

Mesenchymal Chondrosarcoma of the Naso-Sinus Region: A Case Report

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Abstract-- Mesenchymal chondrosarcoma (MCS) of the naso-sinus region is an uncommon malignant tumor characterized by aggressive local behavior and a high risk of recurrence. Representing less than 0.5% of paranasal sinus neoplasms, it poses diagnostic and therapeutic challenges due to its rarity and nonspecific early presentation. We report the case of an 18-year-old male with persistent right nasal obstruction and intermittent epistaxis whose histopathological diagnosis confirmed a mesenchymal chondrosarcoma of the naso-sinus region. This report offers a detailed description of the clinical features, endoscopic findings, imaging characteristics, histopathological evaluation, therapeutic approach, and outcome.

A comprehensive review of the literature is also presented to contextualize this case within current scientific knowledge and to highlight key diagnostic and therapeutic insights.

Keywords-- Mesenchymal chondrosarcoma, Paranasal sinuses, Endoscopic surgery, Imaging, Histopathology, Adjuvant radiotherapy, Multidisciplinary care.

I. INTRODUCTION

Malignant tumors of the nasal cavity and paranasal sinuses are rare, accounting for less than 1% of all cancers and approximately 3% of head and neck malignancies. Among these tumors, sarcomas representing cartilaginous differentiation are particularly uncommon. Mesenchymal chondrosarcoma (MCS) is a rare and aggressive subtype of chondrosarcoma, first described in the mid-20th century, distinguished histologically by a prominent primitive mesenchymal component and biologically by aggressive growth and local invasiveness. The intricate anatomy of the nasal cavity, the proximity to vital structures such as the orbit and skull base, and the nonspecific early symptoms contribute to frequent delays in diagnosis.

The clinical presentation of MCS in the naso-sinus region often overlaps with benign processes such as chronic sinusitis or inflammatory polyps, leading to diagnostic challenges. Definitive diagnosis rests on careful clinical evaluation, high-resolution imaging (CT and MRI), and thorough histopathological analysis including immune histochemical profiling.

Optimal management typically involves a combination of surgical resection with clear margins and adjuvant radiotherapy, guided by a multidisciplinary team of specialists.

This manuscript describes an illustrative case of naso-sinus mesenchymal chondrosarcoma, delineates key diagnostic and therapeutic considerations, and situates the discussion within the broader context of existing literature.

II. CASE REPORT

Presentation

An 18-year-old male presented to our otolaryngology clinic with a three-month history of progressive right nasal obstruction, intermittent epistaxis, and occasional headaches. There was no significant past medical history, no history of facial trauma, and no known occupational or environmental exposures. The patient denied anosmia, visual changes, facial pain distinct from sinus pressure, systemic symptoms such as fever, night sweats, or weight loss.

On initial examination, the patient appeared wellnourished with stable vital signs. There was no cervical lymphadenopathy, and cranial nerve examination was normal. Anterior rhinoscopy revealed a firm, non-tender mass occupying most of the right nasal cavity, with mild bleeding on contact.

Endoscopic Findings

Nasal endoscopy using a rigid 0-degree endoscope revealed a lobulated, irregular mass filling the right nasal cavity and displacing the nasal septum medially. The surface of the lesion was polypoid with areas of superficial hemorrhage and mild mucosal inflammation. No ulceration or necrotic centers were clearly visible. The left nasal cavity was normal (figure 1)

The lesion did not visibly invade the nasopharynx or oral cavity. Examination of the posterior nasal cavity and oropharynx revealed no additional abnormalities. These findings necessitated advanced imaging and biopsy for definitive diagnosis.



Figure 1: Polylobulated right nasal mass visualized endoscopically.

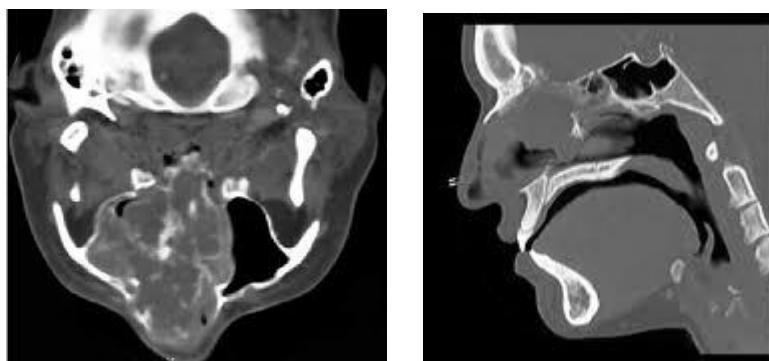


Figure 2: Axial and sagittal sections: Heterogeneous lobulated expansile lesion with septations and calcifications, involving the sphenoid body and extending to both choanae.

IV. MAGNETIC RESONANCE IMAGING (MRI)

MRI revealed a mass hypointense on T1-weighted sequences and heterogeneously hyperintense on T2-weighted images. Post-contrast sequences showed uneven enhancement, consistent with a vascular tumor with variable cellularity.

III. DIAGNOSTIC EVALUATION

Imaging Studies

Computed Tomography (CT)

A non-contrast multidetector CT scan of the paranasal sinuses demonstrated a heterogeneous soft tissue mass within the right nasal cavity, extending into the right maxillary and ethmoid sinuses. The lesion exhibited irregular margins with areas of bone erosion involving the medial wall of the maxillary sinus and the ethmoid labyrinth. Focal calcifications were identified within the mass, suggesting chondroid matrix formation.

The lamina papyracea appeared intact, and there was no evidence of orbital invasion or intracranial extension (figure 2).

Adjacent soft tissue planes were clearly identified, and the imaging ruled out extension into the orbit or skull base. The superior soft tissue resolution provided by MRI was crucial for surgical planning (figure 4).

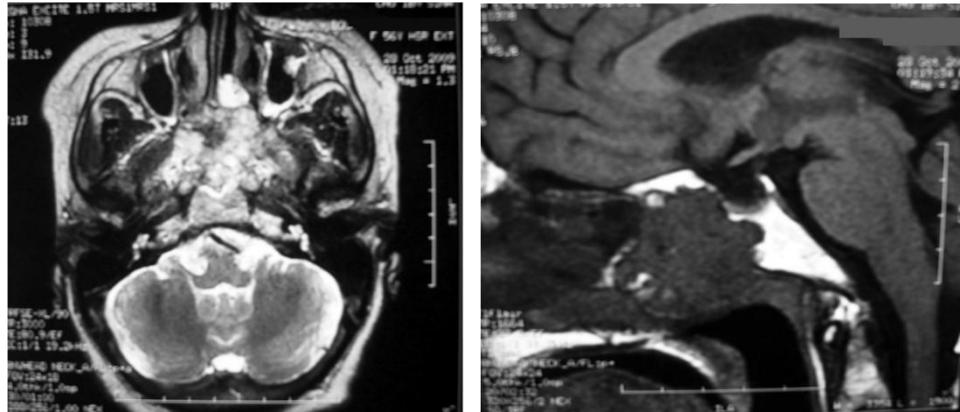


Figure 3: Heterogeneous lobulated expansile lesion containing septations with calcifications, occupying the sphenoid body and extending to both choanae.

V. HISTOPATHOLOGICAL EXAMINATION

An endoscopic biopsy was performed under local anesthesia. Histological analysis revealed a biphasic tumor composed of densely cellular areas of small, undifferentiated mesenchymal cells with hyperchromatic nuclei and frequent mitoses, interspersed with regions of immature cartilaginous matrix. Foci of differential chondroblastic differentiation were evident.

Immunohistochemical staining demonstrated strong positivity for S-100 protein and vimentin, supporting the cartilaginous and mesenchymal origin of the tumor. The tumor cells were negative for cytokeratin AE1/AE3, ruling out carcinoma. These findings supported a diagnosis of **mesenchymal chondrosarcoma, Grade II (intermediate)** according to World Health Organization criteria (figure 4).

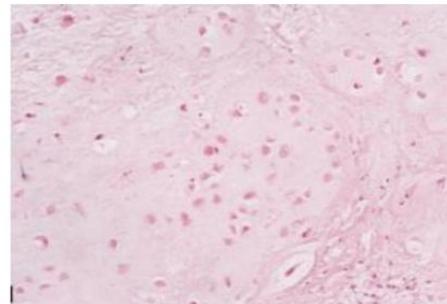


Figure 4: Histological section showing spindle cells and cartilaginous matrix (H&E, x200).

VI. DISCUSSION

Epidemiology and Pathogenesis

Chondrosarcomas are malignant neoplasms characterized by the formation of cartilage matrix. They represent a heterogeneous group of tumors, accounting for 10–20% of primary bone sarcomas. Mesenchymal chondrosarcoma is a rare variant, constituting approximately 3–10% of all chondrosarcomas and an exceptionally small fraction of head and neck sarcomas.

While the exact etiology of MCS remains unclear, several genetic and molecular features have been identified. Mutations in IDH1 and IDH2 are frequently detected in chondrosarcomas and may contribute to oncogenesis by altering cell metabolism and differentiation pathways. Other genetic aberrations involving TP53 and genes regulating mesenchymal differentiation have been described. Although radiation exposure has been implicated in secondary chondrosarcoma development, no such history was evident in our patient.

VII. CLINICAL PRESENTATION AND DIFFERENTIAL DIAGNOSIS

The clinical manifestations of naso-sinus MCS are often nonspecific and include unilateral nasal obstruction, intermittent epistaxis, facial pressure or pain, and occasionally, headaches. Symptoms such as diplopia, proptosis, or vision changes arise only when the tumor invades the orbit. Neurological deficits may signal skull base involvement, emphasizing the need for prompt imaging.

Differential diagnosis for a unilateral nasal mass with bone erosion includes inflammatory polyps, inverted papilloma, esthesioneuroblastoma, carcinoma (e.g., squamous cell carcinoma, adenocarcinoma), osteosarcoma, melanoma, and other sarcomas such as synovial sarcoma or fibrosarcoma. Therefore, distinguishing features on imaging and histopathology are critical for accurate diagnosis.

Role of Imaging

Multimodal imaging is essential. CT scanning provides excellent characterization of bony architecture and reveals calcifications within the tumor matrix, a hallmark suggestive of cartilaginous tumors. **MRI** offers superior soft tissue contrast, delineating tumor margins, involvement of adjacent critical structures, and relationship to the orbit and skull base. Although FDG-PET/CT may be useful for detecting distant metastases, its routine use in naso-sinus chondrosarcoma remains limited.

Histopathological Features

Histologically, MCS displays a characteristic biphasic pattern consisting of undifferentiated mesenchymal cells interspersed with islands of differentiated cartilage. The high mitotic index and cellular atypia distinguish it from conventional chondrosarcoma. Immunohistochemistry supports the diagnosis and excludes other malignancies, with S-100 and vimentin positivity being typical. Cytokeratin negativity helps differentiate from epithelial tumors.

VIII. THERAPEUTIC STRATEGIES

Surgery

Complete surgical excision with negative margins is the cornerstone of treatment and offers the best chance for long-term control. Surgical approaches vary based on tumor extent:

- *Endoscopic endonasal resection* is preferred for localized lesions without extensive invasion, offering reduced morbidity and excellent visualization.
- *Open approaches* are reserved for extensive tumors, involvement of critical structures, or when negative margins cannot be achieved endoscopically.
- *Combined craniofacial techniques* may be required for tumors invading the anterior skull base.

Achieving negative margins significantly influences local control and overall prognosis.

Radiotherapy

Adjuvant radiotherapy is generally indicated in cases of positive or close surgical margins, high histologic grade, or residual disease. Modern techniques such as intensity-modulated radiation therapy (IMRT) and proton therapy enable precise dose delivery while sparing adjacent critical structures.

Chemotherapy

The role of chemotherapy in MCS remains debated, with limited evidence supporting its routine use. It may be considered in cases of unresectable disease, distant metastases, or recurrence, although response rates vary.

Prognosis and Follow-Up

Mesenchymal chondrosarcoma is associated with a high risk of local recurrence and potential for distant metastases. Prognosis depends on histologic grade, completeness of surgical resection, and early detection of recurrence. Regular follow-up with endoscopic examination and periodic imaging is essential. MRI is recommended at intervals of 6–12 months postoperatively, with more frequent assessments during the first two years after treatment.

IX. CONCLUSION

Mesenchymal chondrosarcoma of the naso-sinus region is a rare but aggressive malignant tumor that demands a high index of suspicion, particularly in young patients presenting with persistent unilateral nasal obstruction and epistaxis. Early and accurate diagnosis necessitates a combination of clinical evaluation, advanced imaging techniques, and histopathological confirmation. Complete surgical resection with clear margins followed by adjuvant radiotherapy when indicated remains the most effective treatment strategy. A multidisciplinary approach and vigilant long-term follow-up are essential for improving outcomes.

Ongoing research into molecular markers, advanced imaging modalities, and targeted therapies is needed to further refine management and prognosis for patients with this challenging diagnosis.

REFERENCES

- [1] Thompson LD. Chondrosarcoma of the head and neck: a comprehensive review. *Head Neck Pathol.* 2014.
- [2] Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F. WHO Classification of Tumours of Soft Tissue and Bone. IARC; 2020.
- [3] Gheriani HA, et al. Sinonasal chondrosarcoma: clinical features and outcomes. *J Craniofac Surg.* 2011.
- [4] Yohannan MD, et al. Mesenchymal chondrosarcoma of sinonasal tract. *Am J Otolaryngol.* 2019.
- [5] Enzinger FM, Weiss SW. Soft Tissue Tumors. 6th ed. Elsevier; 2018.
- [6] Cornelis F, et al. Chondrosarcoma of sinonasal region: analysis of 15 cases. *Laryngoscope.* 2009.
- [7] Lewis JS Jr, et al. Mesenchymal chondrosarcoma: updated clinicopathologic review. *Histopathology.* 2017.
- [8] Amary MF, et al. IDH mutations in chondrosarcoma. *J Pathol.* 2011.
- [9] Patel ZM, et al. Sinonasal chondrosarcoma. *Otolaryngol Clin North Am.* 2006.
- [10] Clase M, et al. Genetic aberrations in head and neck chondrosarcomas. *Oral Oncol.* 2014.
- [11] Mertens F, et al. Cytogenetics of chondrosarcoma. *Cancer Genet Cytogenet.* 2006.
- [12] Lisan Q, et al. Sinonasal mesenchymal chondrosarcomas: symptoms at presentation. *Eur Ann Otorhinolaryngol Head Neck Dis.* 2016.
- [13] Kountakis SE, et al. Sinonasal sarcomas clinical review. *Otolaryngol Head Neck Surg.* 2003.
- [14] Lund VJ, Stammberger H. Sinonasal Oncology. Thieme; 2015.
- [15] Perzin KH, et al. Sinonasal neoplasms infiltrating orbit. *AJNR Am J Neuroradiol.* 1986.
- [16] Mineck CW, et al. Intracranial spread of sinonasal tumors. *Neurosurgery.* 1988.
- [17] Som PM, Curtin HD. Head and Neck Imaging. 5th ed. Elsevier; 2011.
- [18] Schild JA, et al. CT imaging in chondrosarcoma diagnosis. *Radiology.* 1995.
- [19] Aiken AH, et al. MRI features of chondrosarcoma. *AJR Am J Roentgenol.* 1996.
- [20] Delgado AF, et al. MRI differentiation of sinonasal tumors. *Eur Radiol.* 2007.
- [21] Even A, et al. Role of PET/CT in bone sarcomas. *Clin Nucl Med.* 2013.
- [22] Barnes L, et al. Surgical Pathology of the Head and Neck. 3rd ed. CRC Press; 2016.
- [23] Triantafyllou A, et al. Sinonasal carcinoma: differential diagnosis. *Histopathology.* 2012.
- [24] Thompson LD, et al. Osteosarcoma of sinonasal region. *Ear Nose Throat J.* 2000.
- [25] Bell D, et al. Sinonasal melanoma. *Head Neck.* 2001.
- [26] Richtsmeier WJ, et al. Histology of mesenchymal chondrosarcoma. *Mod Pathol.* 2008.
- [27] MacDonald KL, Sim FH. Mesenchymal chondrosarcoma pathology review. *Cancer.* 1996.
- [28] Agaimy A, et al. Immunohistochemistry in chondrosarcoma. *Am J Surg Pathol.* 2010.
- [29] Miettinen M, et al. Immunoprofile in sarcoma differential. *Histopathology.* 2014.
- [30] Castelnovo P, et al. Endoscopic resection of sinonasal tumors. *Otolaryngol Clin North Am.* 2016.
- [31] Harvey RJ, et al. Endoscopic management of sinonasal sarcomas. *Laryngoscope.* 2012.
- [32] Weber PC, et al. Open approaches in sinonasal malignancies. *Otolaryngol Clin North Am.* 2013.
- [33] Kraus DH, Dubner S. Combined approaches in skull base surgery. *Otolaryngol Clin North Am.* 2001.
- [34] De Crevoisier R, et al. Radiotherapy in chondrosarcoma. *Int J Radiat Oncol Biol Phys.* 2004.
- [35] Amdur RJ, et al. Adjuvant radiotherapy results in skull base sarcomas. *Head Neck.* 2008.
- [36] Verma V, et al. Proton therapy in skull base tumors. *Front Oncol.* 2017.
- [37] Grimer RJ, et al. Chemotherapy in bone sarcomas. *Cancer Treat Rev.* 2014.
- [38] Briccoli A, et al. Mesenchymal chondrosarcoma outcomes with chemo. *Oncology.* 2010.
- [39] Patel UA, Hwang PH. Surveillance strategies in sinonasal malignancies. *Otolaryngol Head Neck Surg.* 2015.
- [40] Gelderblom H, et al. Prognostic factors in chondrosarcoma. *Ann Oncol.* 2008.
- [41] Evans HL, et al. Chondrosarcoma grading and outcomes. *Cancer.* 1977.
- [42] Martinez SR, et al. Impact of orbital/intracranial invasion in sinonasal tumors. *J Surg Oncol.* 2010.
- [43] Goldstein DP, et al. Recurrence patterns in sinonasal sarcomas. *Head Neck.* 2011.
- [44] Hanna E, et al. Long-term survival in head and neck sarcomas. *Cancer.* 2013.
- [45] Marioni G, et al. Sinonasal sarcomas outcomes and treatment patterns. *Eur Arch Otorhinolaryngol.* 2014.
- [46] Lewis VO, et al. Mesenchymal chondrosarcoma clinical predictors. *Clin Orthop Relat Res.* 2011.
- [47] Laitinen M, et al. Molecular characteristics of chondrosarcoma. *Pathol Oncol Res.* 2018.
- [48] Frezza EE, et al. Sinonasal malignancies: multidisciplinary management. *J Surg Oncol.* 2010.
- [49] Llorente JL, et al. Sinonasal carcinoma review. *Cancer Treat Rev.* 2014.
- [50] Cornelius RR, et al. Surgical margins in skull base sarcomas. *J Neurosurg.* 2011.