



Case Report

Myeloid sarcoma of the skull base: A case report and systematic literature review

Prazwal Athukuri¹, A. Basit Khan, Ron Gadot, Monira Haque, Sungho Lee, K. Kelly Gallagher, Martha P. Mims, Gustavo A Rivero, Andrea Barbieri, Akash J. Patel, Ali Jalali¹

Department of Neurosurgery, Baylor College of Medicine, Houston, Texas, United States.

E-mail: Prazwal Athukuri - prazwal.athukuri@bcm.edu; A. Basit Khan - abdul.khan@bcm.edu; Ron Gadot - ron.gadot@bcm.edu; Monira Haque - monira.haque@bcm.edu; Sungho Lee - sungho.lee@bcm.edu; K. Kelly Gallagher - kkgallag@bcm.edu; Martha P. Mims - mmims@bcm.edu; Gustavo A Rivero - garivero@bcm.edu; Andrea Barbieri - andrea.barbieri@bcm.edu; Akash J. Patel - akash.patel@bcm.edu; *Ali Jalali - ali.jalali@bcm.edu



*Corresponding author:

Ali Jalali, MD, PhD,
Department of Neurosurgery,
Baylor College of Medicine,
Houston, United States.

ali.jalali@bcm.edu.

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ABSTRACT

Background: Myeloid sarcoma (MS), or chloroma, is a rare extramedullary malignant tumor that consists of undifferentiated granulocytic cells, and it is most commonly associated with acute myeloid leukemia (AML). Intracranial MS accounts for 0.4% of MS cases, and involvement of the skull base and visual dysfunction is rarely reported. However, the optimal treatment and response to treatment of skull base MS in the presence of visual symptoms is unknown.

Case Description: A 30-year-old male with a history of AML presented with rapidly progressive vision loss and a sellar and parasellar mass with bilateral cavernous sinus and optic nerve encasement. The patient underwent endoscopic endonasal transsphenoidal biopsy revealing intracranial MS. He was treated postoperatively with high-dose intravenous and intrathecal cytarabine and had complete restoration of his vision by postoperative day 11. A systematic review of the literature identified six cases of skull base MS, five of whom presenting with visual symptoms. All patients underwent systemic chemotherapy with cytarabine and/or cyclophosphamide, with infrequent use of intrathecal chemotherapy or radiation. Those with reported visual outcomes were diagnosed 4 months or longer after symptom onset and demonstrated no visual improvement with treatment.

Conclusion: Skull base MS is a rare disease entity with a high prevalence of visual dysfunction. Our patient's complete disappearance of intracranial disease and resolution of visual symptoms with systemic and intrathecal chemotherapy highlight the importance of timely diagnosis and appropriate treatment without a need for direct surgical decompression.

Keywords: Acute myeloid leukemia, Chloroma, Myeloid sarcoma, Parasellar, Skull base

BACKGROUND

Myeloid sarcomas (MSs) are rare extramedullary manifestations of hematologic malignancies such as acute myeloid leukemia (AML), chronic myeloid leukemia (CML), or other myeloproliferative disorders. Frequently termed chloromas due to their green hue, MS is rare with an incidence of 2.5–9% in AML. Moreover, while the mean age of MS is 34.8 years, 60% of all cases of MS occur in children <15 years of age.^[16,20] MS can be diagnosed at any point in the course of the underlying malignancies and rarely may present preceding a leukemia diagnosis.

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MS masses typically manifest within bony or periosteal structures due to direct spread from bone marrow. Due to its rarity, MS is a challenging diagnosis to make with an overall misdiagnosis rate as high as 40%.^[14] Biopsy is the preferred method of diagnosis.^[6,14,19] Rarely, MS manifests within the cranium, termed intracranial myeloid sarcoma (IMS).

IMS is rare, with an estimated incidence of 0.4% in patients with AML and has a mixed prognosis with 84-month overall survival rate of 58.6%.^[12,19] Leukemic cell infiltrates can migrate from the bone marrow into the underlying brain parenchyma, rarely resulting in intra-axial masses.^[6] IMS most commonly presents with headache, vision changes, weakness, altered mental status, and nausea/vomiting of decreasing frequency of presentation.^[12] The treatment of IMS depends on location, extent of systemic disease, age, and functional status of the patient. Although there are no established guidelines for treating IMS through large prospective randomized studies, the mainstay of treatment centers around radiotherapy or systemic chemotherapy. Common chemotherapy regimens include cytarabine-containing remission induction chemotherapy for all MS masses. A low-dose radiotherapy as consolidation treatment can be applied with good prognosis.^[4]

The most common locations for IMS are the temporal lobe, cerebellum, and falx or parasagittal locations, in sum accounting for 30.9% of IMS.^[12] Less than 5% of IMS are located on the skull base.^[12] Here, we report a case of IMS in the sellar and parasellar region presenting with rapidly progressive vision loss. Furthermore, we review the available literature regarding skull base MS to identify the clinical presentations, imaging findings, laboratory workup, and treatment outcomes of reported cases of these neoplasms.

METHODS

All information pertaining to this case was obtained from the electronic medical record. Patient's informed consent was not needed since no identifiable information was disclosed. A simultaneous systematic review of the literature using the online PubMed and EMBASE was conducted using the dedicated search terms "sellar" or "skull base" and "MS" or "chloroma." Given limited results, no date restrictions were set. Articles were included specific case details on skull base MS masses which were reported. Abstracts were screened sequentially for content and relevance; articles were excluded if full text was unavailable in English. The remaining articles were read in full.

CLINICAL PRESENTATION

The patient is a 30-year-old man with a history of AML diagnosed 7 years prior and treated with systemic chemotherapy treatment with fludarabine, idarubicin, and

cytarabine. The patient has been in clinical remission for 1 year after diagnosis, but was lost to follow-up. He presented to our service complaining of rapidly progressive bilateral vision loss and diplopia for the past 2 months. Previously, he had presented to an optometrist who prescribed new glasses. He, additionally, reported 2 months of headaches, nausea, vomiting, and a 26-pound weight loss. Imaging revealed a sellar mass consistent with a pituitary lesion but without the typical appearance of a pituitary macroadenoma. Examination of the right eye revealed restricted extraocular movements in all directions, ptosis, a relative afferent pupillary defect, and minimal light perception. The left eye demonstrated normal extraocular movements and normal pupillary reactivity, but severely restricted visual fields and decreased acuity of foveal vision. Notably, the patient had rapid fluctuations in his visual examination. On dilated fundoscopic examination, there was bilateral Grade IV papilledema.

Magnetic resonance imaging (MRI) demonstrated a lobulated extra-axial mass centered in the sella turcica and expanding in multiple directions along the floor of the anterior cranial fossa and crossing bilateral cavernous sinuses into the middle cranial fossae. A component of the mass additionally extended posteriorly and superiorly into the suprasellar cistern and well as inferiorly in the prepontine cistern. Extracranially, the mass was noted to extend into bilateral orbital apices as well as the sphenoid and ethmoid sinuses. Bilateral internal carotid arteries and cavernous sinus contents as well as optic nerves were encased within the tumor. The mass was hypointense on T1 and isointense on T2 with homogenous enhancement on postcontrast T1 [Figure 1]. Computed tomography (CT) scan of the head demonstrated erosion of the ethmoidal portion of the anterior skull base. The tumor appeared hyperdense on CT scan [Figure 2]. CT scans of the chest, abdomen, and pelvis were unremarkable. Laboratory evaluation revealed decreased ACTH, FSH, and LH. He was started on dexamethasone for optic nerve compression and adrenal insufficiency.

An endoscopic endonasal transsphenoidal biopsy was performed. The intrasphenoidal contents consisted of mucosal cysts with mostly necrotic debris and some viable tissue, which was removed and sent for examination. The anterior wall of the sella was then opened and the mass was encountered immediately under the dura. This was debulked and sent for frozen and permanent pathology. There was no CSF leak, and a multilayer closure was performed. Pathologic review of the tissue showed sheets of blast cells with immunophenotype consistent with MS [Figure 3]. The specimen was positive for CD43, CD34, CD117, and myeloperoxidase (MPO). Flow cytometry study showed 68% aberrant myeloblasts. Next-generation sequencing showed *KRAS* and 2 *CEBPA* (c.1027 del and



Figure 1: Preoperative MRI. Postcontrast T1 (a) sagittal, (b) axial, and (c) coronal magnetic resonance imaging demonstrating a mass (white arrow) involving the sellar and parasellar region.

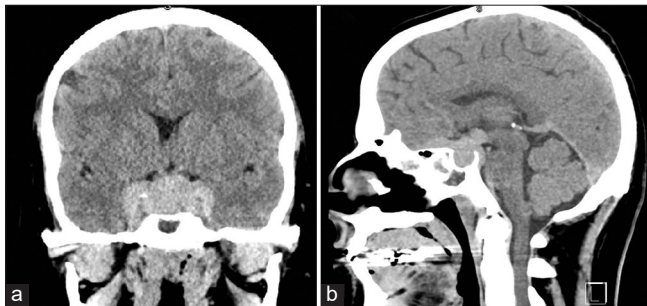


Figure 2: Preoperative CT. (a) Coronal and (b) sagittal computed tomography imaging demonstrating a hyperdense sellar mass.

c.937_939dup) mutations at 46%, 44.6%, and 25.7% variant allele frequency, respectively. His bone marrow showed no evidence of AML.

Immediately after surgery, the patient reported only a slight improvement in vision. On postoperative day (POD) 2, high-dose intravenous cytarabine at 2000 mg/m² over five doses was initiated. On POD 4, his vision had improved to right eye 20/100 and left eye 20/100 centrally when reading at a distance of 12 inches and with normal extraocular movements. By POD 11, the patient had recovered to essentially normal eye function on ophthalmology examination. He was given intrathecal cytarabine on POD 14 over four doses. Abnormal myeloblast ratio in the CSF dropped from 84% on POD 0 to <1% by POD 42 [Table 1]. In the initial flow cytometry, majority of the cells were aberrant myeloblasts with bright CD45+ lymphocytes, granulocytes, and monocytes comprising only 4.5%, 3.6%, and 1.7% of total cells, respectively. T-cells showed a normal CD4:CD8 ratio of 2.7 and normal expression of the pan T-cell antigens CD3 and CD5. B-cells were polytypic with a κ : λ ratio of 1.0, indicating no abnormalities. In the final flow cytometry, bright CD45+ lymphocytes and CD14+ monocytes comprised 23.5% and 0.9% of total cells, respectively, and granulocytes comprised the majority of cells analyzed with <1% of aberrant myeloblasts. The hospital course was complicated by pancytopenia and neutropenic fever

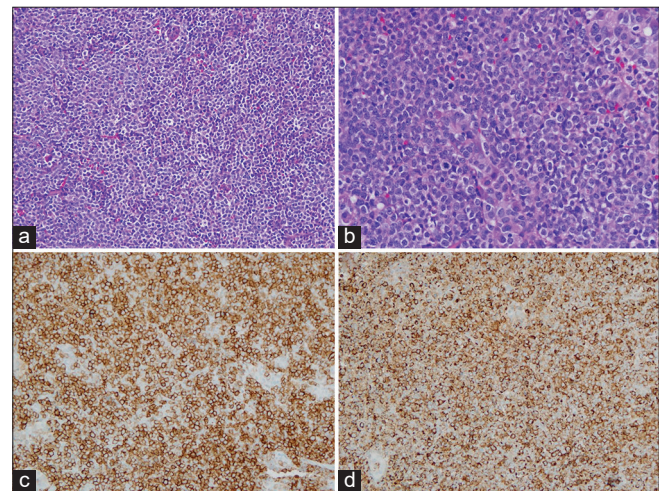


Figure 3: Myeloid sarcoma: the tumor shows sheets of intermediate sized blasts with scant cytoplasm, irregular nuclear contours, finely dispersed chromatin, and prominent nucleoli (a). Mitotic figures are numerous (b). The neoplastic cells strongly express CD117 (c) and myeloperoxidase (d).

Table 1: Postoperative flow cytometry.

Marker	Day 0	Day 44
CD45 (dim)	+	–
CD34	+	–
CD13	+	–
CD33	+	–
CD117	+	–
CD7	+	–
CD56	+ (small subset)	–
CD19	+ (small subset)	–
CD10	–	–
cTDT	–	–

and the patient required continued treatment for adrenal insufficiency and hypothyroidism from panhypopituitarism due to the mass. Follow-up imaging nearly a month after surgery and chemotherapy showed near-total disappearance of the mass and resolution of the portions encasing the

Table 2: Prior reports of skull base myeloid sarcoma.

Patient	Author	Age/ sex	Presentation	Imaging characteristics	Diagnostic and treatment approach	Time from symptom onset to presentation	History of leukemia diagnosis; medullary and extramedullary findings	Preoperative ophthalmologic findings	Postoperative ophthalmologic findings	Preoperative hematological function	Postoperative hematological function	Immuno histochemistry and flow cytometry	Clinical outcome
1.	Fruauff <i>et al.</i> ^[8]	14- y.o.* M*	Neck pain, headaches, weight loss, papilledema	Large, bilobed, enhancing, high attenuation lesion starting in the posterior fossa	Exploratory neurosurgery with systemic and CNS* chemotherapy for potential AML* Needle core biopsy followed by craniotomy due to concerns of herniation. Four cycles of cytarabine (systemic chemotherapy)	1-2 months	Unremarkable	Bilateral papilledema	NR	Unremarkable	NR	NR	NR
2.	Grier <i>et al.</i> ^[9]	41-y.o. F*	Mutism, bradykinesia	Contrast-enhancing left-sided middle cranial fossa with extension into subtemporal fossa and parapharyngeal space on MRI*	Transsphenoidal excision followed by chemotherapy	2 days	Unremarkable; medullary and extramedullary findings were not reported	NR	NR	Unremarkable	NR	MPO+, CD34+, CD13+, CD33+, CD117+, CD20-, CD10-, aberrant expression CD19, partial expression CD56	Improved mental status after chemotherapy. Lost to follow- up after 7 months
3.	Novello <i>et al.</i> ^[13]	54-y.o. M	Acute onset of right cranial nerve 3 palsy, leg pains	Lesions in the sellar, upper clival, and right parasellar region, isointense/slightly hyperintense on CT*, dislocation optic chiasm and pituitary stalk compressing and invading right cavernous sinus	Transsphenoidal excision followed by chemotherapy	“Acute”	Myeloproliferative neoplasms diagnosis in 1998, Myelofibrosis diagnosis in 2008. Postoperative acute myeloid leukemia from the megakaryoblastic lineage	Right cranial nerve 3 palsy	NR	Massive splenomegaly, anemia, thrombocytopenia	Postoperative myeloid leukemia	CD34+, CD42b+, CD45+, CD61+, LAT, MPO-, CD68-, glycophorin-, CD71-	Clinical condition worsened and patient died 2 months after surgery
4.	Suzuki <i>et al.</i> ^[17]	39-y.o. M	Left nasal obstruction and swelling, chronic rhinosinusitis, left proptosis	Left ethmoidal mass involving maxillary sinus, orbit, and skull base on CT and MRI	Surgery for tissue sampling, followed by remission induction therapy with daunorubicin and cytarabine. Salvage chemotherapy with radiation therapy. Myelodysplastic syndrome treated with allogenic hematopoietic stem cell transplantation (allo-HSCT*)	NR*	Unremarkable; marrow with excess myeloblasts; megakaryoblasts and giant neutrophils during complete remission period	Left-sided proptosis	NR	Leukocytosis composed of myeloid blast cells, mild anemia, and thrombocytopenia	Unremarkable	MPO+, CD34+, CD33+, HLA- DR+, CD56-, CD3-, CD20-, CD56-, Tdt-, 94% myeloblasts	Complete remission with no evidence of recurrence after allo-HSCT at 12-month follow-up

(Contd...)

Table 2: (Continued).

Patient	Author	Age/ sex	Presentation	Imaging characteristics	Diagnostic and treatment approach	Time from symptom onset to presentation	History of leukemia diagnosis; medullary and extramedullary findings	Preoperative ophthalmologic findings	Postoperative ophthalmologic findings	Preoperative hematological function	Postoperative hematological function	Immuno histochemistry and flow cytometry	Clinical outcome
5.	Thakar <i>et al.</i> ^[18]	20-y.o. M	Garcin's hemibase syndrome, 1 episode of generalized seizure, hypoesthesia on left side of the face (V1 and V2), facial weakness (wasting of the left masseter muscle), lower motor neuron facial paresis with prominence on the left side, sensorineural deafness on the left ear, palate deviated to the right side, gag reflex absent on the left side (difficulty swallowing), diplopia, complete left eye ophthalmoplegia, absent left corneal reflex	Multiple, extra-axial, contrast- enhancing dural-based lesions in bilateral sphenopetro-clival regions. Lesion in the right parietal convexity extending into the subgaleal plane, bilateral lesions in the sphenopetro-clival skull base regions, mirroring each other, with the left side having a larger component in the posterior fossa extending to the foramen magnum on MRI	Surgery followed by cyclophosphamide- based chemotherapy and radiation therapy	4 months	Unremarkable	Diplopia, complete left eye ophthalmoplegia, absent left corneal reflex	Clinical condition remained constant	Unremarkable	NR	MPO+, CD117+, CD43+, epithelial member antigen-, CD3-, CD20-, CD68-, CD34-, CD99-, CD1a-	Clinical condition remained constant while imaging showed marginal decrease in size of the lesions in the 15-month follow-up. Repeat bone marrow biopsy showed persistence of nonleukemic status
6.	Xu <i>et al.</i> ^[19]	29-y.o. F	Progressive visual loss in the right eye, 20 kg* (~44 lb*) weight loss	Saddle area-occupying lesions on CT	Endoscopic sellar region tumor resection, CHOP* regimen chemotherapy. Modified chemotherapy regimen (high-dose cytosine arabinoside) and consolidation chemotherapy after correct diagnosis of myeloid sarcoma	1 year	Unremarkable; abnormal bone density in limbs, ribs, sternum, spine, and pelvis. Improved after correct treatment (cytosine arabinoside)	Progressive visual loss in the right eye	No improvement in blindness in the right eye. Not reported after correct diagnosis.	Anemia (low RBC* and Hg* levels)	Refractory anemia	LCA+, CD4+, CD43+, CD68+, CD117+, CD99+, CD1a+, CD34+, CD3+/-, CD8+/-, MPO-, TdT-, AE1/3-, EMA-, S-100-, CD10-, CD20-, CD79a-, Ki-67+	No improvement on the blindness in the right eye. Refractory anemia and sternal tenderness after surgery and first round of chemotherapy. 8 months later, correct diagnosis was made. Three months following that, the patient condition is stable and follow-up will be continued.

(Contd...)

Table 2: (Continued).

Patient	Author	Age/ sex	Presentation	Imaging characteristics	Diagnostic and treatment approach	Time from symptom onset to presentation	History of leukemia diagnosis; medullary and extramedullary findings	Preoperative ophthalmologic findings	Postoperative ophthalmologic findings	Preoperative hematological function	Postoperative hematological function	Immuno histochemistry and flow cytometry	Clinical outcome
7.	Current Case	30-y.o. M	Acute pain distress, 26 lb weight loss, diplopia, decreased vision in the left and right eye, limited eye movements, severe papilledema, right V2 and V3 hypesthesia, and general neurologic decline	Large extra-axial sellar/ suprasellar mass involving the sphenoid sinus, bilateral cavernous sinuses, all cranial fossa, infiltration into pituitary stalk and hypothalamus, encasing bilateral internal carotid arteries and optic nerve sheaths with remodeling of the sella turcica on MRI and CT	Endoscopic endonasal transsphenoidal approach followed by postoperative systemic and intrathecal chemotherapy	2 months	AML diagnosis 7 years ago, currently in remission; unremarkable medullary and extramedullary findings	Right eye proptosis, right cranial nerve 6 palsy, diplopia, decreased vision in the right and left eye, severe papilledema	Nearly complete resolution of visual symptoms	Mild anemia, increased PTT	Episodes of neutropenic fever (resolved), pancytopenia (resolved)	68% aberrant myeloblasts, CD43+, CD34+, CD117+, MPO+, CD7+, HLA-DR+, CD13+, CD33+, CD56+/-, CD19+/-, CD45+/-, CD10- cTdT-, CD14- CD16-, CD11b-, CD64-, CD36- cCD79a-, sCD3- cCD3-	No eye pain, no headaches, and stable vision by postoperative day 11. Few hematological and endocrine complications. Follow-up imaging at 6 months shows no evidence of tumor. Near-normal vision with mild difficulty focusing and KPS score of 90 on 8-month follow-up.

*Y.O: years old, M: male, F: female, Kg: kilogram, Lb: pounds, MRI: magnetic resonance imaging, CT: computed tomography, CNS: central nervous system, AML: acute myeloid leukemia, allo-HSCT: allogenic hematopoietic stem cell therapy, CHOP: Cyclophosphamide, Adriamycin, Oncovin, and Prednisolone, NR: not reported, RBC: red blood cells, Hg: hemoglobin

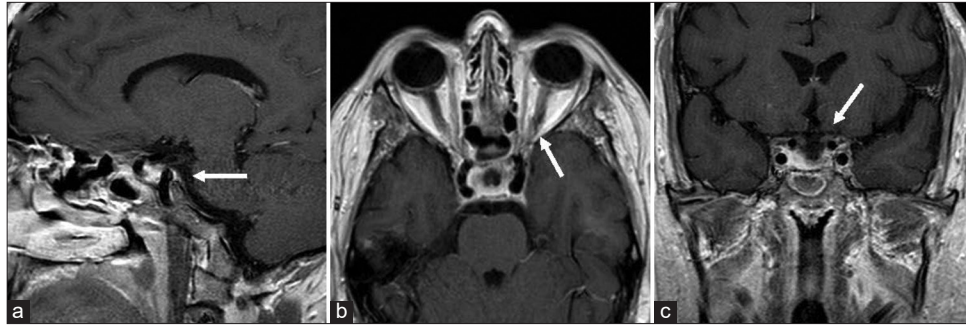


Figure 4: Postoperative MRI (a) T1 sagittal, (b) T1 axial postcontrast, and (c) T1 coronal magnetic resonance imaging demonstrating debulking of the sellar and suprasellar portion of the mass (white arrow).

optics nerves and cavernous sinus contents [Figure 4]. There was no evidence of recurrent tumor on follow-up imaging 6 months after surgery. On 8-month telephone follow-up, the patient's Karnofsky Performance Scale (KPS) score was 90, and he reported near-normal vision with mild difficulty focusing.

SYSTEMATIC REVIEW OF THE LITERATURE

Using dedicated search terms, our systematic review identified 19 articles. After reviewing the abstracts of each, 10 articles were excluded for not satisfying inclusion criteria. The full texts of the remaining nine articles were read in full, after which three additional articles were excluded for lack of specific case details on skull base MS masses. Our systematic review, thus, identified six previously reported cases of skull base MS masses [Supplementary Figure 1]. Data regarding demographics, presenting symptoms, imaging characteristics, leukemia diagnosis, metastases status, preoperative and postoperative symptoms, and findings were obtained from the included articles [Table 2].^[8,9,13,17-19]

Clinical presentation

Four patients were male and two patients were female with ages ranging from 14 to 54 years. All patients presented with skull base or sellar region MS masses. Five of the six patients presented with symptoms of ophthalmoplegia (2), vision loss (2), and diplopia (1). Three of the six patients presented within 2 months of symptoms. Two of the six patients presented after 4 months of symptoms. One report did not indicate the time from symptom onset to presentation. Two of the six patients also presented with significant weight loss as noted in our patient. Three of the six patients presented with anemia and thrombocytopenia. Only one patient had a previously diagnosed myeloproliferative neoplasm, whereas the other five patients presented with a MS mass without any history of leukemia.

Imaging findings and laboratory workup

Four of the six patients presented with solitary MS masses in the skull base or sellar region whereas the other two also had abnormal findings on bone marrow biopsy. The MS masses were localized in the anterior, middle, and posterior cranial fossae, including the parasellar, sphenopetroclival, and orbital regions of the skull base. Five of the six case reports described abnormal findings with immunohistochemistry and flow cytometry consistent with MS, with a common finding of aberrant myeloid cell population. Four cases were CD34+, indicating enhanced progenitor activity.^[15] Three cases were MPO+ and CD117+, which indicate myeloid differentiation.^[1] Two cases were CD33+, which has been detected on blasts in AML.^[7]

Treatment regimen and outcomes

Of cases with vision loss, four reports did not indicate whether the vision findings changed with treatment. The cases with vision loss did not undergo a direct decompression of optic nerves in the acute setting. One patient did not respond to chemotherapy and died after 2 months of worsening clinical condition and progressive systemic disease. All patients underwent surgical biopsy followed by systemic chemotherapy. One case specified that the patient underwent intrathecal chemotherapy. Two of the six patients received radiation therapy as part of multimodal treatment. Four of the six patients received either cytarabine or cyclophosphamide, whereas, in two patients, no details of the regimen were reported.^[10,20] Two reports commented on visual outcomes after treatment.^[18,19] These were diagnosed 4 months or longer after symptom onset and demonstrated no visual improvement with treatment.

DISCUSSION

Here, we report a case of a 30-year-old man with a history of AML in remission who presented with rapidly progressive vision loss. MRI and CT revealed a mass in the sellar and

parasellar region with encasement of bilateral optic nerves and cavernous sinus components, which prompted surgical biopsy. Pathological review revealed abnormal myeloblasts and immunophenotype consistent with MS. The patient received systemic cytarabine chemotherapy and had complete recovery of vision and eye movement within a week of starting the treatment. A systematic review of the literature was performed to identify characteristics of skull base or sellar region MS. To the best of our knowledge, there are only six other reported cases in the literature. Similarly to our patient, five of the six previously reported patients presented with visual dysfunction. In all reported cases, surgical biopsy followed by systemic chemotherapy was the mainstay of treatment.

IMS masses appear hyperdense on noncontrast CT and demonstrate intense homogenous enhancement on CT.^[16] Furthermore, they appear isointense to hypointense on T1-weighted and mildly hyperintense on T2-weighted MRI with homogenous contrast enhancement.^[12,16] Despite these typical characteristics, there is a high rate of misdiagnosis with imaging alone with an overall misdiagnosis rate as high as 40%.^[14] The differential diagnosis for intracranial masses with similar imaging characteristics includes meningioma, hematoma, abscess, and intracranial metastasis.^[3,6,11] Alternative diagnoses with evidence of leukemic infiltration include meningeal disease (i.e., carcinomatous meningitis), intravascular tumor aggregates (i.e., carcinomatous encephalitis), or focal tumor masses other than MS.^[12,20] Consequently, a biopsy is frequently required to confirm the pathological diagnosis of IMS.^[6,14,19] To avoid a false-negative biopsy, immunophenotyping of various myeloid cell developmental stages is necessary.^[12] The most commonly expressed markers in IMS in descending order of frequency are MPO, CD117, CD34, CD45, CD68, and CD43.^[12] Our patient was positive for CD45 (dim), MPO, CD117, CD34, and CD43. MPO and CD117 are sensitive for myeloid differentiation.^[1] Pathologic and cytogenetic studies of 13 MS tissue samples indicated that the presence of CD34 suggests that the mass is not *de novo*.^[1] The listed markers, along with a history of AML, confirmed the diagnosis of IMS due to relapse following clinical remission of AML in our patient.

Prognosis of MS is worse when associated with relapse of AML, myeloproliferative disorder, or myelodysplastic syndrome.^[14] AML is the most common etiology of IMS followed by CML.^[12] IMS can present as part of active AML (19.7% of the cases), precede the diagnosis of AML (18.2% of the cases), or be the initial manifestation of AML relapse (62.1% of the cases) as in our patient.^[12] Based on our systematic review, a combination of surgical biopsy combined with a high dose of cytarabine or cyclophosphamide is frequently used to treat skull base and parasellar MS [Table 2].^[5,12] Chemotherapy regimens with cytarabine, used in cases of AML remission, can help to result in remission and decreased progression of acute leukemia in

the blood or marrow space.^[10,20] In cases of isolated MS masses that develop in relapse after chemotherapy, reinduction of AML chemotherapy in conjunction with hematopoietic stem cell therapy should be considered.^[4] A meta-analysis of case reports and case series on the treatment of IMS indicated that 53.7% of the patients underwent surgery, of which 24.3% of the patients achieved gross-total resection and 75.7% of the patients underwent subtotal resection or biopsy. About 54% of the patients underwent chemotherapy, and 44.4% of the patients underwent radiotherapy. Both radiotherapy and chemotherapy alone were statistically significant in reducing mortality rate in IMS.^[2,12] Surgery alone did not reduce mortality in IMS. In our patient, even though both optic nerves were encased by tumor through their intracranial and orbital course, direct surgical decompression was deemed high risk and unnecessary. However, he had a complete resolution of visual symptoms after biopsy and subsequent systemic and intrathecal chemotherapy, which is unique to what was observed in Thakar *et al.* and Xu *et al.*^[18,19] A quicker time from symptom onset to presentation in our case resulted in better vision outcomes. Disappearance of the cavernous portion of the mass with treatment also resulted in resolution of ophthalmoplegia and diplopia. Our experience suggests that timely diagnosis through biopsy followed by an appropriate chemotherapy regimen can adequately treat skull base MS and reverse the associated vision loss.

CONCLUSION

We report a case of MS of the sellar and parasellar region diagnosed through endonasal biopsy and managed with chemotherapy. We, additionally, reviewed the extant literature reporting six cases that discuss this rare disease entity. Rapid diagnosis and treatment led to complete resolution of visual and neurological symptoms in our patient without a need for direct surgical decompression. Accordingly, MS should be considered on the differential diagnosis in patients with similar clinical and imaging profiles.

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Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

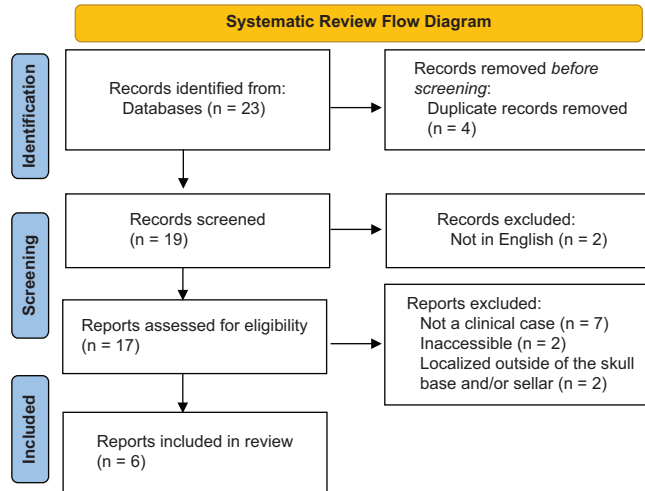
There are no conflicts of interest.

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SUPPLEMENTARY FIGURE



Supplementary Figure 1: Systematic review flow diagram.