CLINICAL CASE REPORT

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Locally Aggressive Behavior and Intracranial Extension in Nasal Chondromesenchymal Hamartoma: Two Pediatric Cases

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Introduction/Background: Nasal chondromesenchymal hamartoma (NCMH) is a rare benign mesenchymal tumor that predominantly affects infants and young children. Although traditionally considered non-aggressive, some cases may demonstrate locally invasive behavior, including skull base erosion and intracranial extension.

Case Presentation: We report two pediatric cases of NCMH with aggressive clinical evolution. The first case involved a newborn presenting with respiratory distress due to a nasal mass. Despite initial surgical resection, early recurrence occurred within five months, with extension to the skull base requiring further partial resection. The second case involved a 9-year-old girl presenting with progressive visual loss, headache, strabismus, and cerebrospinal fluid rhinorrhea. Imaging revealed a large lesion involving the anterior skull base, suprasellar region, clivus, and paranasal sinuses. Following subtotal endoscopic endonasal resection, the patient developed multiple recurrences requiring repeated surgical interventions and developed sixth cranial nerve palsy.

Conclusions: NCMH may exhibit unexpectedly aggressive behavior despite its benign histopathology. These cases highlight the importance of achieving maximal safe resection, close long-term imaging surveillance, and a multidisciplinary management strategy, particularly in cases of incomplete resection.

Keywords: Nasal chondromesenchymal hamartoma, Child; Skull base neoplasms, Intracranial extension, Recurrence

INTRODUCTION

Nasal chondromesenchymal hamartoma (NCMH) is a rare benign mesenchymal lesion that predominantly affects infants and young children. Histologically, it is characterized by a heterogeneous composition of cartilaginous tissue, spindle cells, and cystic components [1, 2].

Although traditionally described as a non-aggressive tumor, because of its benign histology, accumulating evidence indicates that a subset of NCMH cases may exhibit locally aggressive behavior in selected cases, including bone remodeling, skull base erosion, and intracranial extension. The pathogenesis of NCMH remains poorly understood; however, recent studies have suggested a potential association with mutations in the DICER1 gene [1, 2].

Due to its rarity, the natural history and optimal management strategies of NCMH remain poorly defined. In this context, we present two pediatric cases with aggressive clinical evolution, characterized by intracranial extension and recurrence, highlighting the importance of long-term follow-up and careful multidisciplinary management.

CASE REPORTS

Case 1

Clinical Presentation:

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A female newborn was admitted to the neonatal intensive care unit due to respiratory distress. Physical examination revealed a pink nodular lesion occupying the right nasal cavity.

Imaging Findings:

Magnetic resonance imaging (MRI) demonstrated a heterogeneous solid-cystic mass causing deviation of the nasal septum and displacement of the medial orbital wall, without clear evidence of intracranial extension at initial presentation.

Treatment and follow-up:

A transnasal resection was chosen because it was the epicenter of the lesion. Histopathological analysis confirmed the diagnosis of nasal chondromesenchymal hamartoma (NCMH). The goal of complete resection was achieved. However, the patient developed an encephalocele that required correction with a new procedure to reconstruct the base of the skull. Despite initial treatment, follow-up imaging at five months revealed significant tumor recurrence with extension to the skull base. Given the extent of the lesion, a partial resection was performed. At five-year follow-up with a multidisciplinary team, the patient remains clinically stable, with radiological stabilization of the lesion and no need for adjuvant therapy. An evaluation of DICER1 gene would be interesting for genetic activation in this case; however, it was not available for the patient.

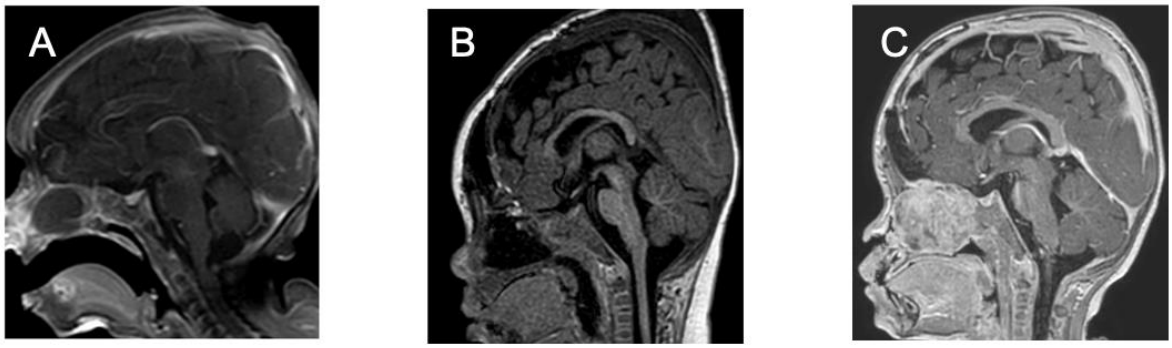

Figure 1. Sagittal T1-weighted contrast-enhanced MRI demonstrating: (A) a preoperative lesion in the right nasal cavity, heterogeneous, with a solid-cystic appearance, showing mild superior bulging of the floor of the anterior cranial fossa, with areas of bone erosion; (B) postoperative findings demonstrating total resection of the lesion; and (C) a recurrent lesion with its epicenter in the ethmoid cells, with infiltration of the skull base, along with dural thickening and enhancement in the basal frontal region adjacent to the olfactory fossa.

Case 2

Clinical Presentation

A 9-year-old female presented with progressive right-sided visual loss, periorbital headache, strabismus and proptosis.

Imaging findings

Magnetic resonance imaging (MRI) revealed a large lesion with heterogeneous enhancement involving the suprasellar region, anterior cranial fossa, clivus, and paranasal sinuses.

Treatment and follow-up:

The initial goal was complete transnasal resection of the lesion; however, this was not feasible due to clival invasion and involvement of the internal carotid artery. In this context, a subtotal resection was performed, focusing on debulking the orbital component. Histopathological examination confirmed the diagnosis of nasal chondromesenchymal hamartoma (NCMH). At nine months of follow-up, a residual

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disease progression was observed, involving the lamina papyracea and the clivus, requiring a second surgical intervention. This procedure was performed via a transcranial approach due to extension into the middle cranial fossa. The lesion was resected and the region was subsequently reconstructed. One year later, further disease progression was noted, associated with sixth cranial nerve (abducens nerve) palsy, requiring an additional transcranial procedure. Since then, three further surgical procedures have been performed with the aim of achieving orbital decompression and debulking of the nasopharyngeal component up to the level of the clivus and the regions adjacent to the bilateral carotid arteries, through combined endoscopic endonasal and supraorbital infraciliary approaches. There was no sign of cerebrospinal fluid (CSF) fistula or CSF rhinorrhoea before or after these approaches. Due to the progression of the lesion after three years of follow up, the patient developed divergent strabismus and is currently undergoing multidisciplinary evaluation and is waiting for a surgical approach with ophthalmology. There is currently no plan for chemotherapy or radiotherapy, but the patient will maintain clinical and radiological follow up with a multidisciplinary team to monitor the growth of the lesion which is not stable yet. An evaluation of DICER1 gene would be interesting for genetic activation in this case; however, it was not available for the patient

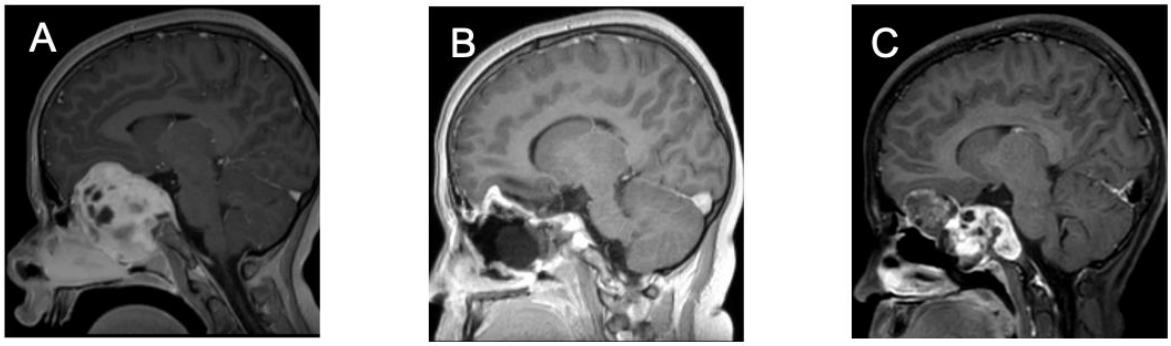

Figure 2. Sagittal T1-weighted contrast-enhanced MRI demonstrating: (A) a preoperative lesion with alcifications affecting the sellar, suprasellar, hypothalamus, clivus, frontobasal, and nasoethmoidal regions, with bone remodeling of the sphenoid regions, the lamina papyracea of the medial orbital walls with extrinsic compression on the lateral rectus muscles, involving the pituitary stalk and the optic chiasm, orbital fissures, optic foramina, invasion with lateral deviation of the intracavernous segments of the internal carotid arteries, invasion of the cavernous sinuses and bilateral trigeminal cavum, posterior extension to the prepontine cistern, resulting in thickening of the bilateral cerebellar tentorium and extension with extrinsic compression on the rectus and orbital gyri; (B) Post-operative findings showing partial ressection with residual lesion in clivus and anterior skullbase; and (C) recurrency of the lesion with intense contrast enhancement in the clivus and parasellar region on the right with some cystic areas in the sphenoid sinus region and superomedial orbital walls bilaterally, compromising the sphenoid wing, which causes slight compression on the medial and superior rectus muscles of the orbits bilaterally. Absence of evident signs of infiltration of the brain parenchyma. Thickening with pachymeningeal enhancement in the frontal basal region and progressing to irregular thickening in the olfactory fossa region.

DISCUSSION

Nasal chondromesenchymal hamartoma (NCMH) is a rare benign lesion, first described by McDermott in 1998 as a tumor resembling chest wall hamartoma [3]. It is characterized by a complex admixture of mesenchymal elements, frequently including cartilaginous and osseous components [4]. Although traditionally regarded as an indolent and non-aggressive entity, a subset of cases may exhibit locally invasive behavior, including bone remodeling, skull base erosion, and, more rarely, intracranial extension [4,5]. Despite its benign nature, at least one case of malignant transformation has been reported in the literature [5].

The clinical presentation of NCMH is highly variable and depends on tumor size, location, and extent of involvement. Reported symptoms include nasal obstruction, epistaxis, visual disturbances, and neurological deficits secondary to mass effect. In infants, respiratory distress and feeding difficulties are commonly observed [6].

From an epidemiological standpoint, NCMH predominantly affects children under one year of age, although cases have also been described in adolescents and adults, with the oldest reported patient being

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70 years old [5,7]. A higher prevalence has been observed in males compared to females [5,7], with no clear evidence of sex-linked inheritance.

Notably, an association between NCMH and the autosomal dominant DICER1 syndrome has been described, characterized by a predisposition to multiple neoplasms due to mutations in the DICER1 gene [8]. This molecular alteration may play a role in tumorigenesis and growth patterns. Although its exact implications in NCMH remain unclear, it is plausible that DICER1 mutations contribute to recurrence risk and more aggressive behavior, supporting the consideration of appropriate genetic counseling. “Unfortunately, DICER 1 testing was not available for our two patients, but since the diagnosis, surgical treatment and follow up was done properly, we believe that the test result would not change these treatments proposed. On the other hand, the testing could have improved other aspects of the treatment, such as genetic counseling for family members and screening for other neoplasms associated with DICER 1 syndrome. Thus, we recommend genetic investigation for DICER 1 syndrome in every NCMH diagnosed when available and its absence of DICER 1 testing might be a limitation of this study.”

Imaging plays a central role in the diagnosis, surgical planning, and follow-up of NCMH. Magnetic resonance imaging (MRI) and computed tomography are essential for initial evaluation, particularly in cases with suspected intracranial extension [9]. NCMH typically appears as a heterogeneous lesion with both solid and cystic components, often associated with remodeling or erosion of adjacent bone structures [9,10]. MRI is superior for assessing lesion extent and soft tissue involvement, whereas computed tomography provides better delineation of osseous changes [6]. In the present cases, MRI was fundamental for detecting early recurrence and accurately defining the extent of skull base involvement.

Surgical resection remains the mainstay of treatment, aiming to prevent recurrence while preserving neurological function. However, complete resection may be challenging in lesions involving the skull base, orbit, or intracranial compartment [5]. The proximity to critical neurovascular structures, such as the optic apparatus, internal carotid artery, and cranial nerves, often limits the extent of safe resection. The endoscopic endonasal approach provides a minimally invasive corridor with excellent visualization of midline skull base structures and has been increasingly adopted in selected cases [6]. As illustrated in the second case, extensive lesions involving the suprasellar region and clivus may require staged procedures or repeated interventions due to residual disease and recurrence.

The cases presented herein demonstrate clinically significant and atypical features, including early recurrence, extensive skull base involvement, and the need for multiple surgical interventions. These findings challenge the traditional perception of NCMH as a purely benign and self-limited lesion. Notably, both patients exhibited disease progression despite surgical management, including after gross total resection in the first case, suggesting that incomplete resection—often dictated by anatomical constraints—may play a central role in recurrence and treatment failure.

The current literature on NCMH remains limited, particularly regarding cases with intracranial extension and recurrence, with approximately 50 cases reported in the English-language literature [5,7]. While most reports describe favorable outcomes following complete resection, with low recurrence rates, the present cases highlight the existence of a more aggressive subset. Furthermore, the combination of extensive intracranial involvement and multiple recurrences, as observed in the second case, appears to be exceptionally rare.

Therefore, long-term follow-up with serial imaging is essential in the management of NCMH, especially in patients undergoing subtotal resection. Our cases demonstrate the varied spectral behavior of this lesion. The first patient remains stable with no signs of recurrence, while the second case is currently under multidisciplinary review due to its more aggressive profile. Early detection of recurrence allows timely intervention and may reduce morbidity. Additionally, these cases underscore the importance of a multidisciplinary approach involving neurosurgery, otolaryngology, radiology, and genetics to optimize surgical strategy and clinical outcomes.

CONCLUSION

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Nasal chondromesenchymal hamartoma (NCMH), despite its benign histological features, may rarely exhibit locally aggressive behavior, with possible intracranial extension and recurrence. The present cases highlight the potential for an atypical clinical course in selected patients, emphasizing the value of long-term imaging follow-up and multidisciplinary management. Further studies are warranted to clarify the factors associated with this behavior and to guide evidence-based treatment strategies.

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DISCLOSURES

Ethical approval

This study was conducted in accordance with the principles of the Declaration of Helsinki. According to institutional policy, ethical approval was not required for this anonymized case report involving two patients.

Consent to participate

Informed consent was obtained from the patients' legal guardians for the use of clinical data and imaging for research purposes.

Conflict of interest

The authors declare no conflict of interest related to the materials, methods, or findings presented in this manuscript.

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Artificial intelligence

The authors affirm that no artificial intelligence tools were used in the writing, editing, or generation of this manuscript. All content was developed by the authors based on clinical experience, scientific knowledge, and literature review.

CONTRIBUTIONS

José Luís Quelho Filho: Conceptualization, data curation, investigation, formal analysis, visualization, writing – original draft, writing – review & editing.

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Linoel Curado Valsechi: Supervision, validation, methodology, writing – review & editing.

Gustavo Botelho Sampaio: Supervision, validation, methodology, writing – review & editing.

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