

Case Report

Intracranial Chondroma beyond the Skull Base: A Two-Case Series Including an Unusual Presentation with Cerebrospinal Fluid (CSF) Leak and Encephalocele

Stephany Morales Viquez^{1*}, Andrew H Kaye¹, Anna Lazutkin², Ron Eliashar², Samuel Moscovici¹

¹Department of Neurosurgery, Hadassah Hebrew University Medical Center, Jerusalem, Israel

²Department of Otolaryngology, Head and Neck Surgery, Hadassah Hebrew University Medical Center, Jerusalem, Israel

*Correspondence author: Stephany Morales Viquez, Department of Neurosurgery, Hadassah Hebrew University Medical Center, Jerusalem, Israel;
Email: vmorales.sj@gmail.com

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Abstract

Background: Intracranial chondromas are rare, benign cartilaginous tumors typically arising from the skull base. The convexity location is exceptionally uncommon and can mimic more common extra-axial lesions such as meningioma, leading to diagnostic challenges.

Methods: A prospective maintained university medical center registry was utilized to undertake a retrospective review of patients operated with convexity chondroma. Clinical presentation, imaging findings, surgical approach and histopathology were analyzed. A review of literature, particularly concerning atypical presentations such as Cerebrospinal Fluid (CSF) leak, was also conducted.

Results: Both patients underwent successful gross total resection with no perioperative complications or tumor recurrence. One patient presented with a large lesion measured 11 cm in maximum diameter, with stable outcome and recurrence-free at 12-month follow-up. The second patient, a lesion measured 5 cm and was resected and has remained, during 12 years follow-up, stable clinical and radiologically with no recurrence. A CSF leak with associated encephalocele occurred in one case and was managed effectively using an endoscopic endonasal repair in a second operation.

Conclusion: Although rare, convexity intracranial chondromas should be included in the differential diagnosis of extra-axial masses. Optimal outcomes rely on high clinical suspicion and a multidisciplinary approach. These cases illustrate that gross total resection can provide control of these kind tumor, regardless of the size, with excellent long-term outcomes. Clinical significance lies in recognizing that even giant lesions can behave indolently if they are resected completely, emphasizing the importance of accurate diagnosis, multidisciplinary surgical approach, planning and long-term follow-up.

Keywords: Chondroma; Cerebrospinal Fluid Leak; Intracranial Neoplasm; Skull Base; Neurosurgery

Abbreviations

CSF: Cerebrospinal Fluid; MRI: Magnetic Resonance Imaging; ENT: Ear, Nose and Throat; CT: Computerized Tomography; ICP: Intracranial Pressure

Introduction

Intracranial chondromas represent less than 0.5% of intracranial tumors, without gender predominance, in patients between 20 and 40 years of age [1-4]. They mostly originate from the skull base and less likely come from the calvarium, probably from cartilaginous remnants along the sphenopetrosal and sphenoparietal synchondroses. This kind of lesion can either be found isolated or as part of other systemic chondromatoses. Chondromas tend to be slow growing lesions often remaining

asymptomatic for prolonged periods [1-4]. The clinical presentation depends on its location, size and mass effect. It's remarkably rare for the tumors to be arising in the convexity of the skull [1-5].

Radiologically, they appear as well-defined extra axial lesions with calcifications; characteristics can be similar to meningiomas, which are by far more common and present with homogeneously enhancing dural-based mass and dural tail [1-5].

We report two cases of intracranial chondromas and review the literature of this rare entity focusing on the unusual presentation of CSF leak. One of them without any involvement of the skull base presented with CSF leak and anterior skull base fossa encephalocele and the second case involved a right-sided convexity lesion, initially speculated to be a meningioma and with 12 years follow-up. Both of them demonstrate the features of this tumor and their challenging diagnostic process and highlight the importance of a multidisciplinary evaluation in order to reach favorable outcomes.

Case Report

Case 1

A 36-year-old male presented with a several years history of anosmia and three months of spontaneous rhinorrhea. He was examined by an Ear, Nose and Throat (ENT) specialist through a fiber optic laryngoscopy with the finding of a left-sided mass reaching the anterior portion of the nasal cavity. A non-contrast paranasal sinus CT demonstrated a clear bony defect in the cribriform plate and soft tissue density consistent with herniation of the left gyrus rectus through a cribriform plate defect, also extending into the upper nasal cavity (Fig. 1). A Brain MRI was performed, which showed a maximum diameter of 11 cm extra-axial mass in the left hemisphere iso-to hypointense on T1 with a dural-based broad attachment causing marked compression of the adjacent brain parenchyma, including rightward midline shift and effacement of the left lateral ventricle (Fig. 2). He underwent a left craniotomy with complete removal of a large frontoparietotemporal extra-axial lesion. During the surgery a solid vascular mass was resected, debulking was done using the high-speed drill. One week later after the surgery, an endoscopic endonasal transsphenoidal procedure was carried out in order to repair the bony defects in the anterior skull base. Histopathology results confirmed a chondroma. The postoperative course went onto a smooth recovery. At 12 month follow-up, the patient remained asymptomatic and without evidence of recurrence.

Case 2

A 46-year-old male presented with a 2 years history of progressive headaches and dizziness. Neuroimaging demonstrated well-defined, 5 cm extra-axial and dural-based right frontal convexity lesion. The lesion was isointense to hypointense on T1-weighted imaging and caused compression of the adjacent brain parenchyma, but no significant midline shift. A Computerized Tomography (CT) showed an isodense extra-axial lesion along the right convexity, along with associated focal calvarial hyperostosis, initially suspected to be a meningioma (Fig. 3). A right craniotomy with resection of the lesion was performed. Intraoperatively, a firmly dural-based mass, adherent to the underlying dura was found. Histopathology showed a low-grade cartilaginous tumor consistent with a chondroma. The postoperative course was uneventful. At 12 years of follow-up, the patient remains recurrence-free and asymptomatic.

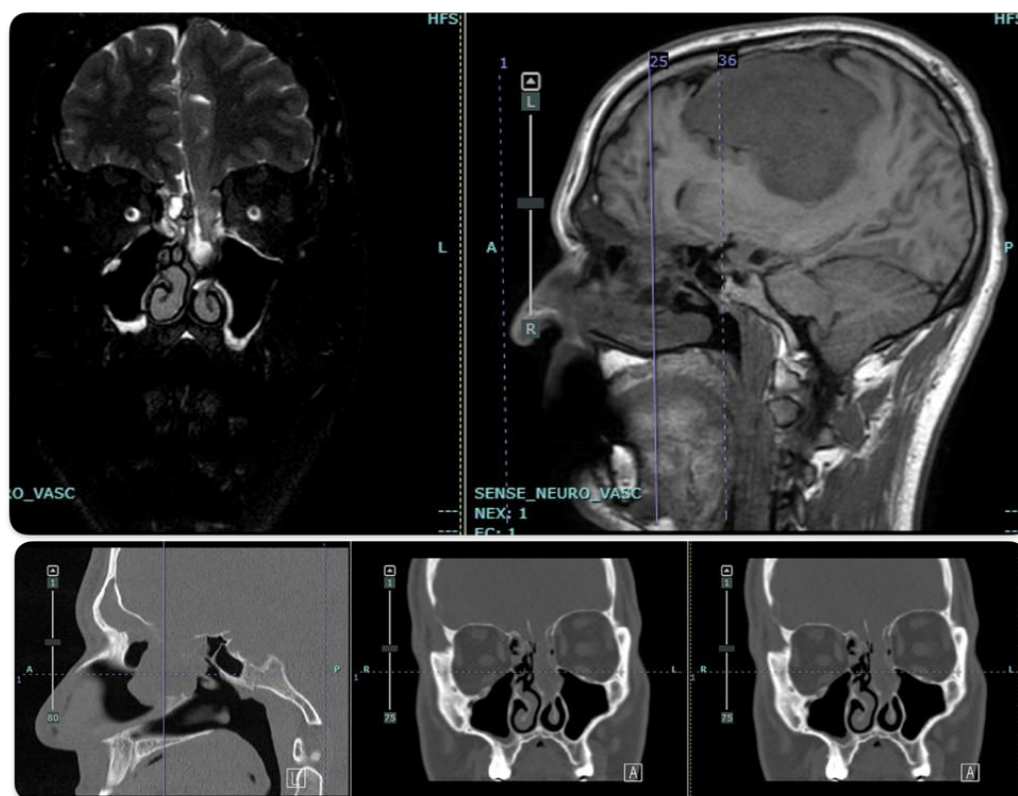


Figure 1: Preoperative MRI and CT scans Paranasal Sinuses (Non-Contrast). MRI (T2 Coronal & T1 Sagittal): Findings of a well-defined encephalocele extending from the left gyrus rectus through a bony defect in the left cribriform plate into the superior nasal cavity.

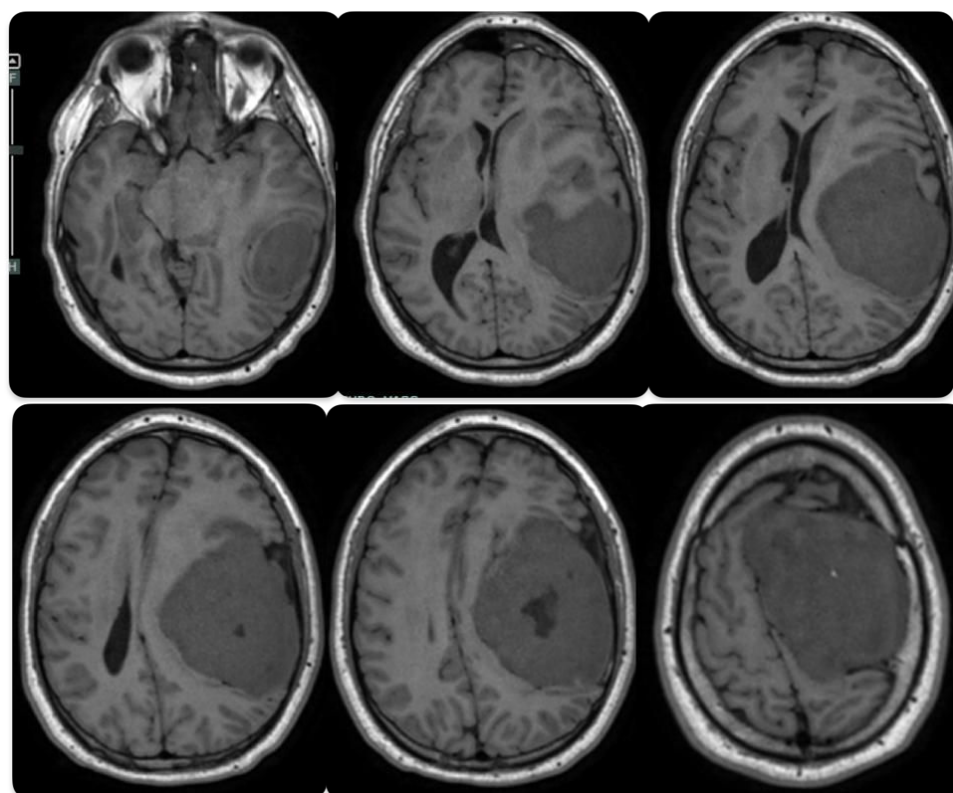


Figure 2: Preoperative MRI. MRI brain axial T1 shows Large left frontal extra-axial mass: Iso- to hypointense on T1 with a dural-based broad attachment, the lesion causes marked compression of the adjacent brain parenchyma, including rightward midline shift and effacement of the left lateral ventricle.

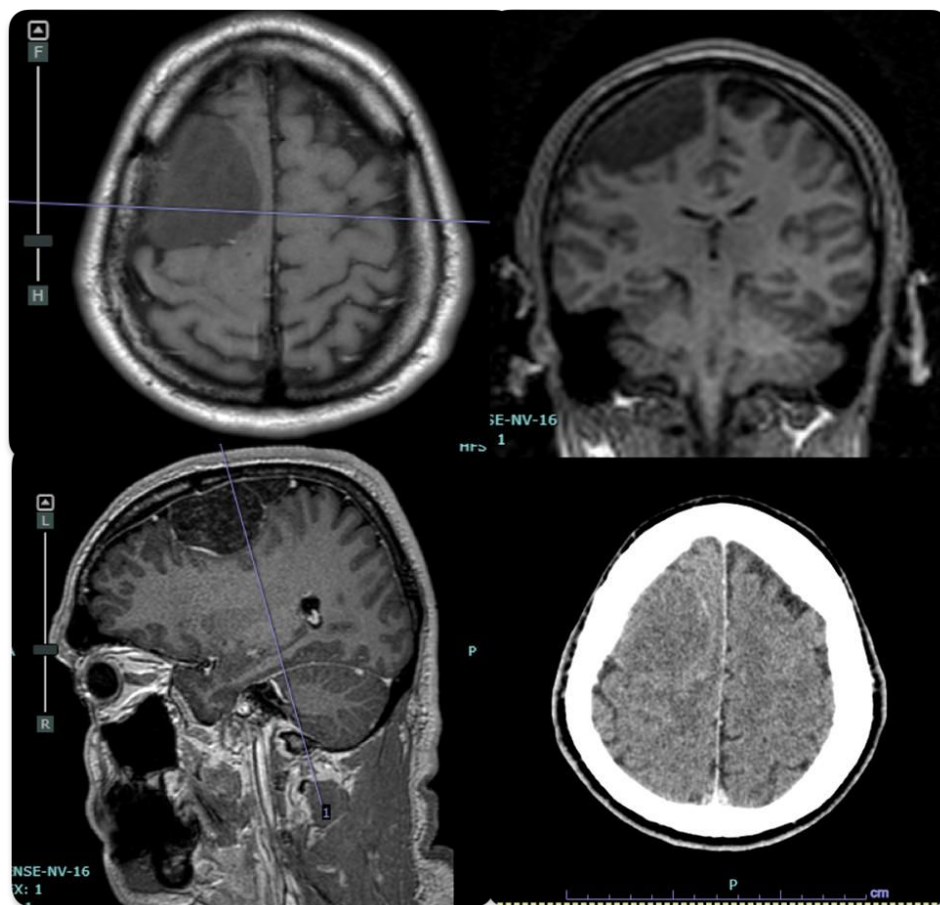


Figure 3: Preoperative MRI and CT scan views show: A well-defined, extra-axial, dural-based mass located in the right convexity region. On axial head CT finding of a isodense extra-axial lesion along the right convexity, associated focal calvarial hyperostosis.

Discussion

The radiological diagnosis of intracranial chondromas is difficult because of their rarity and imaging similarity with more frequent extra-axial masses, such as meningiomas, chondrosarcomas and chordomas [2-4]. The lack of pathognomonic radiological traits makes preoperative diagnosis even more challenging, accentuating the need for histopathological confirmation. While CT imaging may reveal an extra-axial hyperdense lesion and some irregular calcification with hyperostotic adjacent bone or some erosion on a CT a MRI shows an isointense extra-axial lesion with heterogeneous pattern of enhancement on T1. These findings are not exclusive to chondromas and should be interpreted within clinical context [1-5].

Both of our patients were initially approached as more common entities, so they serve as examples of an atypical presentation of chondromas [6]. It is unusual to find a giant extra-axial lesion of the convexity without any continuity with the skull base that presents as a CSF leak after a cribriform plate defect with herniation of the gyrus rectus. We hypothesize that the chronic elevation in Intracranial Pressure (ICP), most likely coming from the space-occupying effect of the giant lesion, caused the continuous erosion of the anterior skull base. This erosion eventually led to dural breach and consequently CSF leak and encephalocele. It also explains the ICP compensation with CSF leak, preventing symptoms of raised ICP or herniation. According to the literature the most common site for chondromas is the skull base and may case localized bone remodeling, but it is exceedingly uncommon for them to present as CSF leak following the result of a high ICP [7].

Decision making in the surgical plan was fundamental in managing this case. It was decided to initially remove the tumor, which was the basis for the raised intracranial pressure and then repair the skull base deficit, responsible for the CSF leak. Endoscopic endonasal techniques, especially in association with vascularized flaps, are demonstrated to be successful in repairing CSF leaks with efficient ablation of skull base lesions [4,6,8]. Combining advanced imaging and intraoperative navigation improves surgical accuracy, reduces perioperative complications and allows the resection of the entire lesion [9-11].

Our second case, involving a convexity chondroma, supports the diagnostic dilemma that these lesions can present. Even though intracranial chondromas most commonly arise from the skull base, a convexity location is an extremely rare location and the lesion can be easily mistaken for a meningioma, due to the characteristic of extra-axial lesion, dural attachment and homogeneous enhancement. Maybe some calcification patterns can increase a hint; but ultimately, the pathology confirms the diagnosis [9-11].

Complete resection remains to be the treatment of choice and is associated with a good long-term prognosis [1,4]. Although these lesions are firm and avascular, they are usually encapsulated and a complete resection can be achieved. A multidisciplinary approach that combines the expertise of neurosurgeons with otorhinolaryngologists ensures comprehensive preoperative evaluation, effective operative approach and optimized postoperative care, which is required for achieving the best results, particularly in complex skull base cases [1,3,4].

From a pathological point of view, it may be difficult to distinguish between chondromas and chondrosarcomas; borderline lesions exists and a long-term surveillance is recommended. The long term prognosis of chondromas is favorable within complete resection, but long-term surveillance is also recommended [1,12].

Conclusion

Intracranial chondromas, particularly those arising from the convexity of the calvarium, are exceedingly rare and can closely mimic more common extra-axial lesions such as meningiomas. Their diagnosis remains challenging due to non-specific radiologic features and their unusual clinical presentations. As illustrated in our series, one patient presented uniquely with CSF leak and encephalocele - an atypical manifestation not previously well described in this context - likely resulting from chronic intracranial hypertension secondary to tumor mass effect.

Conflict of Interests

The authors have no conflict of interest to declare related to this article.

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Informed Consent

Written consent was taken from the patients.

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