Surgical Outcome of Large Solid Posterior Fossa Hemangioblastoma without Preoperative Embolization

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Abstract

Background Large solid hemangioblastoma in the posterior fossa has an abundant blood supply as an arteriovenous malformation. The presence of adjacent vital neurovascular structures makes them vulnerable and difficult to operate. Complete surgical resection is always a challenge to the neurosurgeon.

Material and Method We share the surgical difficulties and outcome in this case series of large solid hemangioblastomas without preoperative embolization as an adjunct. This study included five patients (three men and two women, with a mean age of 42.2 years). Preoperative embolization was attempted in one patient but was unsuccessful. All the patients have headache (100%) and ataxia (100%) as an initial symptom. A ventriculoperitoneal shunt was inserted in one case before definite surgery due to obstructive hydrocephalus. The surgical outcome was measured using the Karnofsky Performance Status (KPS) score.

Result The tumor was excised completely in all the cases. No intra- and postoperative morbidity occurred in four patients; one patient developed transient lower cranial nerve palsy. Mean blood loss was 235 mL, and no intraoperative blood transfusion was needed in any case. The mean follow-up period was 14.2 months. The mean KPS score at last follow-up was 80.One patient had a KPS score of 60.

Conclusion Our treatment strategy is of circumferential dissection followed by en bloc excision, which is the optimal treatment of large solid hemangioblastoma. The use of adjuncts as color duplex sonography and indocyanine green video angiography may help complete tumor excision with a lesser risk of complication. Preoperative embolization may not be needed to resect large solid posterior fossa hemangioblastoma, including those at the cerebellopontine angle location.

Keywords
- solid hemangioblastoma
- circumferential dissection
- embolization

Introduction

Hemangioblastoma is a benign, highly vascular central nervous tumor, most likely seen in the fifth to sixth decades of life, accounting for 1.5 to 2.5% of intracranial tumors and 7 to 10% of posterior fossa tumors.[1–3] It commonly has a cyst with a small nodule in most cases of posterior fossa location (most commonly in the cerebellar hemisphere), whereas tumors with solid consistency are common in the brainstem, cerebellar vermis, and spinal cord.[2,4] In the posterior fossa, the frequency of hemangioblastoma is as follows: cerebellum (70%), brainstem (24.3%), cerebellopontine angle (CPA; 1.8%), craniocervical junction (1.6%), and very rarely in the fourth ventricle.[1,3,5]

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The term "large" is usually used to described a hemangioblastoma of ~3 cm in size, whereas the term "giant" is used for a tumor greater than 4 cm.[5,6] A hemangioblastoma is usually sporadic, although it is associated with the Von Hippel–Lindau (VHL) disease in 20 to 30% of the cases, where multiple tumors may be present.[1,4] Male dominance is seen in sporadic cases, whereas in the case of VHL disease, female predominance is seen with early age of onset.[1] VHL disease usually shows multiple lesions, but there are very few reports of hemangioblastomatosis (disseminated hemangioblastoma) with the sporadic disease.[7]

Large solid hemangioblastoma behaves like an arteriovenous malformation (AVM), and it is a challenge to operate on these tumors because of their high vascularity and their large sizes, which add complexity for the surgical excision of these tumors in the posterior fossa.[4,8] The surgical principle with circumferential dissection and en bloc excision has been followed for the large solid hemangioblastoma, but many of the studies have elaborated on the role of preoperative embolization followed by piecemeal excision of the tumor.[5,9–11]

The main limitation of embolization in such large hemangioblastoma in the posterior fossa is that the tumor may share the same supply as a normal neural structure or have multiple large artery feeders, so it may not be feasible to use embolization.[2] The risk of cerebellar infarction and intratumoral bleeding causing increased peritumoral edema with resulting brainstem compression may be devastating in such large solid hemangioblastoma.[4,10,12]

We share our experience of a case series of large solid hemangioblastoma and their surgical outcome without using preoperative embolization as an adjunct.

Patients and Method

Study Design

This study is a case series that is retrospective in design. All the large solid posterior fossa hemangioblastoma cases were reported from a single center and operated on in the division of neurosurgery, Raipur, India, between 2018 and 2020. Only large, purely solid hemangioblastoma (size >3 cm) were included. All the cases have been evaluated for the VHL disease using clinical criteria, as discussed by Van Leeuwaarde et al,[13] and they were all negative.

Patients

The patient charts containing demographical, radiologic, surgical findings, and clinical status were reviewed at the last follow-up. The patient's age, gender, presenting symptoms and neurologic examination, tumor size and location, intraoperative blood loss, extent of resection, postoperative complications, and the last follow-up examination were recorded and illustrated in ►**Table 1**.

Preoperative Considerations

Since preoperative diagnosis for hemangioblastoma was not clear, digital subtraction angiography (DSA) was not done in all the cases. We attempted embolization in one patient, but we were unsuccessful because of the inability to negotiate the microcatheter into the feeding artery. A ventriculoperitoneal (VP) shunt was inserted in one case before the definitive surgery for preoperative obstructive hydrocephalus. A surgical procedure was preferred as per the location, and midline suboccipital craniotomy and retromastoid craniotomy approaches were considered.

Surgical Technique

The character of solid hemangioblastoma is like AVM, and it requires individually tailored approaches to achieve adequate surgical exposure. We used somatosensory evoked potential (SSEP), motor evoked potential (MEP), and cranial nerve monitoring during the resection of hemangioblastoma of the cerebellum or the CPA.

We follow the surgical technique for our solid hemangioblastoma as the principle of surgery for AVM. The blood supply of hemangioblastoma is mainly derived from meningeal branches and intracranial feeding arteries. In the initial

Table 1 Summary of the clinical characteristics of five patients with large solid hemangioblastoma

Case	Age/sex	Clinical presentation	Tumor location/ tumor size	Surgical procedure	Intraoperative blood loss (mL)	Outcome (Karnofsky Performance Score) at last follow-up	Follow-up period (mo)
1	31 y/male	Headache with raised ICP features; Cerebellar ataxia	Left cerebellar hemisphere/ 4.6 × 4.3 × 3.4 cm	Total removal	250	80	17
2	45 y/female	Headache with raised ICP features; Cerebellar ataxia	Right CPA and cerebellomedullary cistern/3.4 × 2.8 cm	Total removal	400	60	3
3	59 y/male	Headache; Cerebellar ataxia	Right cerebellar/ 3.7 × 3.4 cm	Total removal	200	80	18
4	40 y/male	Headache; Cerebellar ataxia	Right cerebellar/ 3.1 × 2.7 cm	Total removal	150	90	24
5	36 y/female	Headache; Cerebellar ataxia	Left cerebellar/ 3.2 × 2.4 cm	Total removal	175	90	9

Abbreviations: CPA, cerebellopontine angle; ICP, intracranial pressure.

stages of surgery, these feeding arteries can be divided, and the draining vein should be preserved till the end of the surgery. Therefore, intraoperative color duplex sonography and intraoperative near-infrared indocyanine green video angiography (ICG-VA) have a detrimental role in distinguishing feeding arteries from draining veins. After distinguishing these with the help of these adjuncts, we gradually divide the feeding artery around the tumor to diminish the tumor's blood supply and thus shrink the tumor circumferentially. In the end, the draining vein is cut and coagulated, and tumor is removed en bloc. Like the AVM, venous drainage must be preserved until complete devascularization of the tumor is achieved, as intraoperative hemorrhage and diffuse tumor swelling with resulting brainstem compression may complicate removal of the lesion.

Registration and Ethics

The ethics committee approved this study with the understanding that no patient identity will be disclosed, and informed consent was taken from all the participants.

Result

Patients Characteristics

In the study, three men and two women were present with a mean age of 42.2 years (range: 31–59 years). All the cases were presented with headache and cerebellar ataxia. The tumor size of all the five cases was 3 cm^3. Four patients had a lesion in the cerebellum and one patient in the CPA and the cerebellomedullary cistern.

Case Illustrations

Case 1

A 31-year-old man presented at our institute with a severe headache with vomiting, difficulty walking, and blurring vision for the last 6 months. On physical examination, papilledema with asymmetrical field defects and left cerebellar signs, including nystagmus, were present. No features of VHL disease were present on clinical examination. Head computed tomography (CT) and magnetic resonance imaging (MRI) showed a $4.6 \times 4.3 \times 3.4$ cm T1 hypointense, T2, and fluid-attenuated inversion recovery (FLAIR) hyperintense lesion with contrast enhancement with fourth ventricle effacement and compression of the brainstem (►**Fig. 1**). No features of calcification or hemorrhage were present. DSA was done, showing a hypervascular tumor fed by the left superior cerebellar artery (SCA), anteroinferior cerebellar artery (AICA), and small tributaries from left the posteroinferior cerebellar artery (PICA) (►**Fig. 1**). Our neuroradiology team attempted embolization, but the microcatheter could not negotiate; hence, the procedure was abandoned. Complete excision of the tumor was done using midline suboccipital craniotomy and posterior arch of C1 removal. The

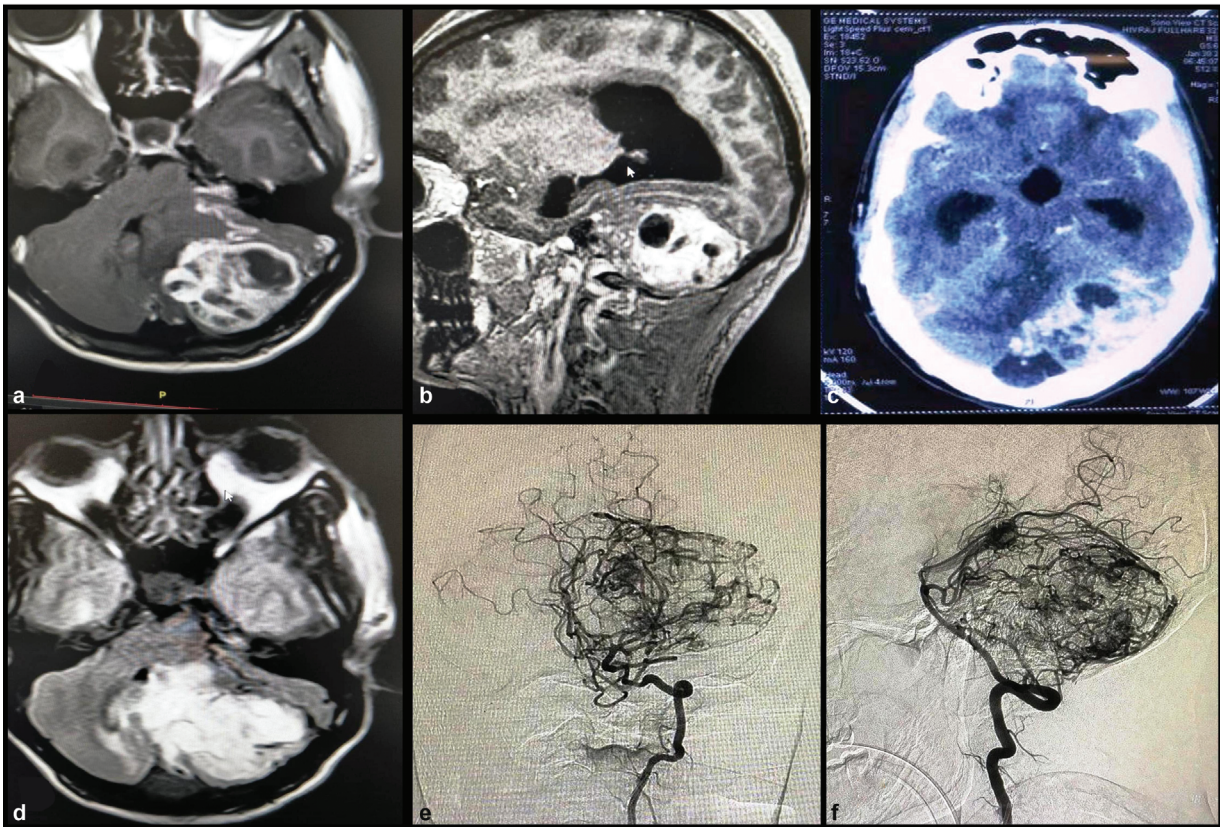

Fig. 1 Illustrative case 1. Magnetic resonance (MR) imaging (**a**,**b**,**d**) showing a large heterogeneously contrast-enhancing predominately solid lesion with dilated tortuous venous supply in the left cerebellum. (**c**) Contrast-enhanced computed tomography (CT) scan confirm the hyperdense lesion. (**e**) Anteroposterior and (**f**) lateral digital subtraction angiography (DSA) images show the feeders from superior cerebellar artery (SCA) and anteroinferior cerebellar artery (AICA) with dense tumor blush suggesting hypervascular lesion.

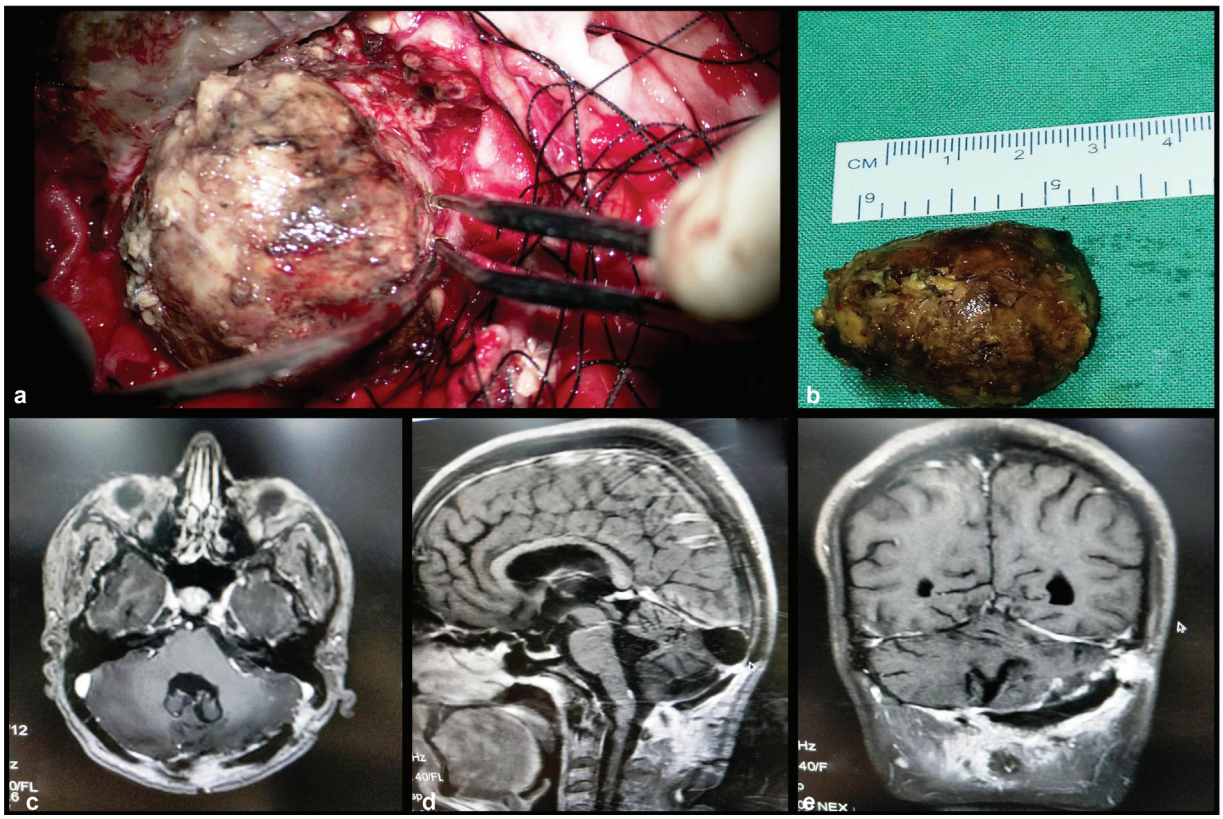

Fig. 2 Illustrative case 1. (**a**) Intraoperative image showing circumferential dissection with (**b**) complete en bloc excision. (**c–e**) Post-operative magnetic resonance imaging (MRI) confirms the complete excision of the tumor.

postoperative neurologic examination did not reveal any neurologic complications.

Histologic examination revealed typical hemangioblastoma consisting of numerous vessels, capillary mesh, and stromal cells. At the 6-month follow-up, the patient was asymptomatic. Follow-up MRI showed complete excision of the tumor (►**Fig. 2**).

Case 2

A 45-year-old woman presented with severe headache, blurring of vision, and difficulty walking for the last 5 months. On physical examination, papilledema and cerebellar signs were present but without cranial nerve deficits. No features of VHL disease were found. MRI wed a T1 iso-hypointense, T2 hyperintense lesion (3.4×2.8 cm) with homogenous contrast enhancement in the right cerebellomedullary cistern and the CPA with significant compression of the brainstem and hydrocephalus (►**Fig. 3**). The patient was taken for a left VP shunt in an emergency, and definitive surgery was done 2 days after the shunt procedure. Modified retromastoid suboccipital craniotomy was done in the park-bench position, removing the foramen magnum ring, posterior arch of C1, and part of the condyle. Gross total excision was done using circumferential dissection and en bloc excision (►**Fig. 4**). The patient developed lower cranial nerve palsy in the immediate post-operative period, which was managed by the nasogastric tube feeding. It gradually improved in 3 months of follow-up. Immediate post-operative CT imaging showed gross total excision of the tumor (►**Fig. 4**). Histopathology confirms the hemangioblastoma.

Outcome and Follow-up

There was no requirement of intraoperative blood transfusion, and mean blood loss was ~235 mL. One patient developed transient lower cranial nerve palsy in the postoperative period, whereas the other four patients had no postoperative complications. Gross total excision was achieved in all the cases, confirmed in the follow-up imaging. The mean follow-up period was 14.2 months. At the last follow-up, the mean KPS score was 80. Only one patient had a KPS score of 60.

Discussion

Hemangioblastomas are intra-axial, slow-growing, benign tumors, and with unknown histogenesis.[5] These are usually intra-axial tumors, but these tumors' unusual locations, such as CPA, may be explained by the exophytic growth of the intrinsic hemangioblastoma or origin from the vessels around the seventh to eighth nerve complex.[4] The uniqueness of hemangioblastoma is their abundant blood supply, which multiplied to several times in solid and large lesions, matching with an AVM; hence, the risks of intraoperative bleeding and postoperative complication are very high.[2,5,14] In earlier times, the mortality rate was as high as 50% in cases of solid type in operative series of Young and Richardson[14] and Okawara.[15]

The feature that differentiates hemangioblastoma from AVM is the true capillary network in hemangioblastoma. It may explain rare instances of preoperative bleeding in the hemangioblastoma, unlike AVM, despite its highly vascular

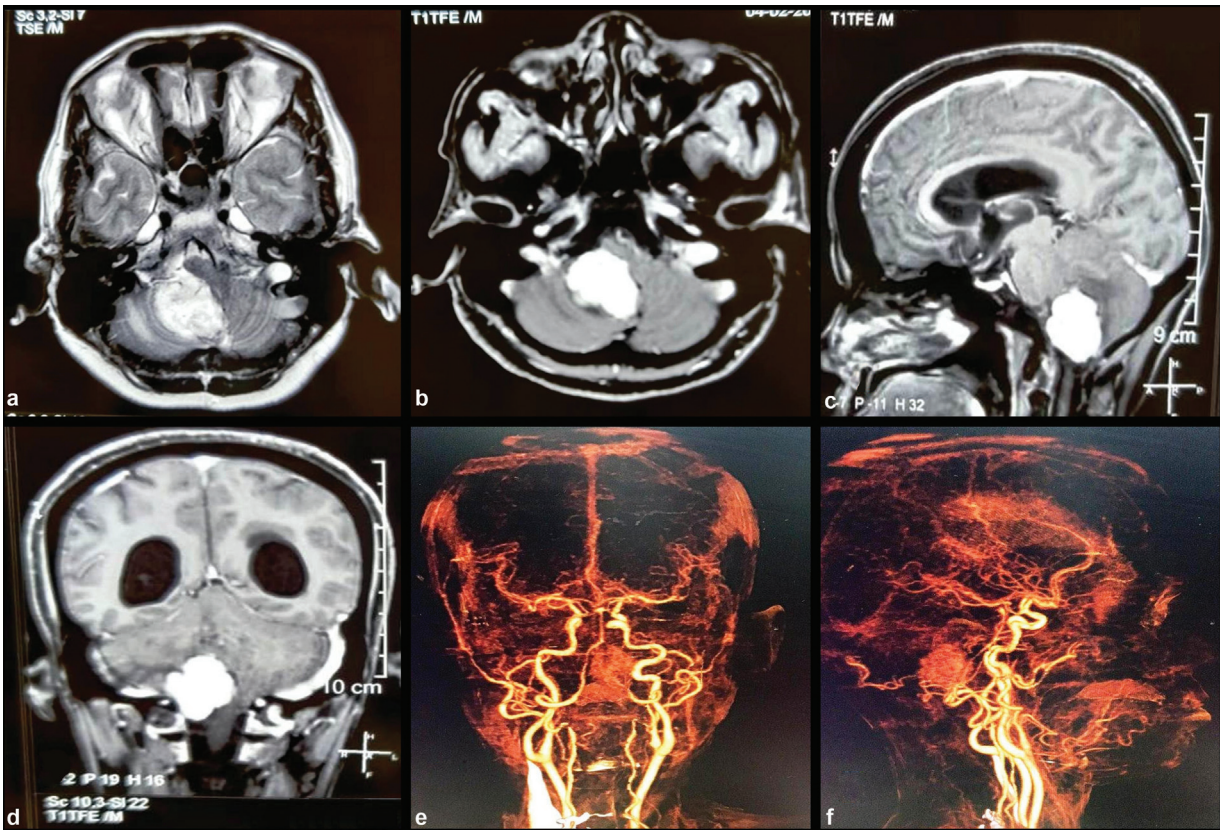

Fig. 3 Illustrative case 2. (**a–d**) Magnetic resonance imaging (MRI) shows a large hyperintense lesion in the right cerebellopontine cistern. It is extending into the cerebellomedullary cistern and with significant brainstem compression. (**e,f**) Computed tomography (CT) angiography confirms the hypervascular lesion with multiple feeders.

nature.[12] The risk of hemorrhage has also been studied in hemangioblastoma and increased age, large size, solid type, and spinal location are risk factors. Hemangioblastomas are usually supplied by the meningeal branches and intracranial feeding arteries, so careful coagulation of these feeding arteries is very important while dissecting the tumor. It is the key strategy for resecting these lesions with delayed tackling of the tumor's venous drainage. Intraoperative color duplex sonography helps identify the arterial feeders and excise them carefully.[2] Approaches to these large solid posterior fossa hemangioblastomas are individually tailored, but often standard procedures such as the midline suboccipital approach, retromastoid approach, transcondylar approach, and rarely translabyrinthine approach allow the safe removal.[5,8,16] The circumferential dissection and "en bloc" excision are the basic principles in such tumors, but it is still challenging to operate these tumors in the posterior fossa because of the narrow corridor and adjacent vital neurovascular structures.[3–5,17] The circumferential dissection has been supported by most of the studies,[2,5,18] whereas a few studies have favored multimodality treatment with preoperative embolization, followed by piecemeal excision of large solid hemangioblastoma.[6,9]

Intraoperative color duplex sonography and near-infrared ICG-VA are of utmost importance in large solid hemangioblastoma.[2,19–21] These adjuncts help identify early-filling arterial feeders first, followed by late-filling small feeders, and finally draining veins. Additionally, nonfeeding arteries can be identified with the help of ICG-VA.[19,20] We have used both these modalities in our case series, and we recommend these modalities to be used regularly in such hypervascular lesions as hemangioblastomas.

Usually, embolization works only in the cases with definite tumor-feeding arteries, seen in angiography, and not providing the normal blood supply to the adjacent vital neural structures.[2] The duration of preoperative embolization varies from 24 hours to a maximum of 3 weeks.[3,6,9] The superiority of onyx and n-butyl cyanoacrylate (NBCA) over the polyvinyl alcohol and other agents has been proven in various studies.[9,11,22] The risk of increased edema after embolization exists as small embolizing particles may drift from the arterial feeder and capillary network to large venous channels causing venous congestion and increased edema.[11,22] A large systemic review by Ampie et al compared both the embolization and nonembolization groups and hypothesized that there is no benefit of embolization. It did not increase the gross total resection rates, decrease the estimated blood loss, and the incidence of complications in the embolization group. Not only does embolization fail to mitigate surgical risks, but also the embolization itself carries a significant risk of complications (as the risk of hemorrhage increases to 8%, which is 1.6% in the nonembolized group). Therefore, embolization may not be considered mandatory for the management of intracranial hemangioblastoma.[10]

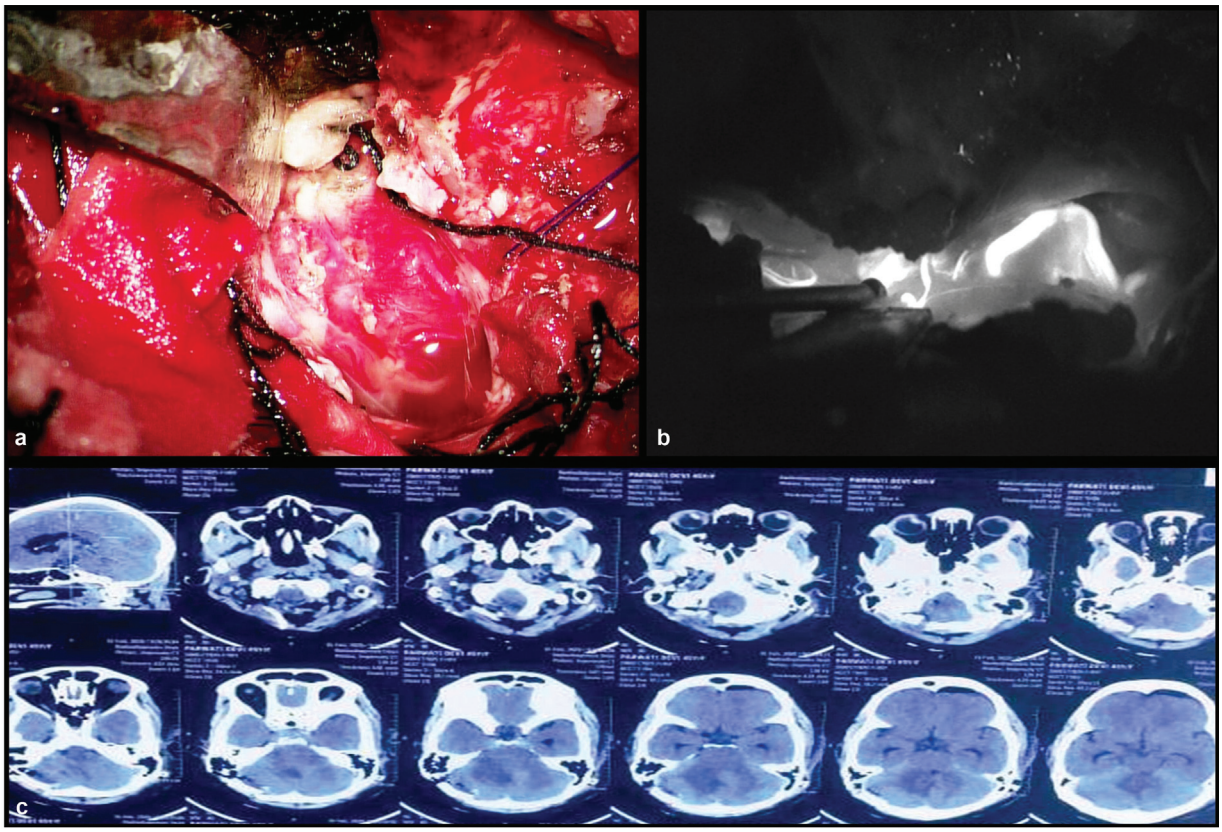

Fig. 4 Illustrative case 2. (**a**) Intraoperative picture showing a typical bright red cherry colored lesion with multiple dilated vessels. (**b**) Intraoperative indocyanine green videoangiography (ICG-VA) confirms the multiple small feeders with large dilated draining veins. (c) A postoperative computed tomography (CT) scan shows complete excision of the tumor.

Histopathologically, these tumors show a biphasic pattern with large vacuolated stromal cells and a dense capillary network. Histopathologic confirmation of solid hemangioblastoma also requires Immunohistochemistry (IHC) confirmation as lipid-filled vacuolated stromal cells may mimic clear-cell metastatic renal cell carcinoma and the Rosenthal fibers from adjacent reactive brain tissue may mimic pilocytic astrocytoma.[5] The IHC marker positive in hemangioblastoma, in decreasing order of frequency, was for vimentin, vascular endothelial growth factor, neuron-specific enolase, reticulin, CD 56, S-100, and inhibin, which all stained at more than 80%.[1,5]

Complete excision was achieved in more than 90% of the cases in recent studies.[18,23] Many clinical outcome factors have been studied in several studies, but tumor characteristic (solid type) is the most consistent factor in predicting the long-term clinical outcome in hemangioblastoma.[3,17] The tumor recurrence rate is 5 to 17% without VHL disease and around 17 to 75% with VHL disease.[3,17,18,23] Incomplete tumor resection because of multicentricity or tumor cells in the cystic wall or brainstem involvement may be the reason for recurrence of these tumors.[2] Cerebrospinal fluid (CSF) dissemination has been noted in rare instances as a cause of recurrence, even after complete excision of these tumors with only 14 cases reported till now.[14]

The most common complications were intracranial (31.5%), consisting of postoperative hemorrhage and hydrocephalus and pseudomeningocele formation.[1] Infection (3.15%) and cerebral ischemia (2.85%) were the other complications.[23] In CPA hemangioblastoma, facial nerve paresis followed by lower cranial paresis are the most troublesome complications and may be permanent.[3] In our series, we did not experience any postoperative complication, except a transient lower cranial nerve palsy in one case. Complete resection could be achieved in all the cases.

Since hemangioblastomas are radioresistant tumors, the role of radiosurgery is minimal in posterior fossa hemangioblastoma and are only employed in cases of residual, recurrent, multiple, or nonoperable cases.[4,5,18,23]

Strength and Limitations of the Study

Management of large posterior fossa hemangioblastoma is challenging, and there is a need for more evidence related to surgical outcomes. Our study targeted this particular patient group and we obtained good surgical results. One of the limitations of the study is the number of cases included in the series, which might be too small for precise conclusions and for challenging the role of embolization in their management. Additionally, this study includes a search of a considerable number of case series from the literature, which gives the readers a reasonably broad perspective about the role and outcomes of surgery in posterior fossa hemangioblastoma and questions the role of embolization in each case.

Conclusion

We propose the circumferential dissection with en bloc excision of posterior fossa solid hemangioblastoma and avoid

attempting intratumoral decompression. We also recommend adjuncts such as intraoperative color duplex sonography and ICG-VA regularly in large solid hemangioblastoma. We do not favor preoperative embolization as the role of embolization is not yet evident. The difficulty in obliterating the tumor's blood supply and the risk of obliterating adjacent vital structures' normal blood supply may be the limiting factors for embolization.

Statement of Ethics
All authors contributed equally to this work. All procedures performed in the study involving patients were following our institution's ethical standards, and any personal information and identity of patients were not disclosed in the study.

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None.

Conflict of Interest
None declared.

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