clinical evolution: In the immediate postoperative period the patient evolved favorably, she was extubated and transferred to intensive care unit, awake, in Glasgow 14, with a slight motor deficit in LL, with photoreactive and isochoric pupils, operative wound covered with dry dressings. The clinical evolution was favorable, being transferred to intermediate care unit, and then to general care. Immediate postoperative non-contrast brain CT showed resection of the lesion without bleeding in the operative bed. *(Fig. 3)* A brain CT with contrast prior to discharge showed a subtotal resection, leaving only small contrast-capturing remains. On postoperative day 10 with improvement in lower limb strength, discharge and follow-up on an outpatient basis were decided.

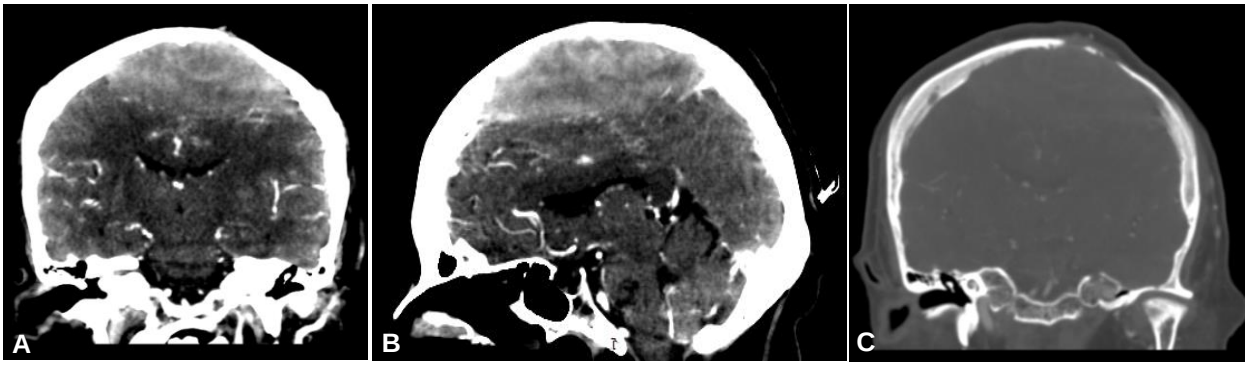

Fig 1. Brain tomography (CT) showing **(A, B, C)** a bilateral lesion at the frontoparietal level, a contrast-binding agent that erodes the cranial calvaria and causes a mass effect that collapses lateral ventricles.

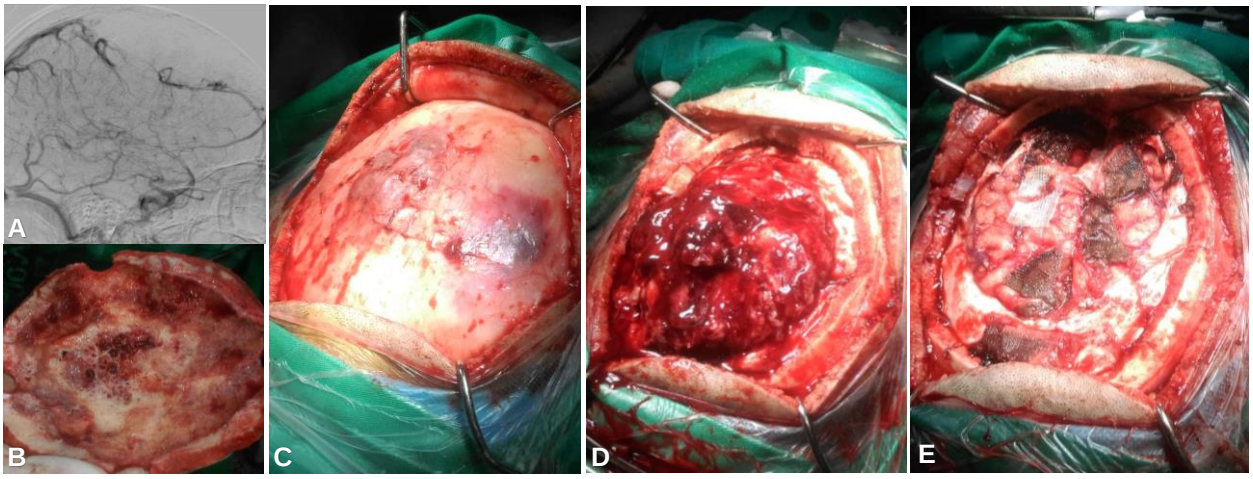

Fig 2. (A) Cerebral angiography showing the absence of the superior sagittal sinus at the level of the lesion. **(B, C, D, E)** Intraoperative images showing bone involvement, lesion characteristics, and the operative bed at the end of resection.

In the outpatient control, the patient had a Glasgow Scale of 15, without motor deficit, with good healing of the operative wound, and a Karnofsky Scale of 90. The pathological study was reported as plasma cell dyscrasia with compromised bone tissue compatible with plasmacytoma or multiple myeloma. The diagnosis of solitary intracranial plasmacytoma was concluded as there were no laboratory findings or images that suggest a systemic involvement, as in the case of multiple myeloma.

DISCUSSION

Plasmacytomas are infrequent tumors, and their intracranial location is even rarer, so this case is relevant, as has been described in other publications.[1,3,12,13,14] In the case reports they are described as lesions that are initially confused with other more frequent pathologies such as meningiomas; This coincides with our case since we were only able to make the precise diagnosis with the pathological anatomy.

Plasmacytoma affects mainly the male sex [1,3,12], which is reflected in most publications, differing from our case as it is a female patient. The age of presentation of our patient coincides with that reported in the literature since most of them present between the fifth and sixth decade of life [3,4]. The clinical manifestations in our patient were characterized mainly by a motor deficit (paraparesis), which is a presentation that is not observed in most of the patients reported in other series, [1,3,12-14] where the symptoms are local pain or increased blood pressure. progressive volume in the affected region.

In cases of solitary lesion not associated with multiple myeloma, as is our case, complete surgical resection and subsequent radiotherapy offer a good long-term prognosis. Although there are also reports where it is stated that being a radiosensitive tumor, radiotherapy is the first line [14]. Our case required surgery because the patient presented compromised motor function due to the mass effect of the tumor and required decompression together with the maximum possible resection. During the intraoperative period, we could observe that macroscopically it did not have the appearance of a meningioma, so at that time we suspected other possible diagnoses, which led us to make a maximum possible resection, leaving only a fragment closely adhered to a cortical vein.

The motor deficit could be explained by the great mass effect of the lesion that, in addition to compromising the superior sagittal sinus, also produced the collapse of the lateral ventricles. As well described, these lesions are initially confused with meningioma-like tumors [3,12] which also occurred in our case. Imaging examinations are not conclusive since, being contrast-enhancing lesions, involving the meninges and adjacent bone, they resemble other more common lesions. This makes it difficult to make an accurate diagnosis in the preoperative period, having to wait for the result of the pathological anatomy.

Patients with plasmacytoma, despite having had a good response to treatment, either with surgery and/or radiotherapy, should have a close follow-up for a long time due to recurrences in a high percentage [16]. In our case, at six months after having operated, no size increase in the residual lesion has been shown, even this lesion is not appreciated in the current tomography and the patient remains with good clinical evolution in the outpatient controls. We believe that surgical management is important especially in injuries with a large mass effect that generates neurological compromise. Our patient will continue to be followed by Neurosurgery and by the medical Oncology Department.

CONCLUSION

Solitary intracranial plasmacytomas are rare tumors whose radiological image is uncharacteristic, giving rise to erroneous interpretations, classifying it as a meningioma in most cases. The definitive diagnosis of plasmacytoma always requires a pathological study as it does not have its own clinical or imaging characteristics.

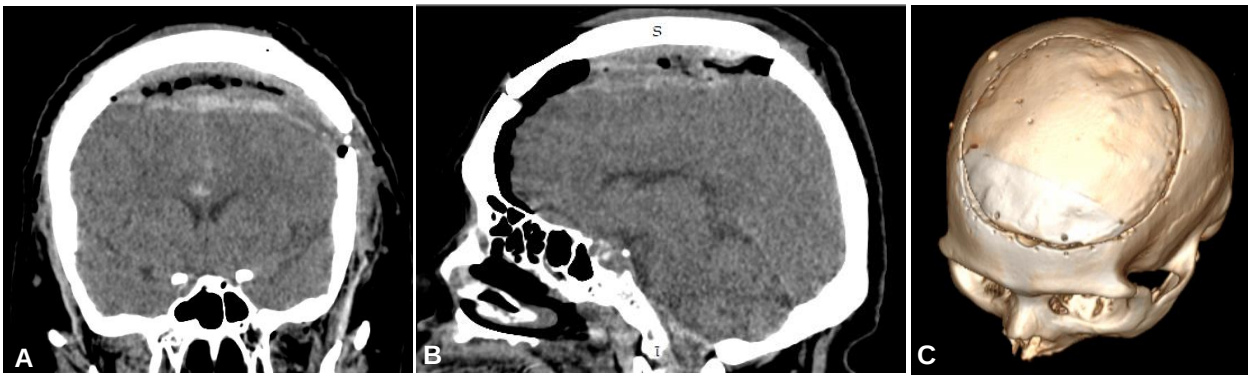

Fig 3. Post-surgical brain tomography showing **(A, B)** resection of the meningeal lesion without evidence of bleeding in the operative bed. **(C)** The wide craniectomy is appreciated, which required a cranioplasty with an acrylic cement mold.

A plasmacytoma should be suspected as it has an expansive intracranial mass, osteolytic, homogeneous, hyperdense, and with enhancement to the contrast medium. Patients with plasmacytoma have a good response to treatment with surgery and/or radiotherapy but should be closely followed due to the possibility of recurrence or progression to multiple myeloma.

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Disclosures
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