INTRODUCTION

Chondrosarcoma is the third most-common primary malignancy of bone after osteosarcoma and myeloma, affecting mainly long bones. Rarely, it can develop in the craniofacial bones, accounting for 8% of all head and neck sarcomas and only 0.1% of all head and neck malignancies. Occurrence of chondrosarcoma in the nasal cavity is uncommon, with less than 200 cases reported in the literature dating from 1950.

Sinonasal involvement is extremely rare, with nasal congestion and obstruction being the leading symptoms. We present a rare case of large chondrosarcoma of the nasal cavity and paranasal sinuses, primarily presenting with ocular symptoms.

KEYWORDS
chondrosarcoma, paranasal sinuses, vision loss

CASE PRESENTATION

A 57-year-old female, with no significant medical history except for nasal septoplasty many years ago, was admitted to our clinic due to progressive decrease of visual acuity in her right eye (RE) for the past 4 months, accompanied by slight periorbital headache. At referral her best corrected visual acuity (BCVA) on Snellen chart was finger counting in the RE and 0.6 in the left eye (LE). Intraocular pressure was normal in both eyes. Although clinically not striking, bilateral proptosis of 22 mm in the RE and 19 mm in the LE was documented on Hertel exophthalmometry (Figure 1). Pupillary reflex and ocular motility were normal, except for slight convergence insufficiency of the RE. No diplopia was present. Slit lamp examination was notable for immature cataract but was otherwise unremarkable. Fundus examination revealed...
incipient epiretinal membrane in the RE, which was confirmed with optical coherence tomography. B-scan ultrasound of the RE showed an extraconic, inferonasally located, large, vaguely demarcated defect of the orbital echo, with propagation of ultrasound into the ethmoid sinus. On standardized ultrasound, the lesion measured up to 12 mm with high internal echogenicity and inhomogeneous structure. The remaining part of the orbital tissue as well as optic nerve diameter were normal (Figure 2). Based on the ultrasound finding, patient underwent thorough investigation including laboratory testing, multi-slice computed tomography (MSCT), and 3 Tesla magnetic resonance imaging (3T MRI) of orbits and sinuses, as well as ear, nose, and throat examination and audiology testing. MSCT and 3T MRI of orbits and sinuses revealed extensive expansive soft tissue density formation within paranasal cavities located in the area of ethmoid cells with destruction of septa, lamina papiracea, and nasal septum, with progression to upper and middle nasal conchae and the sphenoid sinus. The lesion did not have clearly defined contours and was imprinted in the right maxillary sinus with obstruction of the osteomeatal complex. The right maxillary sinus was almost completely obstructed by tumorous formation, retention cysts, and polypoid thickening of the mucosa. Osteodestruction of the lamina cribrosa and planum sphenoidale in the area of mediagittal structures was visible with erased contour of the anterior part of the sella turcica. Proptosis of both bulbs was detected, more pronounced on the right side, where the optic nerve, superior oblique, and medial rectus muscles were suppressed contralaterally (Figure 3). Laboratory testing was unremarkable. Nasal fiberendoscopic examination revealed narrowing in the area of the roof of both nasal cavities, seemingly originating from the ethmoids, with expansion of the tumor mass from the ethmoid toward the nasopharynx. Audiometry and tympanometry were performed to test hearing. Tonal audiogram was unremarkable in the speech area, but it revealed perceptual decline in high frequencies up to 35 dB in the right side and up to 45 dB in the left side. Tympanometry and stapedius reflex were unremarkable. Trans-nasal endoscopic biopsy of the tumor mass was performed and the histopathology analysis confirmed the diagnosis of moderately differentiated chondrosarcoma. The patient was presented to a multidisciplinary oncology team and it was decided to proceed with endoscopic surgery. Intraoperatoratively, it was evident that the tumor involved

**FIGURE 1** Bilateral proptosis with retraction of the upper eyelid, more pronounced on the right side

**FIGURE 2** A, B-scan echography of the RE: an extraconic defect of the orbital echo inferonasally toward the ethmoid sinus. B, A-scan standardized ultrasound measurements
the nasal cavity and septum, both ethmoid sinuses, sphenoid sinus and clivus with extradural endocranial extension. Central tumor debulking was done using a microdebrider, and was followed by sharp dissection of tumor tissue from the periorbita and dura. Clival remnants were drilled to the compact bone. Postoperatively, pulse corticosteroid therapy was administered due to compressive optic neuropathy. The patient also developed syndrome of inappropriate antidiuretic hormone secretion which was treated by endocrinologist and all signs and symptoms subsided within a week. On postoperative ophthalmologic examination, there was evident improvement of BCVA on both eyes: on the RE to 0.01 and on the LE to 0.9 on Snellen chart. Poor visual acuity in the RE is considered an expected repercussion of the associated epiretinal membrane and optic neuropathy.

3 | DISCUSSION

Sinonasal tumors are heterogeneous group of tumors arising from different tissues in the area of nasal cavity, paranasal sinuses, and skull base. Sarcomas, especially chondrosarcomas, are very rare in this region. Diagnosis of cartilaginous tumors is challenging considering varying histopathology and clinical behavior and is based on histopathological analysis. Chondrosarcomas are divided into three histologic grades based on their mitotic rate, nuclear size, staining pattern, and the degree of cellularity, with grade I being well differentiated, grade II moderately differentiated, and grade III poorly differentiated. Vast majority are low- to-intermediate-grade tumors with low metastatic potential, with a minority of high-grade tumors with high metastatic potential and poor outcome. Clinical behavior and prognosis depend on histologic grade with 5-year survival rates being 90%, 81%, and 43% for grade I, II, and III, respectively. Furthermore, the lowest recurrence rate is observed among patients with grade I (15%) and the highest recurrence rate among grade III tumors (33%). The most common presenting symptoms are local swelling and pain, but due to the proximity of the orbit, ocular symptoms and signs may appear first in cases of malignancy in paranasal sinuses. Most of the cases of nasal cavity chondrosarcoma report nasal obstruction and congestion as the leading symptom. There are only few reports of nasal cavity and paranasal sinuses chondrosarcoma presenting with ocular pathology. Due to their relatively poor response to conventional radiotherapy and chemotherapy, surgical resection is the main modality of treatment, but desired complete resection is not often feasible due to the proximity to vital structures, which was the case in our patient. However, in our case, endoscopic tumor resection produced a good result, improving the patient’s symptoms. Close follow-up with regular MRI scans is planned to look for any tumor recurrence.

4 | CONCLUSION

Chondrosarcomas of the paranasal sinuses are quite rare and pose a compelling diagnostic and treatment challenge. Very rarely will they initially present with ocular symptoms as with our patient. A comprehensive range of medical investigation enables timely recognition and treatment.

AUTHOR CONTRIBUTIONS

Author 1: substantial contributions to the conception of the work, analysis and interpretation of data, drafting the work and revising; Author 2: substantial contributions to the conception of the work, analysis and interpretation of data, drafting the work and revising; Author 3: analysis and interpretation of data, revising the work; Author 4: analysis and interpretation of data, revising the work; Author 5: analysis and interpretation of data, revising the
work; Author 6: analysis and interpretation of data, revising the work.

ACKNOWLEDGEMENTS
None.

CONFLICT OF INTEREST
The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT
Data sharing not applicable – no new data generated.

ETHICAL APPROVAL
All procedures were in accordance to the tenets of the Declaration of Helsinki.

CONSENT
Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

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How to cite this article: Kos E, Miletić D, Kuzmanović Elabjer B, Bušić M, Lorencin M, Jurlina M. Rare case of nasal cavity chondrosarcoma presenting only with unilateral decreased vision. Clin Case Rep. 2022;10:e05843. doi:10.1002/ccr3.5843