

## Review Articles

# Management Difficulties of Cerebeller Haemangioblastoma: A Case Report and Literature Review

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### Abstract

*The surgical removal of solid deep-seated haemangioblastomas remains challenging, because treatment of these lesions is often complicated by severe bleeding associated with the rich vascularity of this tumor and by severe neural tissue injury associated with the difficulty of en bloc resection. A 55 year-old man, was admitted to us with inability to stand & walk due to imbalance and altered level of consciousness. He has family history of Von Hippel Lindau (VHL) disease. He underwent microsurgery followed by gamma knife radiosurgery 13 years back for cerebeller haemangioblastoma. Magnetic resonance imaging (MRI) revealed intra-axial mass with multiple large cysts in cerebellum with severe compression on cerebellum and brainstem. Preoperative angiography revealed a hypervascular tumor in the posterior fossa, he underwent microsurgery through the previous midline suboccipital craniotomy involving the removal of the arch of C-1. Patient recovered slowly in post operative period.*

**Keyword:** Cerebeller haemangioblastoma, Von Hippel Lindau disease, Radiosurgery, Microsurgery

### Introduction

Cushing and Bailey used the term 'Haemangioblastomas' to describe tumors arising from the endothelial cells of the central nervous system.<sup>1</sup> Haemangioblastomas are solid or cystic benign vascular tumors that may arise anywhere in the body including central nervous system.<sup>2</sup> Though histologically benign, their development is often unfavorable due to high frequency of recurrence and multicentricity especially when occurring in a familial set-up i.e., Von Hippel Lindau (VHL) syndrome.<sup>3</sup> They account for 1.5-2.5% of all intracranial and 7-12% of posterior fossa tumors. In spite of some advancement of endovascular and radiosurgical therapy in the treatment of these tumours, they are rarely effective in isolated form; Microsurgical removal CNS haemangioblastomas is the treatment of choice, though it is often complicated and difficult, because of tumor hypervascularity and location. In addition, though surgical mortality, morbidity rates have been reduced, but still there may be mortality and morbidity, even with the advantages of modern microsurgical techniques.<sup>4,5,6,7</sup>

Here, we report on a male patient with recurrent haemangioblastoma in the cerebellum with Von Hippel Lindau disease who underwent partial microsurgical excision

followed by radiosurgery 13 years back. The patient was successfully treated by redo microsurgical excision.

### Case report

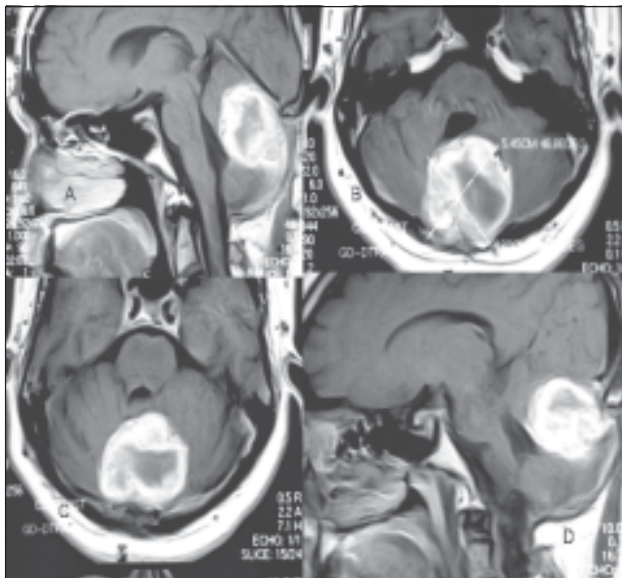
A seventy year old office executive presented to us with progressive imbalance of limbs and trunk for which he could not stand and walk for last few days along with altered level of consciousness. There was history of intention tremor and scanning of speech for last 14 years that were worsening for the last months. He had hiccough and vertigo. There was no history of visual or any other cranial nerves dysfunction. His family history was positive for Von Hippel Lindau disease. He underwent microsurgical operation of tumour + ventriculo-peritoneal (V-P) shunt for cerebeller haemangioblastoma in Singapore (1995). There was massive peroperative bleeding (nine units of blood transfused) for which operation could not be proceeded and about eighteen hours was needed for haemostasis followed by one month ICU support. Post operatively patient developed scanning of speech and intention tremor. Later on patient was referred for gamma knife radiosurgery to Japan (1995). After radiosurgery his condition was static with cerebeller dysarthria and intention tremor but he was just able to manage his office with difficulties. Higher psychic function were

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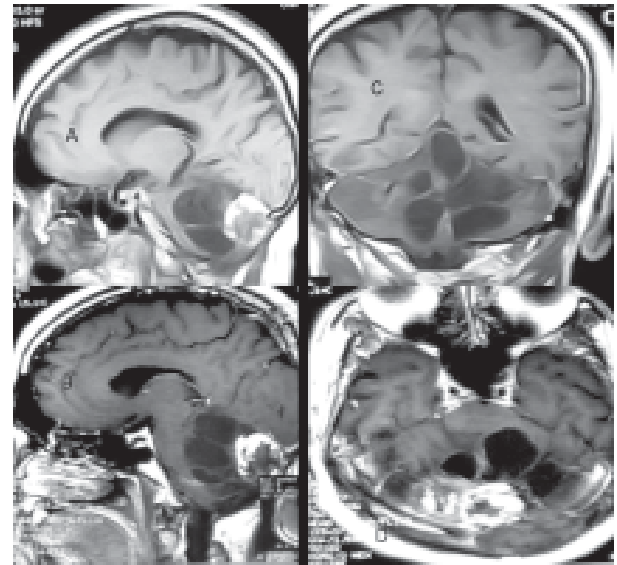
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normal except speech which was scanning cerebeller dysarthric type. Cranial nerves including fundoscopic examination were normal. His cerebeller signs and long tract signs were bilaterally positive. V-P shunt was functioning and abdominal examination revealed no abnormality. Preoperative MRI in 1995 showed highly vascular solid cerebeller haemangioblastoma(Figure-1A &1B). Postoperative and post radiosurgical MRI in 1997/98 persisted reduced size vascular tumour (Figure- 1C&1D) . MRI on 2008(before second operation) showed recurrent cerebeller tumour (vascular) with multiple big cyst almost occupying the whole posterior cranial fossa along with post radiosurgical changes and midline suboccipital craniectomy (Figure-2). Other parts of brain were free of tumour. Cerebral angiography showed posterior cranial fossa highly vascular tumour blush with surrounding avascular areas (cystic areas).Spinal MRI and abdominal ultrasonogram were normal. His Hb was 16gm%.

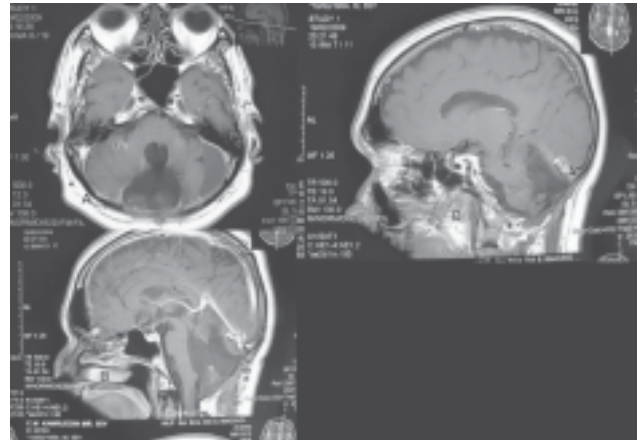
Peroperatively tumour was vascular , fibrous and adherent to tentorium & brain(due to radiosurgery).We excised the tumour along with excision of superficial cyst and marsupialization of deep cyst that was adhere to the brain stem. Postoperative period was uneventful .At the end of six months after operation patient can walk without support and write with some difficulties. Dysarthria persisted like that of preoperative state. Post operative(after second operation) contrast MRI showed complete excision of the tumor (Figure-3).



**Fig.-1:**A&B;MRI of brain, before first operation, saggital and axial section. C &D;MRI of brain axial and saggital section after first operation and radiosurgery.



**Fig.-2:**A&B;MRI of brain, before second operation, saggital section. C &D; MRI of brain coronal and axial section just before second operation.



**Fig.-3:** MRI of brain saggital sections (A&B) and axial section (C) after second operation.

**Discussion and literature review**

Haemangioblastomas may occur sporadically or as part of VHL complex.<sup>8</sup> the male/female ratio is closer to 1-1.5:1.<sup>8-18</sup> The average age at presentation was ranged from 38-49.5 years.<sup>9-17</sup> VHL syndrome is seen in 23-40% cases of cerebeller haemangioblastoma.<sup>10</sup> Our patient had VHL syndrome and he presented at the age of 41 years.

The triad of headache, vomiting and cerebellar symptoms has been the most common clinical features in literature.<sup>9-17</sup> The incidence of papilloedema has ranged from 39-70% and cerebellar involvement from 40-80%. Cranial nerve involvement seen in 17-35% cases.<sup>9-17</sup> This patient initially presented with Features of raised Intra Cranial Pressure(ICP) for which he underwent V-P shunt.

There may be polycythemia and is believed to be due to secretion of Erythropoietin-like substance by the neoplastic tissue. The average hemoglobin is 16-18 g% reported in literature.<sup>14, 17</sup> Before the last operation patient's Hb was 16g%.

Radiologically, 60-72% are cystic tumors and rest are solid.<sup>9-15</sup> Initially the tumour of our patient was solid but at recurrence it had cystic component (in formation of cystic component radio surgery might get some influence!) Tumors may in the cerebellar hemispheres accounted for 60% cases, vermian tumors in 28%, while brainstem (subpial) tumors is seen 12 % patients. Hydrocephalus is seen in 20% cases.<sup>14</sup>

In spite of some advancement of endovascular and radiosurgical therapy in the treatment of these tumours, they are rarely effective in isolated form; Microsurgical removal CNS haemangioblastomas is the treatment of choice, though it is often complicated and difficult, because of tumor hypervascularity and location. Complete excision is the goal of the surgery. The cyst wall need not be removed. The solid nodule tumor mass is removed microscopically by dissecting the gliotic margins away from the tiny vascular pedicles that are coagulated. To avoid intraoperative swelling, the large draining veins have to be preserved until the arterial feeders to the mural nodule have been isolated and resected. Piecemeal removal must be avoided to prevent catastrophic bleeding. Some authors have advocated the use of preoperative embolisation as an adjunct to microsurgical removal. Though radiosurgery can help in small, solid and difficult location, it is rarely curative and recurrence is most likely along with radiation change in brain, meninges and tumour itself that make tumour excision difficult that we faced in our case. Pan et al treated 20 haemangioblastomas with Gamma-Knife (GK).<sup>19</sup> Niemala et al treated 10 patients with 11 haemangioblastomas with GK.<sup>20</sup> They suggested that a solitary small or medium-sized haemangioblastomas usually shrink or stop growing after radiosurgery

The mortality rate in available literature has varied from 4-36%.<sup>8-23</sup> 15%-20% patients developed postoperative complications.<sup>8,10-17,22,23</sup>

The common postoperative morbidity was due to chest infection, lower cranial nerve paresis and meningitis.

Factors predicting a poor outcome are multiple haemangioblastomas, association with retinal haemangioblastomas and/or onset of disease at less than 30 years age.<sup>10</sup> Predominantly solid and midline tumors had a significantly higher mortality when compared to cystic and hemispheric tumors, which can be partly explained by their proximity to the brainstem.<sup>12,13,17</sup>

Dela Monte stated that recurrence was correlated with younger age (< 30 years) and VHL syndrome.<sup>24</sup> Also, cystic tumors were more frequent, had a longer survival while recurrence was less.<sup>11</sup> In literature, recurrence has ranged from 16-30%.<sup>8,10,12,22,23,24</sup>

**Conflict of interest:** None

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