“Chondrosarcomas of the skull base" Analyzing a rare pathology.

Andrés Cervio[1], Mauro Ruella[1],Jazmin Fraire[1], Lucila Domecq Laplace[1] , Ruben Mormandi [1], Francisco Marco del Pont[1], Ricardo Marengo[2]

1-Department of Neurosurgery. Fleni, Buenos Aires, Argentina.

2-Department of ENT. CIAC, CEMIC, Buenos Aires, Argentina.

Introduction

Chondrosarcomas are malignant tumors that develop from cells derived from chondrocytes. They constitute about 20% of cancers of the skeletal system and affect people between 40 and 70 years old

Only 2% are located at the skull base and the petroclival location is the most common. Symptoms vary depending on the location of the tumor.

Keywords: Chondrosarcomas; Skull base; Endoscopic endonasal approach; parasellar

Methods and Materials

Retrospective, monocentric study of a cohort of patients operated on for skull base chondrosarcomas at our institution in the last 10 years. Their clinical, imaging, and surgical characteristics were analyzed. A bibliographic research was carried out to compare with other series, management and results.

Results

Eleven patients were included. The average age of 45.6 years (range 23-72) and predilection for the male sex (8:3). The average follow-up was 51 months (range 8-138).

The most common symptoms were cranial nerve involvement and the most common topography was parasellar.

An endoscopic endonasal approach (EAA) was performed in 8 patients and 5 underwent craniotomy. One patient underwent re-operation and 67% were treated with postoperative radiotherapy/radiosurgery.

The outcome was favorable with a mean KPS of 90.

Table 1.Summary of cases demonstrating clinical presentation, imaging findings and treatment.

Age	Gender	Clinical presentation	Location	Volume	Approach	Adyuvancy	Postop. KPS	Follow up (months)
72	M	Headache Diplopia	Ethmoidal	33 mm^3	Microscopic Transnasal	Radiotherapy	100	138
23	F	Headache III CN	Parasellar	32 mm^3	Pterional	No	90	115
30	M	Diplopia IV CN	Parasellar	3.9 mm^3	EEA	No	90	69
65	M	Diplopia VI CN Epistaxis	Parasellar	1.0 mm^3	EEA	Radiotherapy	90	8
72	M	V&VII CNs	Petroclival	2.4 mm^3	EEA	Gamma-Knife	90	32
44	M	I&IV CNs	Anterior fossa	47 mm^3	EEA x2 Pterional x2	Radio-Chemo therapy	90	47
38	F	V CN	CPA	2.8 mm^3	EEA	Radiotherapy	80	14
36	M	Diplopia VI CN	Petroclival	9.3 mm^3	EEA	No	90	10
31	M	Diplopia III&VI CNs	Parasellar	3.6 mm^3	EEA	GammaKnife	90	24
72	F	Headache Gait instability	Petrooccipital	12 mm^3	Retrosigmoid	Radiotherapy	40	9
43	M	Diplopia V&VI CNs	Parasellar	50 mm^3	Pterional	Radiotherapy	90	118

Abbreviations: CN (Cranial nerve); CPA (Cerebello-Pontine angle); EEA (Endoscopic Endonasal Approach)

Results

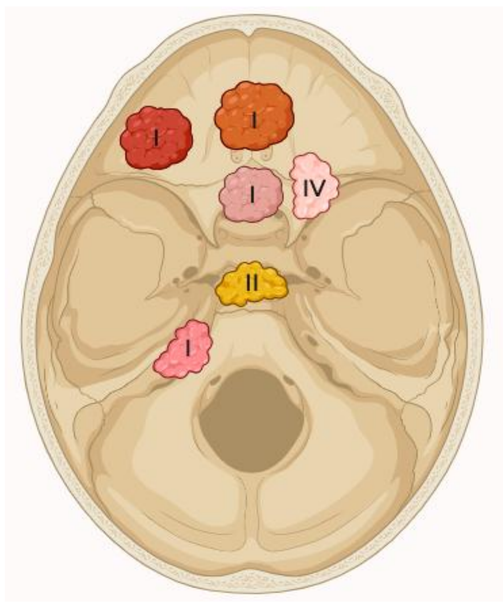

Figure 1. The illustration depicts the most frequent locations of chondrosarcomas in our experience. Clival and para sellar were the most frequently seen, making the EEA approach the most selected one.

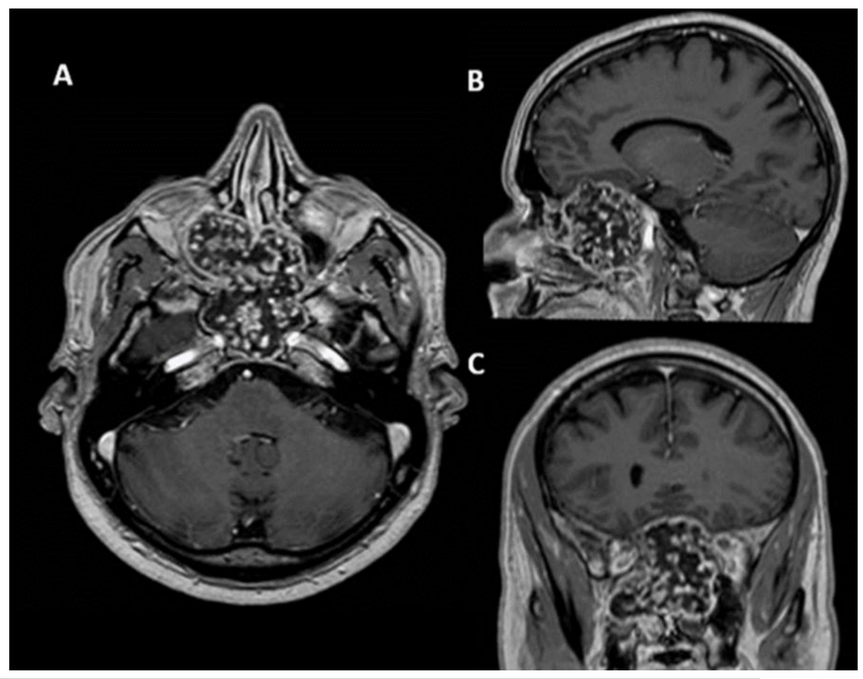

Figure 2. MRI with contrast demonstrating a huge espheno-ethmoid-orbitoclival lesion with heterogeneous enhancement and bony erosion.

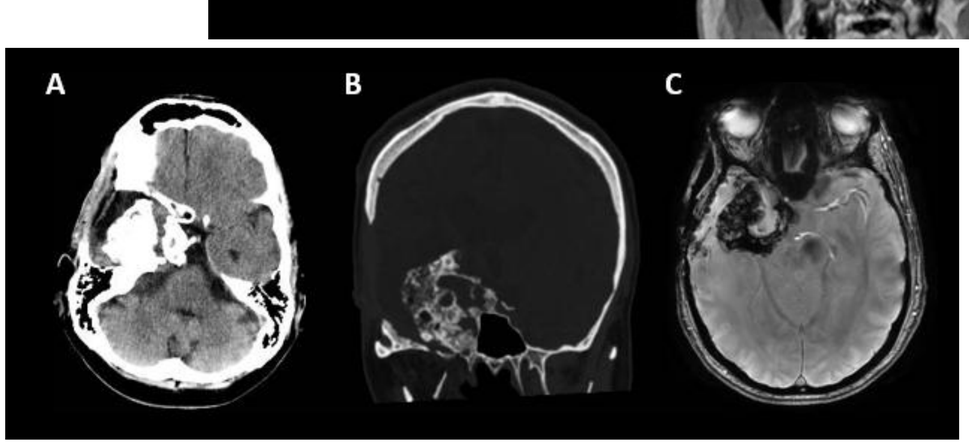

Figure 3. CT scan and MRI with susceptibility-weighted images (SWI) aid in analyzing bony erosion and calcifications of the tumor.

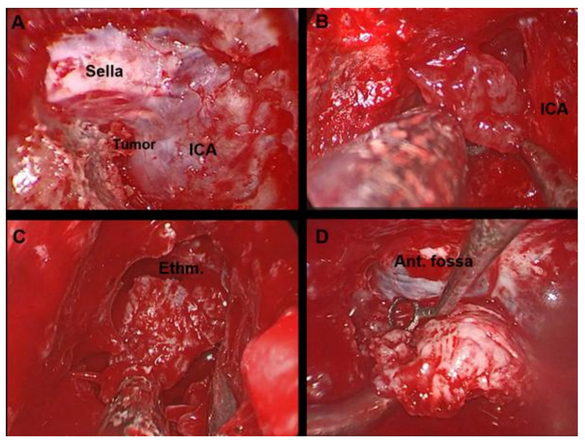

Figure 4, Intraoperative EEA images of patients with chondrosarcoma. This type of tumor usually requires the uncovering of ICA (A) to access tumors with parasellar extension and CS involvement (A and B). It is also often necessary to extend the approach into the anterior fossa (C-D) or transpterygoid approaches for adequate tumor exposure.

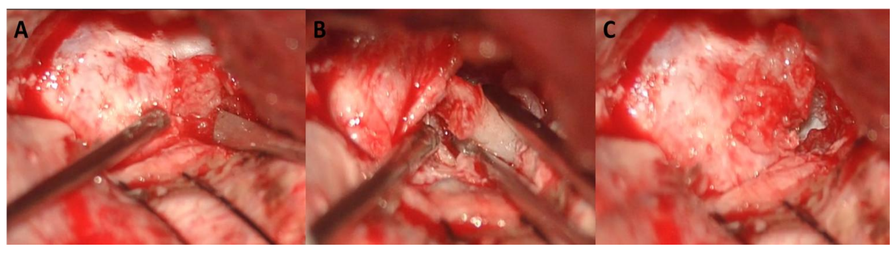

Figure 5. Intraoperative transcraneal approach with extradural peeling of middle fossa (A) and clinoidectomy (B) allowing access to calcified extraaxial tumor removal and decompression of neural structures (B yC)

Conclusions

Chondrosarcomas are rare tumors of the CNS and involve the skull base even more. Surgical resection is the gold standard for treatment as they are usually **resistant** to adjuvancy. Thorough preoperative evaluation of images, appropriate selection of the approach as well as neuroanatomical knowledge are of vital importance. Endoscopic endonasal resection has been reported only in exceptional cases.

Contact

[Andres Cervio]
[Fleni]
[Montañeses 2325, Buenos Aires, Argentina, ZC 1428]
[acervio@fleni.org.ar]
[+54 9 11 5777-3200-int 2702]

References

- Rosenberg AE, Nielsen GP, Keel SB, et al. Chondrosarcoma of the base of the skull: a clinicopathologic study of 200 cases with emphasis on its distinction from chordoma. Am J Surg Pathol 1999;23:1370–8.
- Korten AG, ter Berg HJ, Spincemaille GH, et al. Intracranial chondrosarcoma: review of the literature and report of 15 cases. J Neurol Neurosurg Psychiatry 1998;65:88–92.
- Bloch O, Parsa AT. Skull base chondrosarcoma: evidence-based treatment paradigms. Neurosurg Clin N Am 2013;24:89–96.
- Gay E, Sekhar LN, Rubinstein E, et al. Chordomas and chondrosarcomas of the cranial base: results and follow-up of 60 patients. Neurosurgery 1995;36:887–96 [discussion 896–7].
- Debus J, Schulz-Ertner D, Schad L, et al. Stereotactic fractionated radiotherapy for chordomas and chondrosarcomas of the skull base. Int J Radiat Oncol BiolPhys 2000;47:591–6.
- Hug EB, Slater JD. Proton radiation therapy for chordomas and chondrosarcomas of the skull base. Neurosurg Clin N Am 2000;11:627–38.
- Bouropoulou V, Bosse A, Roessner A, Vollmer E, Edel G, Wuisman P, Harle A: Immunohistochemical investigation of chordomas: Histogenetic and differential diagnostic aspects. Curr Top Pathol 80:183–203, 1989.
- Maira G, Pallini R, Anile C, Fernandez E, Salvinelli F, La Rocca LM, Rossi GF: Surgical treatment of clival chordomas: The transsphenoidal approachr evisited. J Neurosurg 85:784–792, 1996.
- Hug EB, Slater JD. Proton radiation therapy for chordomas and chondrosarcomas of the skull base. Neurosurg Clin N Am 2000;11:627–38.
- Sbaihat A, Bacciu A, Pasanisi E, et al. Skull base chondrosarcomas: surgical treatment and results. Ann Otol Rhinol Laryngol 2013;122:763–70.
- Sekhar LN, Pranatartiharan R, Chanda A, et al. Chordomas and chondrosarcomas of the skull base: results and complications of surgical management. Neurosurg Focus 2001;10(3):E2
- Mesquita Filho PM, Ditzel Filho LF, Prevedello DM, et al. Endoscopic endonasal surgical management of chondrosarcomas with cerebellopontine angle extension. Neurosurg Focus 2014;37(4):E13
- Samii A, Gerganov V, Herold C, et al. Surgical treatment of skull base chondrosarcomas. Neurosurg Rev 2009;32(1):67–75 [discussion: 75].)