

Neurosurgical Management of a Giant Parasagittal Meningioma in a Patient with Neurofibromatosis Type 2: A Case Report

Citar como:

Aguilar-Zegarra L, Váscones-Román FF, Caucha Y. Neurosurgical Management of a Giant Parasagittal Meningioma in a Patient with Neurofibromatosis Type 2: A Case Report. Rev Méd Hered. 2025; 36(3): 247-251. DOI: 10.20453/rmh.v36i3.5841

Recibido: 02/10/2024

Aceptado: 26/06/2025

Declaration of financing and conflicts of interest:

The authors declare that there were no external sources of funding, and they declare that they have no financial relationships with organizations that may have an interest in the submitted article.

Authorship contribution:

LA, FFVR: develop the idea, write the case, and approve the final manuscript. **YCM:** reviewed and approved the final manuscript. The authors share full responsibility for all aspects presented in this clinical case report.

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Manejo neuroquirúrgico de un meningioma parasagital gigante en una paciente con neurofibromatosis tipo 2: reporte de caso

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SUMMARY

Parasagittal meningiomas are intracranial tumors that develop along the superior sagittal sinus. Although they are generally benign, their proximity to vital vascular structures poses significant challenges for surgical management. In patients with neurofibromatosis type 2 (NF2), an autosomal dominant genetic disorder, there is a high predisposition to develop multiple tumors in the central nervous system, including meningiomas and schwannomas. The report discusses a 23-year-old woman with a family history of NF2 who presented with a large parasagittal meningioma and bilateral schwannomas, along with symptoms such as headaches, hearing loss, vertigo, and seizures. MRI revealed a right frontoparietal meningioma invading the superior sagittal sinus. Successful craniotomy and resection showed a poorly vascularized meningioma invading the sagittal sinus. This case highlights the surgical difficulties associated with sagittal sinus meningioma invasion and emphasizes the importance of multidisciplinary care and long-term follow-up due to the high risk of tumor recurrence in NF2 patients.

KEYWORDS: Neurofibromatosis 2, Meningioma, Brain neoplasms.

RESUMEN

Los meningiomas parasagitales son tumores intracraneales que se desarrollan a lo largo del seno sagital superior, y aunque generalmente son benignos, su proximidad a importantes estructuras vasculares presenta desafíos significativos para el manejo quirúrgico. En pacientes con neurofibromatosis tipo 2 (NF2), una enfermedad

genética autosómica dominante, existe una alta predisposición a desarrollar múltiples tumores en el sistema nervioso central, incluidos meningiomas y schwannomas. Se presentó el caso de una mujer de 23 años con antecedentes familiares de NF2 quien presentó un meningioma parasagital gigante y schwannomas bilaterales, experimentando dolores de cabeza, pérdida de audición, vértigo y convulsiones. La resonancia magnética mostró la presencia de un meningioma frontoparietal derecho que invadía el seno sagital superior. La craneotomía y resección exitosa revelaron un meningioma poco vascularizado con invasión del seno sagital. Este caso subraya los desafíos quirúrgicos de la invasión del meningioma en el seno sagital, enfatizando la necesidad de atención multidisciplinaria y seguimiento a largo plazo debido al alto riesgo de recurrencia del tumor en pacientes con NF2.

PALABRAS CLAVE: Neurofibromatosis 2, meningioma, neuroma acústico.

INTRODUCTION

Parasagittal meningiomas are common intracranial tumors, responsible for 20-30% of intracranial meningiomas, and develop along the superior sagittal sinus, a key venous structure for brain drainage⁽¹⁾. Although most meningiomas are benign, their growth can cause significant brain compression, manifesting clinically with headaches, seizures, and severe motor deficits. Due to their proximity to vital brain structures, the surgical treatment of these tumors is complex, with a high risk of postoperative complications such as thrombosis or hemorrhage, in addition to a considerable risk of tumor recurrence^(2,3). In patients with type 2 neurofibromatosis (NF2), an autosomal dominant genetic disease, the predisposition to developing meningiomas is significantly higher, with approximately 50-70% of these patients presenting with multiple intracranial tumors, including parasagittal meningiomas and bilateral vestibular schwannomas^(4,5). The mutation in the NF2 gene causes the loss of function of the Merlin protein, which is key in tumor suppression, and favors the formation of tumors in the central nervous system.⁽⁴⁾

The objective of this report was to describe the clinical and surgical management of a complex case of giant parasagittal meningioma in a young patient with NF2, highlighting the challenges associated with its proximity to the superior sagittal sinus and the coexistence of multiple schwannomas.

CLINICAL CASE

A 23-year-old woman presented to the hospital with severe headaches, tinnitus, and nausea. The patient reported an onset of symptoms approximately two years ago, characterized by progressive headache,

hearing loss, vertigo, visual acuity reduction, and generalized tonic seizures, with an increase in the frequency and intensity of these episodes in the last six months. The patient has a family history of type 2 neurofibromatosis. Upon initial physical examination, the patient was alert, attentive, and oriented in time, space, and person. She exhibited decreased muscle strength in the left lower extremity, with conserved muscle tone and tropism. Deep tendon reflexes were heightened (+++), with an indifferent left plantar reflex. Decreased sensitivity in the extremities and a motor deficit affecting balance and coordination were observed. No meningeal signs or neck stiffness were detected. Cranial nerve evaluation revealed amaurosis in the left eye with an afferent pupillary defect, severe hearing loss predominantly in the left ear, bilateral tinnitus, and ophthalmoparesis affecting cranial nerves III and VI in the left eye. Higher cognitive functions remained intact.

Magnetic resonance imaging revealed a giant right fronto-parietal parasagittal meningioma with lobulated edges (Figure 1). A hyper-enhancing lesion was also identified in the right choroid plexus, a solid nodular image in the left occipital scalp, and bilateral schwannomas. Based on these findings, the patient was hospitalized for neurosurgical management. During hospitalization, the patient's condition remained stable. A right fronto-parietal craniotomy and tumor resection were performed, revealing a pearly white, fibrous, and poorly vascularized meningioma (Figure 2). A second extra-axial lesion invading the superior sagittal sinus, friable, gray-violet, and also poorly vascularized, was identified. Pathological examination of the tumor was performed, revealing findings consistent with a meningioma, including the presence of psammoma bodies (Figure 3).

The patient was discharged with pharmacological treatment, including acetaminophen for pain, phenytoin as an anticonvulsant prophylaxis, and dimenhydrinate to control nausea. A neurosurgical

follow-up was scheduled for two weeks to evaluate the postoperative status and adjust therapy in case of recurrence.

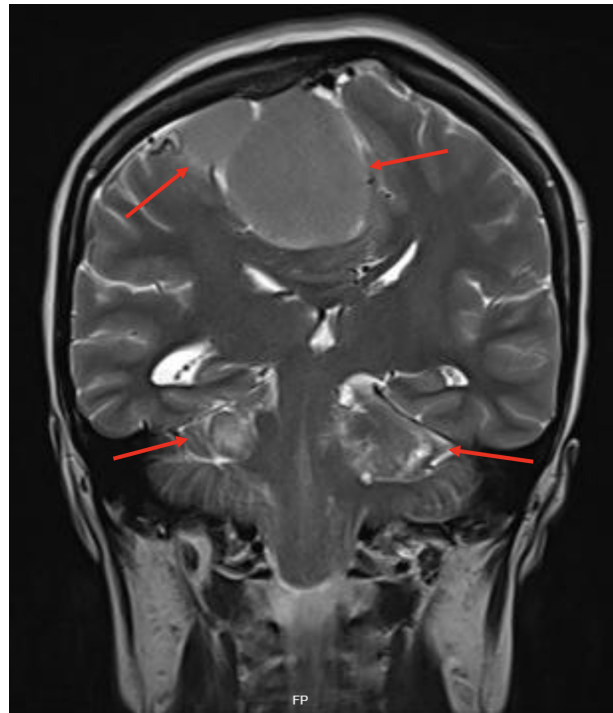


Figure 1. T2-weighted magnetic resonance imaging, coronal section shows an extra-axial expansive lesion in the fronto-parietal region, involving the superior sagittal sinus, with hyperintense signal. The lesion compresses adjacent brain gyri without evident signs of perilesional edema.



Figure 2. Intraoperative image of the surgical specimens shows a set of extracted tumor lesions, highlighting a large, lobulated tumor with a pearly and fibrous appearance, with poor vascularization on its surface, measuring approximately 8 cm. Accompanied by two smaller lobulated masses, also with similar characteristics. The lesions were completely excised and correspond to a giant parasagittal meningioma, according to the previous clinical and radiological context.

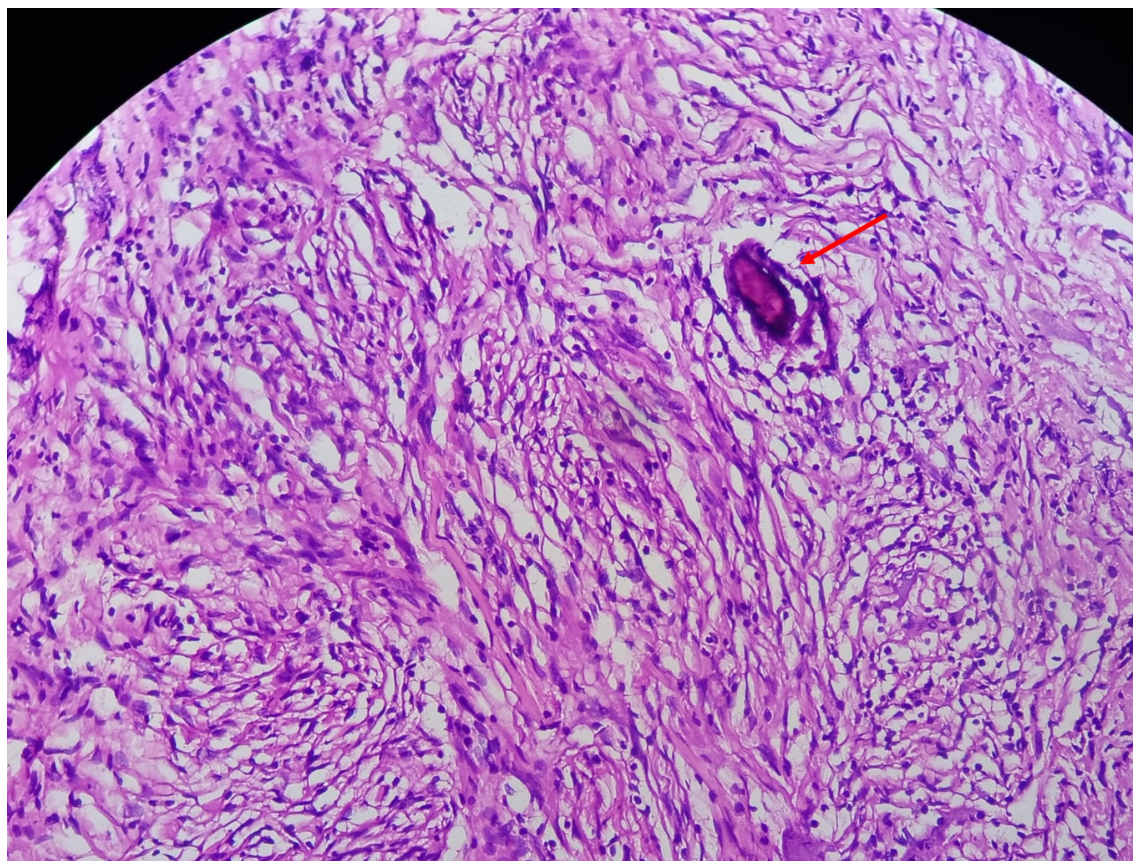


Figure 3. Histopathological examination identifies the presence of a psammoma body within the tumor tissue. This structure appears as a concentrically laminated, round calcified body—commonly associated with meningiomas.

DISCUSSION

This case highlights the complex interplay between two rare neurological conditions: giant parasagittal meningioma and type 2 neurofibromatosis (NF2). The simultaneous presence of meningiomas and schwannomas in patients with NF2 poses significant diagnostic and therapeutic challenges. NF2 is associated with a genetic predisposition to develop multiple tumors of the central nervous system due to mutations in the NF2 gene, which affect the function of Merlin protein, an essential tumor suppressor that regulates cell growth⁽⁵⁾. Between 50% and 70% of NF2 patients develop meningiomas, with a notable prevalence in parasagittal locations, where surgical management is particularly complex due to the proximity to the superior sagittal sinus.^(4,5)

The tumor invasion of the superior sagittal sinus in parasagittal meningiomas is a critical factor that increases the risk of intraoperative complications, such as hemorrhage and sinus thrombosis⁽⁶⁾. Despite advances in surgical techniques, complete resection of

these tumors remains a significant challenge. Subtotal resection may be necessary in some cases to preserve neurological function or due to sinus invasion. In this case, the invasion of the superior sagittal sinus posed a challenge, as the high vascularization of this area increases the risk of severe complications. A recent study suggests that subtotal resection followed by radiotherapy could be a viable strategy to achieve long-term tumor control while minimizing postoperative complications.⁽⁷⁾

The patient's diagnosis was complicated by the presence of multiple tumors and a progressive clinical picture that included hearing loss, vertigo, and generalized seizures, all typical manifestations of bilateral vestibular schwannomas. Magnetic resonance imaging was crucial in identifying the extent of the tumors, allowing for appropriate surgical planning. MRI is undoubtedly the gold standard for evaluating these tumors, providing detailed information about their location, size, and morphological characteristics, essential for precise surgical planning.⁽⁸⁾

Additionally, it has been reported that NF2 patients often require repeated neurosurgical interventions due to the multifocal nature of the disease and the high risk of tumor recurrence. Indeed, between 20% and 30% of meningiomas in NF2 patients recur after surgical resection⁽³⁾, reinforcing the need for long-term follow-up and intensive surveillance with periodic neuroimaging. Larger tumors (greater than 25 mm) and multiple tumors are associated with a worse prognosis. Moreover, the presence of vestibular schwannomas also complicates clinical management, significantly affecting the patient's quality of life due to progressive hearing loss and balance issues that limit daily functionality.⁽¹⁰⁾

In terms of long-term management, a multidisciplinary approach is essential for patients with NF2. Neurosurgical intervention must be accompanied by regular follow-up in collaboration with neuro-oncologists, geneticists, and neurologists to monitor both tumor recurrence and the development of new tumors associated with NF2. Genetic counseling also plays a key role in managing these patients, allowing for early detection in first-degree relatives and a better understanding of the disease's progression.⁽⁹⁾

This case underscores not only the importance of careful surgical intervention but also the need for long-term management that includes continuous radiological surveillance and a comprehensive approach to minimize complications and improve the quality of life in patients with NF2.

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