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Case report: Nasal spindle cell sarcoma. Review and management

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Clinical case

A 32-year-old female patient, with no significant medical history, was admitted to the Otolaryngology Department of the Arzobispo Loayza National Hospital with a four-month history of progressive left nasal obstruction and anterior rhinorrhea on the same side. She also developed progressive left proptosis and, one month prior, presented with left amaurosis. She then sought consultation with the Head and Neck Surgery Department, where a contrast-enhanced CT scan of the facial skeleton was performed.

where a large expansive tumor (54×33×43mm) is visualized occupying the left nasal fossa with extension to the nasopharynx, with lobulated margins, with involvement of the left orbit and left sphenoid sinus with extension to the lateral sinus with displacement of the left carotid artery, for which reason an MRI of the Facial and Cerebral Mass is requested (Figure 1) where it is observed that the lesion partially occupies the left intraorbital space in its inferomedial margin, producing contralateral displacement and elevation of the extraocular muscles, causing erosion and destruction of the medial and inferior wall orbital, at the level of the left sphenoid sinus it contacts the anterior portion of the clivus as well as erosion of the surface and lateral aspect of the sellar region producing compression of the optic chiasm and the intracranial portion of the left optic nerve, it also presents involvement of the lamina cribrosa.

A biopsy was performed in the office, and the pathology report showed spindle cell stroma with atypia, with inconclusive immunohistochemistry. Therefore, a surgical approach was decided upon. The patient underwent endoscopic surgical treatment, with tumor resection via endoscopic maxillectomy, ethmoidectomy, Draf IIB frontal sinusotomy, and wide sphenoidotomy. A fibroelastic-appearing tumor with hemorrhagic areas and a hard, bone-like capsule was successfully removed.

No cerebrospinal fluid leakage was observed in the dura mater, so it was decided to cover the surgical bed with Surgicel and Gelfoam. The sample was sent to pathology with the result: spindle cell sarcoma compatible with malignant tumor of the peripheral nerve sheath, with immunohistochemistry positive for CD56 and focally positive for SOX10 and CD99, with a Ki67 of 30%.

During postoperative evaluations, the patient showed improvement in proptosis in the immediate postoperative period and did not experience rhinorrhea or epistaxis. Nasal irrigation was initiated one week postoperatively, followed by endoscopic wound care every 15 days. Imaging studies were performed one month postoperatively (Figure 1), as well as endoscopic examinations at one month (Figure 2) and three months, which showed no evidence of tumor recurrence.

The patient is currently in the Oncology department, where she is beginning adjuvant radiotherapy. She continues to be followed by our service.

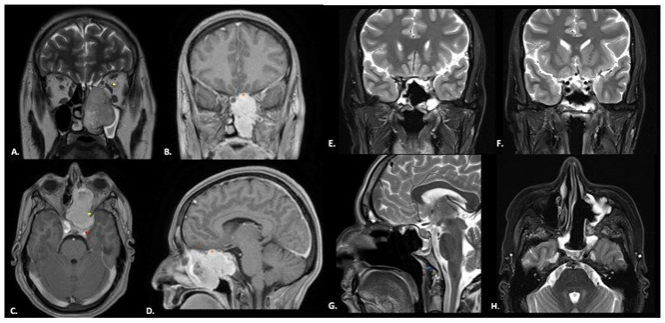

Figure 1: Facial bone MRI with contrast, pre- and post-operative. **(A)** Coronal T1 with contrast: Hypointense tumor with lobulated margins and heterogeneous enhancement after contrast administration, invading the floor and medial wall of the orbit, displacing orbital muscles (medial and inferior rectus) as well as the optic nerve (*yellow). **(B)** Coronal T2: Hyperintense tumor involving the sphenoid plane (*orange) and displacing the left internal carotid artery. **(C)** Axial T1 with contrast: Tumor displacing the lamina papyracea and the left cavernous portion of the internal carotid artery (*red), as well as compressing the left optic nerve. **(D)** Sagittal T2: Involvement of the ethmoid roof, sphenoid plane (*orange), and compression of the sellar region. **(E)** Coronal T2: Wide sphenoidotomy with hyperintense contents in the lateral sinus (*blue); no residual tumor is apparent. **(F)** Coronal T2: Internal carotid arteries and sellar region are observed without tumor pathology. **(G)** Sagittal T2: Wide sphenoid bone with integrity of the meningeal plane. **(H)** Axial T2: Hyperintense mucosa in the maxilla and sphenoid, without tumor remnants.

Discussion

Globally, head and neck cancer ranks sixth. Among all the histological types found in this region, sarcomas are rare, representing only 1% of malignant neoplasms in this area. Of this percentage, most are located in the skin and soft tissues; only 5% occur in the nasal cavity or paranasal sinuses [1].

In pathology, soft tissue sarcomas can be divided into four categories: those with complex karyotypes, those with simple karyotypes with specific reciprocal translocations, those with specific somatic mutations, and those with specific amplifications. The first of these categories includes approximately 50% of soft tissue sarcomas and consists of spindle cell and pleomorphic tumors, among which is the Malignant peripheral Nerve Sheath Tumor (MNTS) [2].

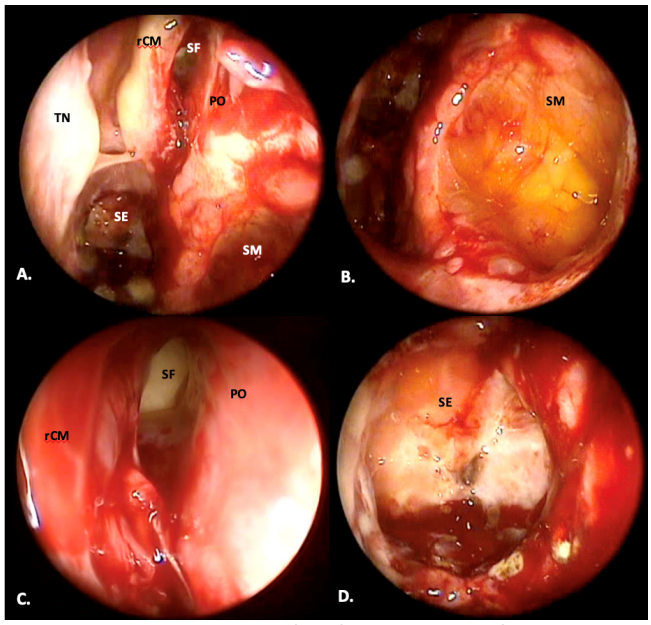

Figure 2: Postoperative Nasal Endoscopy, 1-month post-surgery. **(A)** Left Nasal Fossa. **(B)** Left Maxillectomy. **(C)** Draf IIb Frontal Sinusotomy. **(D)** Extended Left Sphenoidtomy.

TN: Nasal Septum; rCM: Middle Turbinate Remnant; SF: Frontal Sinus; PO: Periorbita; SE: Sphenoid Sinus; SM: Maxillary Sinus.

Neurovascular Tumors (NVTs) are sarcomas that arise from peripheral nerves or other cells associated with the nerve sheath, such as Schwann cells, perineural cells, and fibroblasts [3,4]. They are rare tumors, with an approximate incidence of 0.001% in the general population [5]. They can be associated with Neurofibromatosis type 1 (NF1), with an incidence of 0.16% in this population [6]. Furthermore, radiotherapy can also increase the risk of NVTs in the future. This finding was reported by Ducatman et al., who found 13 cases of post-radiation NVTs out of a total of 120 cases, observing an average latency period of 16.9 years between initial radiotherapy and tumor diagnosis [5]. In the presented case, the patient has no diagnosis of NF1 and no history of radiotherapy exposure.

Regarding the symptoms, at the onset of the disease, these are often common to other nasosinusal diseases such as chronic rhinosinusitis, polyposis, or allergic rhinitis, which delays diagnosis. Therefore, Gore, in his study, highlights the challenge of timely diagnosis and treatment of nasosinusal sarcomas, emphasizing the importance of multidisciplinary management, as well as prolonged follow-up and monitoring. Furthermore, he shows that the type of surgery (endoscopic or open) does not modify survival, which depends more on the tumor stage [7].

From a histopathological and immunohistochemical perspective, TMVNP can mimic other spindle cell tumors, making IHC crucial. In TMVNP, S100 expression is usually patchy, and SOX10 is positive in a subset. In the case reported by Boujida et al., SOX10 was diffuse/intense, S100 was focal, and the Ki-67 index reached 60%; in contrast, our patient showed focal SOX10 positivity, S100 negativity, CD56 positivity, focal CD99 positivity, and a Ki-67 index of 30% [8]. Yao et al. report a positive expression rate in TMVNP of S100 between 50–60%, SOX10 of 27%, and Ki-67 greater than 10% [9].

Magnetic Resonance Imaging (MRI) is the imaging study of choice in these patients, as it aids in staging and surgical planning. Wilson et al. found that Diffusion Restriction (DR) offers particularly high performance in differentiating Nasosinusal Tumors (NMVTs) from benign peripheral nerve sheath tumors,

reporting a pooled sensitivity of 88–93% and a specificity of 94–95% when DR is considered alone or in combination with other features such as perilesional edema, irregular margins, or necrosis. These data support the use of DWI/ADC in aggressive nasosinusal tumors with orbital and skull base involvement, as in the case of this patient [10].

Regarding treatment, the cornerstone is complete resection with negative margins when feasible. The study by Chowdhury et al. on craniospinal Membranous Tumors (MSTs) shows that macroscopic total resection is associated with better survival compared to subtotal resections. Adjuvant radiotherapy improves local control and reduces time to disease progression, although its impact on overall survival is inconsistent; even so, several authors recommend it for high-grade tumors or those with questionable margins [7,11]. Chemotherapy has historically had a limited response. These findings are consistent with the approach taken in this clinical case, where the patient underwent radiotherapy after complete endoscopic resection.

Regarding prognosis, TMVNP maintains high rates of local recurrence (31–75%) and metastasis (22–45%), with 5-year survival as low as 25% in some studies. Adverse factors include tumor size >5 cm, high grade, positive margins, NF1, and Ki-67 >20%. In the study by Chowdhury et al., complete macroscopic resection, low grade, and absence of NF1 tended to improve overall survival; they also recommend adjuvant Radiotherapy (RT) [11]. According to Gore, histological type also plays a role; however, global staging was the most robust predictor in his systematic review [7]. Vengaloor et al. analyzed case reports of patients diagnosed with TMVNP and followed for an extended period, where surgery with adjuvant RT could achieve durable control, even with close/positive margins, reinforcing the need for anatomically wide resections and RT. Furthermore, Vengaloor et al. It presents a clinical case where the patient, thirteen years after completing treatment (surgical plus adjuvant RT), shows no clinical evidence of disease [12].

Therefore, it is important to monitor these patients and, in the event of relapses, to initiate early and timely rescue treatment. Consequently, at the time of writing this clinical case, the patient is still being evaluated periodically in the ENT department of HNAL.

Conclusion

Malignant peripheral nerve sheath tumors are rare neoplasms whose early diagnosis and timely surgical management are essential to improve prognosis and reduce recurrence.

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