



Case Report

Intrasellar cavernous hemangiomas: A case report with a comprehensive review of the literature

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ABSTRACT

Background: Extra-axial cerebral cavernous hemangiomas particularly those found in the sellar region, are extremely rare. Their clinical manifestations and imaging characteristics can mimic those of a pituitary adenoma thus making preoperative diagnosis difficult. Few cases are reported in the literature. We present a case, along with a comprehensive review of the literature regarding specific aspects of diagnosis and management of all similarly reported rare cases.

Case Description: We present the clinical, radiological, and operative data of a rare case of a large intrasellar cavernous hemangioma in a 49-year-old female patient presented with headache and diminution of vision, which was diagnosed intraoperatively during an endonasal endoscopic transsphenoidal approach. Subtotal debulking was performed with immediate postoperative clinical improvement. The patient was then referred for radiotherapy and maintained her clinical improvement since then.

Conclusion: Neurosurgeons should consider this rare pathology in the preoperative differential diagnosis of sellar tumors. Bright hyperintense T2 signal with or without signal voids associated with centripetal delayed contrast enhancement in magnetic resonance imaging images might raise the suspicion which can be further confirmed intraoperatively with frozen sections. Due the reported high vascularity and intraoperative profuse bleeding leading to high operative morbidities, piecemeal subtotal resection followed by radiosurgery may be considered today as the safest and most effective strategy.

Keywords: Cavernous hemangioma, Radiosurgery, Sellar, Transsphenoidal

INTRODUCTION

Cerebral cavernous hemangiomas (CCHs) are benign vascular lesions with an incidence of 0.5% among all individuals.^[2] Although they can occur in all parts of the nervous system, these lesions mostly occur in the cerebral hemispheres.^[19] Approximately 50–80% of CCH are asymptomatic and are usually found incidentally on magnetic resonance imaging (MRI).^[28] However, clinically affected patients might present with headaches, epileptic seizures, intracranial hemorrhage, or focal neurological deficits.^[14] Stereotactic radiosurgery is currently widely used in combination with surgery as a standard of care.^[4]

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Seller cavernous hemangioma (SCH) is a rare pathology, with only 16 cases reported in the literature so far [Table 1]. In this report, we describe a case of SCH in accordance with SCARE criteria.^[1] We also describe the surgical approach used to treat

Table 1: Summary of the clinical and surgical data of previously reported cases of intrasellar cavernous hemangioma in the literature.

Year	Age/gender	Presenting symptom	Radiology modality/lesion extension	Surgical Approach	Extent of resection	Reported results
1965 ^[17]	46/F	H/A, reduced VA, right ophthalmoplegia	Cerebral angiography/sellar and right parasellar	Bifrontal craniotomy	Subtotal resection	Patient died 11 days after surgery
1980 ^[26]	72/F	Double vision, metastatic breast cancer	Autopsy finding/sellar and left parasellar	Autopsy findings	-	-
1984 ^[5]	50/M (1 st surgery at age of 42)	H/A, Seizures, reduced VA, hypopituitarism, impotence	Cerebral angiography, pneumoencephalography/sellar, suprasellar, and left parasellar	Subfrontal craniotomy (in both surgeries)	Total resection	Re-operated after 8 years for recurrence
1991 ^[22]	45/F	H/A, reduced VA, patient had long history of neurofibromatosis	CT and cerebral angiography/sellar and left parasellar	Pterional craniotomy	Subtotal resection	patient died intraoperatively from transcalvarial brain herniation
1991 ^[7]	48/M	H/A, reduced VA,, BTH	CT and cerebral angiography/sellar suprasellar and right parasellar	Subfrontal craniotomy	Biopsy	
1994 ^[20]	41/F	Amenorrhea and galactorrhea. Hyperprolactinemia.	MRI and CT and cerebral angiography/sellar and left parasellar	SLT	Subtotal resection	
2001 ^[10]	41/M	Asymptomatic patient known to have left orbital hemangioma	MRI/right part of the sella	SLT	Total resection	SAH and delayed CSF leak, VP shunt inserted 10 days after surgery
2004 ^[16]	63/M	Reduced VA, BTH, hypopituitarism, Hyponatremia,	MRI and cerebral angiography/sella and left parasellar	EET	Subtotal resection-	Hyponatremia improved
2006 ^[8]	62/F	Diplopia, partial oculomotor palsy	MRI and cerebral angiography/sellar and right parasellar	TST	Subtotal resection	Diplopia improved.
2013 ^[30]	32/F	Blurred vision	MRI/Sella and suprasellar	EET	Subtotal resection	Transient diabetes insipidus
2014 ^[21]	-	H/A, loss of libido, blurred vision	MRI/sella and left parasellar	EET	-	-
2017 ^[32]	64/F	H/A, reduced VA, BTH hyperprolactinemia	MRI/sellar and right parasellar	TST	Subtotal resection	Improved VA,
2018 ^[11]	66/F (1 st surgery at age of 39)	H/A, BTH	MRI/sellar and right parasellar	Subfrontal craniotomy in 1 st surgery EET in 2 nd surgery	Subtotal resection	-
	48/F (1 st surgery at age of 44)	H/A, galactorrhea	MRI/sella and left parasellar	SLT in 1 st surgery EET in 2 nd surgery	Subtotal resection	-
2018 ^[3]	43/M	H/A, impotence, reduced VA, BTH, and 6 th nerve palsy	CT and MRI/sellar and suprasellar	EET	Subtotal resection	
2018 ^[9]	49/M	H/A, Diplopia, ptosis	CT/sella and left parasellar	EET	Subtotal resection	

H/A: Headache, VA: Visual acuity, BTH: Bitemporal hemianopia, SAH: Subarachnoid hemorrhage, VP: Ventriculoperitoneal shunt, SLT: Sublabial transsphenoidal, EET: Endoscopic endonasal transsphenoidal, TST: Transseptal transsphenoidal, CT: Computed topography

this lesion as well as the histopathological characteristics, on which the diagnosis was established.

PRESENTATION OF THE CASE

A 49-year-old female was referred to our hospital as a case of a large symptomatizing nonfunctioning pituitary macroadenoma for surgical management. On thorough clinical evaluation, the patient reported that she has been complaining of chronic bitemporal headache, unexplained easy fatiguability, and a very slowly progressive diminution of vision over the previous 2 years. She also reported having high blood pressure which was adequately controlled by medications. She reported no history of any surgical intervention or any familial diseases. On examination, cranial nerves assessment was unremarkable except for her vision problem. Detailed ophthalmological examination revealed decreased visual acuity in both eyes, more on the left, with bitemporal homonymous hemianopia. Endocrinological assessment and biochemical tests revealed central hypothyroidism which was managed by levothyroxine replacement therapy. Computed topography imaging of the brain showed a slightly hyperdense large sellar/suprasellar mass extending to the cavernous sinuses on both sides with the right parasellar extension. The lesion was expanding the bony sella turcica with indentation and remodeling of the upper clivus [Figure 1].

MRI of the sellar area with intravenous contrast administration showed large sellar/suprasellar/parasellar mass, with heterogenous low signal in T1 images, intermediate high signal in T2 images, central small areas of diffusion restriction in DWI images, and mild patchy enhancement in postcontrast images. It measured approximately 36x41x50 mm on anteroposterior, cephalocaudal, and transverse dimensions, respectively. The lesion was completely encasing the cavernous and clinoid segments of the internal carotid arteries on the right side while partially encasing the right supraclinoid and the left cavernous segments. The lesion also exerted mass effect on the adjacent structures in the form of elevation of the tuber cinereum, and the floor of the third ventricle, with the intracranial segments of the optic nerves and optic chiasm being stretched over and inseparable from the lesion. The pituitary gland and the pituitary stalk could not be visualized [Figure 2].

An endoscopic endonasal transsphenoidal (EET) route was used to approach the lesion with the cooperation of a head and neck surgeon. During opening of the sellar floor, a dark red and firm lesion was encountered that seemed to be of vascular nature. Excessive bleeding was encountered on attempt to debulk the lesion. An intraoperative frozen section biopsy confirmed the vascular nature of the lesion and excluded a pituitary pathology. Considering the unknown nature of the lesion, its high vascularity and propensity for

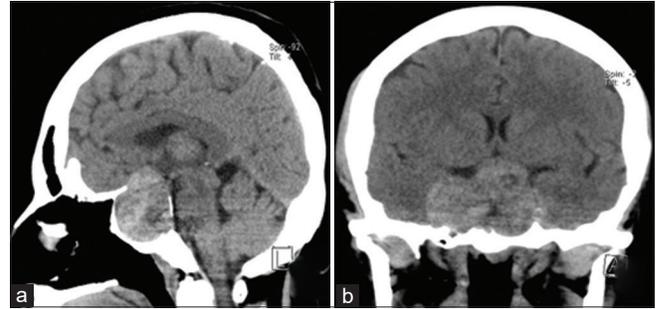


Figure 1: Preoperative CT images. (a) Sagittal nonenhanced CT scan of the brain demonstrating a large sellar/suprasellar hyperdense lesion with superior extension compressing the floor of the 3rd ventricle and expanding the sella turcica and remodeling the upper clivus. (b) Coronal nonenhanced CT scan of the same lesion demonstrating the parasellar extension of the lesion which is more to the right side.

bleeding, the decision was to perform a subtotal resection of the lesion aiming for mass effect reduction which was achieved without complications. Special attention was directed to obtain complete hemostasis. A subsequent histopathological examination confirmed the diagnosis of CCH [Figure 3].

Clinical reassessment during the postoperative period revealed marked improvement of the patient's visual acuity and field. Follow-up MRI showed adequate decompression of the optic chiasm and nerves despite a large intrasellar residual lesion [Figure 4].

The patient was offered a second stage surgery to attain a more radical excision, but she was satisfied with the improvement in her vision and refused. The case was thoroughly discussed in a multidisciplinary team meeting and the patient was referred for stereotactic radiosurgery. At the follow-up visit after 6 months, the patient is still maintaining her clinical improvement.

DISCUSSION

CCHs were initially classified as rare vascular lesions but have recently become an exceeding finding in multiple locations, especially with recent advances in imaging technology.^[29] They are known to reside anywhere within the neuroaxis and can be found in either the intra- or extra-axial regions. Most extra-axial cerebral cavernous hemangiomas reside in the middle cranial fossa with tendency to involve the cavernous sinus.^[18] However, extra-axial CCHs in the sellar region are considered extremely rare. The literature shows very slight female predominance with an average age of 47 years at first presentation [Table 1]. The term SCH was first used in 1980 by Wolfsberger *et al.* to describe an incidentally discovered on autopsy of a female who died of breast cancer.^[26] SCH can mimic the same clinical features of any other tumoral,

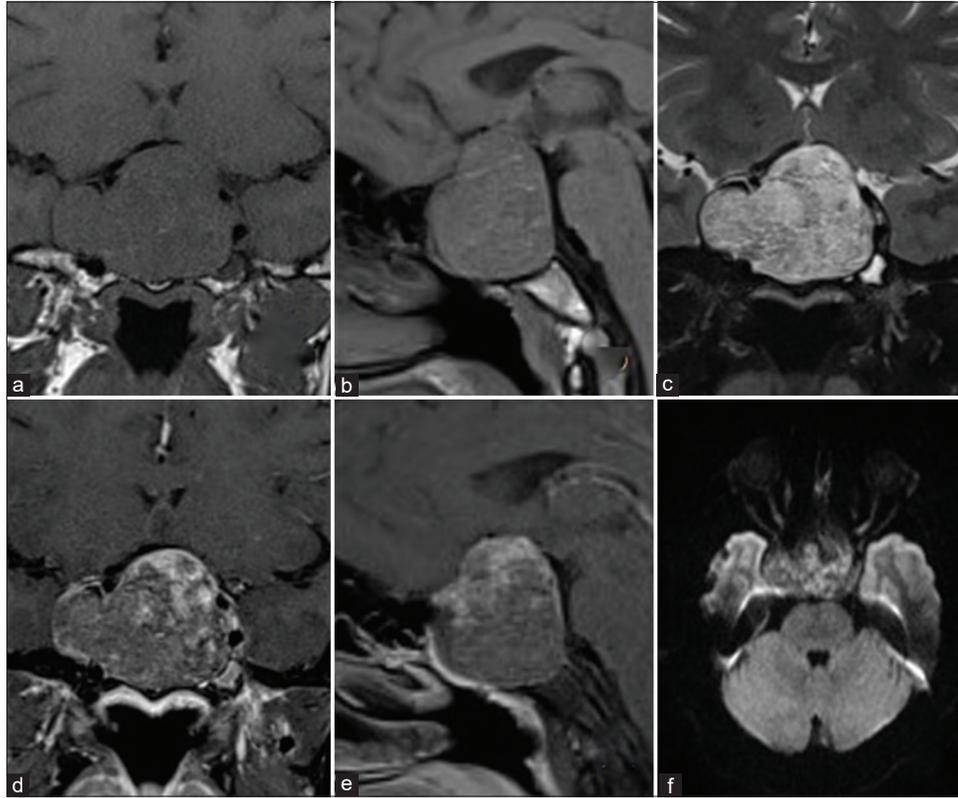


Figure 2: Preoperative MRI images. (a) Coronal T1, (b) sagittal T1, (c) coronal T2, (d) coronal post contrast, (e) sagittal post contrast, (f) axial DWI images of the lesion showing heterogeneous low T1 signal, intermediate high T2 signal, patchy post contrast enhancement, with central areas of diffusion restriction.

vascular, and inflammatory lesion affecting this central region of the skull base.^[31] Cranial nerve palsies caused by cavernous sinus extension or secondary visual impairment due to optic chiasm compression are common clinical findings.^[6] All these clinical manifestations, however, are not characteristic enough to help in the differentiation between sellar cavernous hemangioma from other pathologies more commonly found in the sellar and parasellar regions such as pituitary adenomas, meningiomas, cysts, aneurysms, and Schwannomas.^[3]

In addition, there are no pathognomonic MRI features that can confidently distinguish these lesions preoperatively.^[31] Thus, radiologists and surgeons should be aware of the possibility of SCH when they identify a sellar mass whose characteristics are consistent with those of a pituitary lesion as for pituitary macroadenomas.^[30] However, some MRI features have been reported in the few case studied in the literature which might assist in raising the suspicion of SCH. On MRI, SCH is usually with well-defined borders; brightly hyperintense on T2WI images and might show either of two patterns of contrast enhancement – a rapid homogeneous enhancement or a progressive centripetal enhancement. Centripetal enhancement is where contrast

fills the peripheral part of the lesion in the early stage of dynamic scanning (routinely performed with sellar MRI to detect pituitary adenomas) and then slowly fills the entire lesion in a pattern similarly seen in extra-cranial cavernous hemangiomas as those in the liver^[15,27]. Furthermore, infrequently the T2 signal characteristics of SCH enclose few signal void areas suggesting its vascular nature^[31]. To be noted that although cavernous hemangiomas originally arising from the cavernous sinus are slightly more common, but their growth pattern is differentiated from SCH with cavernous sinus extension by measuring the major portion of the tumor mass.^[30] As in the presented case, the main portion of the hemangioma was located in the sella, and thus, its sellar origin can be verified.

Multiple surgical approaches including pterional and subfrontal craniotomies, or sublabial, transseptal, and endoscopic endonasal transsphenoidal approaches have reportedly been utilized for the treatment of such lesions [Table 1]. Since the mid 90s, all the reported cases [Table 1] had been approached transsphenoidally with the EET technique being the most preferred one. We also advocate performing an EET approach, which has recently become a standard of care for sellar lesions and allow easier access to

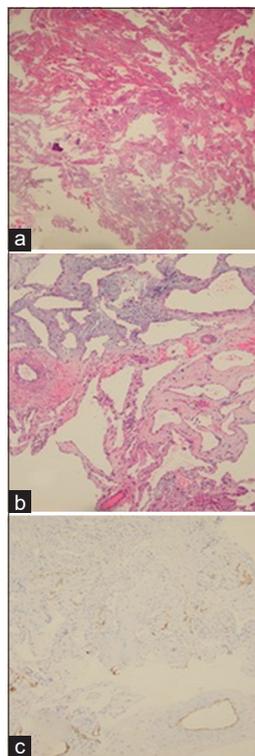


Figure 3: Histopathological slides images. (a and b) Sections from the specimen reveal a neoplasm composed of multiple large vascular channels lined by endothelial cells and separated by loose fibrous tissue with focal lymphocytic infiltrate. Most of them contain red blood cells in their lumina. Few areas of fibrosis and focal whorling is noted. No pituitary gland tissue seen. No evidence of malignancy in the examined biopsy material. (c) CD34 immunohistochemical stain highlighting the small capillaries.

and more feasible debulking of sellar masses, especially in cases, in which lesions likely extend to the suprasellar cistern and/or invade the cavernous sinus. Operative findings seem to be universal among the reporting operating surgeons, describing a firm dark reddish vascular lesion, adherent to surrounding structures, which bleed profusely. If a cavernous hemangioma is suspected, the surgeon may first wish to obtain a frozen section for diagnosis.^[10]

As the sellar cavernous hemangioma are vascular lesions and postulated to be dural based,^[10] significant hemorrhage commonly occurs intraoperatively during attempted resection.^[7,11,16,22] This especially can cause poor endoscopic visualization during surgery. From the 15 reported operated cases in the literature, total resection was achieved in only two cases. For this reason, if intrasellar cavernous hemangioma is suspected or confirmed by frozen section, only subtotal piecemeal resection should be attempted, and particular attention should be paid to obtain complete hemostasis.

Managing residual cavernous hemangioma with adjunctive treatments, such as stereotactic radiosurgery, after confirming

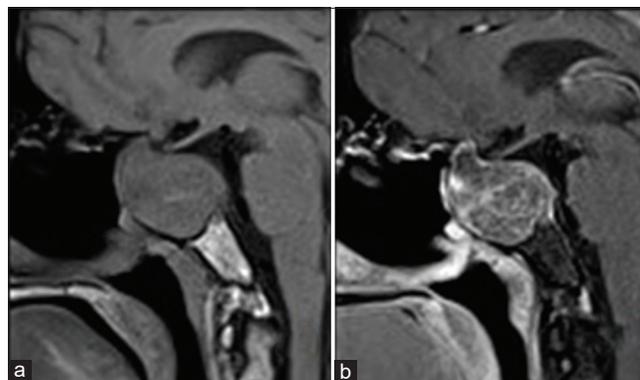


Figure 4: Postoperative MRI images. (a) Sagittal precontrast and (b) postcontrast T1-weighted image, of the immediate postoperative MRI images showing adequate optic nerves decompression despite a large residual sellar cavernous hemangioma

the diagnosis with the surgical biopsy can achieve an excellent outcome and avoid additional morbidities.^[23] Radiotherapy has been successfully used both pre- and postoperatively and is recently considered as an effective treatment resulting in a mean reduction of 54% of tumor volume.^[25] Due to the excellent results of radiation treatment and due to the high chance of subtotal resection, surgery as a definitive management modality is controversial.^[9,12] The reported risks of sellar cavernous hemangiomas treatment with surgical reduction and adjuvant radiosurgery compare favorably with the risks of recurrent hemorrhage from a vascular brain tumor.^[13,24]

CONCLUSION

Sellar cavernous hemangioma is rare vascular lesions that can present with clinical picture and radiological features mimicking pituitary adenomas. Neurosurgeons should consider this rare pathology in the preoperative differential diagnosis of sellar tumors. Bright hyperintense T2 signal with or without signal voids associated with centripetal delayed contrast enhancement in MRI images might raise the suspicion which can be further confirmed intraoperatively with frozen sections. Due the reported high vascularity and profuse bleeding, piecemeal subtotal resection aiming for neural tissues decompression followed by radiotherapy may be considered today as the safest and most effective strategy.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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