CASE REPORT

Lateral ventricle haemangioblastoma: the role of perfusion scanning and embolisation in diagnosis and management

Ian Anderson,¹ Ramesh Kumar,² Tufail Patankar,³ Atul Tyagi¹

¹Department of Neurosurgery, Leeds General Infirmary, Leeds, West Yorkshire, UK

²Department of Neurosurgery, National University of Malaysia Medical Centre, Kuala Lumpur, Malaysia

³Department of Neuroradiology, Leeds General Infirmary, Leeds, West Yorkshire, UK

Correspondence to

Ian Anderson,
ian.anderson@leedsth.nhs.uk

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SUMMARY

We present the case of a 24-year-old man who presented with vertigo and right-sided weakness. Subsequent imaging demonstrated a lateral ventricle haemangioblastoma. This is the first case ever to be treated with surgical excision augmented by preoperative endovascular embolisation, as illustrated with perfusion CT scanning performed pre-embolisation and postembolisation. We present the case followed by a summary of the previously published cases and a discussion of the advantages of perfusion scanning and endovascular embolisation in these highly vascular (and therefore potentially dangerous) lesions.

small cystic components extending to involve the parietal periventricular region and medially the

BACKGROUND

We describe the first case ever of haemangioblastoma of the lateral ventricle to be treated with surgical excision augmented by preoperative endovascular embolisation, as illustrated with perfusion CT scanning (CTP) performed pre-embolisation and postembolisation.

A review of the available literature confirms this to be only the ninth case of lateral ventricle haemangioblastoma to have been described to date. This particular case also highlights the role of perfusion scanning in the preoperative differential diagnostic workup of this rare tumour.

CASE PRESENTATION

A 24-year-old, right-handed, Caucasian man presented with a sudden onset of vertigo and right arm weakness associated with transient slurring of speech and a right facial droop following a period of intense exercise. There was no medical history, except for a 4-month history of recurrent headaches, being treated as sinusitis. The patient's sister is known to have died from a brain tumour (of unknown type) at the age of 9 years. On physical examination, the patient was noted to have mildly decreased power of extension throughout the right upper limb, but no other neurological defect was present.

INVESTIGATIONS

A contrast-enhanced CT scan showed a relatively well-defined, diffusely contrast-enhancing intraventricular mass extending in the body of the corpus callosum and medial left frontoparietal region. Subsequent MRI demonstrated a large, heterogeneous, relatively well-defined intraventricular tumour, with a diffusely enhancing component and

Figure 1 T1-weighted imaging (WI) with gadolinium (A) and T2WI (B) MRI showing an intraventricular tumour, of part cystic and part solid composition and diffuse enhancement and a large calibre blood vessel within it.

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Figure 2 Preoperative CT-perfusion scans demonstrating an increased relative cerebral blood volume (A) and relative cerebral blood flow (B) within the tumour.

body of the corpus callosum (figure 1A, B). A whole spine MRI showed no evidence of metastases and a staging CT (thorax, abdomen and pelvis) was unremarkable.

DIFFERENTIAL DIAGNOSIS

It was felt that this lesion most likely represented a pilocytic astrocytoma and a midline frontal craniotomy and interhemispheric approach were performed with intention to resect, however the operating surgeon encountered an extremely vascular tumour with large calibre surface vessels; macroscopically more suggestive of haemangioblastoma. The decision was taken to postpone resection in order to embolise the tumour first. A

tumour wall specimen was sent for histology prior to closure and the initial frozen section report was pilocytic astrocytoma.

Given the suspicion that this may represent a more vascular lesion than a pilocytic astrocytoma, a CTP scan was performed (GE Medical Systems *CT Perfusion V4.3.1* software) which confirmed a substantial increase in relative cerebral blood volume (rCBV) of 37.6 within the tumour as well as an increase in relative blood flow of 13.5 (figure 2A, B).

TREATMENT

Subsequent cerebral digital subtraction angiography confirmed that this was indeed a highly vascular tumour, supplied by the pericallosal, anterior choroidal and posterior choroidal arteries.

Figure 3 Pre-embolisation (A) and postembolisation (B) digital subtraction angiography showing the relative reduction in tumour vascularity following endovascular embolisation.

Figure 4 Postoperative CT-perfusion scans demonstrating that the relative cerebral blood volume (A) and relative cerebral blood flow (B) within the tumour had reduced following endovascular embolisation.

The tumour bed was embolised with Histoacryl glue and Embospheres particles, before the posterior choroidal artery was distally occluded, in order to devascularise the inferior portion of the tumour completely (figure 3A, B).

A repeat CTP scan was performed several hours postembolisation which showed a 78% reduction in rCBV (from 37.6 to 8.1) and a substantial reduction in relative tumour blood flow (from 13.5 to 6.3; figure 4A, B).

The patient returned to the operating theatre 8 days postembolisation. The tumour was noted to be substantially less vascular than previously and was successfully debulked. There was still some bleeding, which was reported by the operating surgeon to be mainly venous.

OUTCOME AND FOLLOW-UP

Histopathological specimens demonstrated a tumour composed of sheets and nests of large polyhedral stromal cells with foamy cytoplasm in a rich vascular network. Stromal cells were positive for GFAP (<10%), S-100 (80%), CD56

(80%), KI-67 (<5%) and Synaptophysin (20%). They were negative for epithelial membrane antigen (EMA), renal cell carcinoma (RCC), myelinated nerve fibres, human melanoma black 45, protein Melan-A, neurofilament 52 and neuronal-specific nuclear protein. Tumour cells were also positive for neuron-specific enolase (NSE) and Vimentin, and negative for neurofilament protein (NFP). Delicate reticulin fibres were present around small nests in the tumour. A diagnosis of a WHO grade I haemangioblastoma was confirmed.

Postoperatively, the patient had weakness of the right upper and lower limbs. MRI showed tumour residuum in the posterior body of the left lateral ventricle. The decision was made to remove the remainder electively, this time via a transcortical approach. When resected, the tumour was noted to be relatively avascular, and there were no particular difficulties in achieving haemostasis. Further MRI demonstrated substantial excision of the remaining tumour but showed a possible small area of residuum at the edge of the thalamus. This has not changed in 3 years of follow-up imaging (figure 5).

Figure 5 Most recent MRI (T1-weighted imaging with gadolinium) images showing good tumour resection (A) and a small area of possible residuum (B) that has remained stable on serial follow-up scans.

Table 1 Comparison of previously reported cases of lateral ventricle haemangioblastoma

Reference	Patient	Presentation	Description of lesion			VHL association?	Management	Resection	Outcome
			Radiological	Macroscopic	Histological				
Vecchi and Patrassi ²	80 years, F	Incidental, at autopsy	NA	Multiple, solid tumours	NA	Unknown	NA	NA	NA
Diehl and Symon ³	20 years, M	Visual disturbance, headaches, papilloedema	<i>CT</i> : Contrast-enhancing lesion with large draining veins. <i>Angiography</i> : Tumour fed by anterior choroidal artery and tentorial branches	Single, solid, pulsatile, vascular tumour	Multiple, thin walled vessels of varying calibre. 'Foamy' stromal cells	Retinal angiomatosis, epididymal cyst	Surgery: Middle temporal gyral approach	Total macroscopic resection, confirmed with CT	Transient visual field defect. Later re-presented with a cerebellar lesion
Murakami et al ⁶	31 years, F	Headaches, gait disturbance, papilloedema	<i>CT</i> : Cystic cerebellar lesion and right intraventricular lesion	Multiple tumours	Haemangioblastomas	Family history	Surgery	Unclear	Unclear
Ho et al ⁴	44 years, F	Headaches, chronic depression, haemorrhagic change to left fundus	<i>CT</i> : Contrast-enhancing lesion. <i>MRI</i> : Contrast-enhancing lesion. <i>Angiography</i> : Tumour fed by posterior choroidal artery	Single, solid, reddish, vascular tumour	Multiple small vessels. 'Foamy' stromal cells	Family history	Surgery: posterior temporal lobe approach	Total macroscopic resection	Left homonymous lower quadrantanopia
Lyo et al ⁵	36 years, M	Headaches, diplopia, confusion, right abducens palsy	<i>CT</i> : Ring-enhancing solid lesion in the left lateral ventricle wall	Single, solid, vascular tumour	Well differentiated, highly vascular	None	Surgery: middle frontal gyral approach	Total macroscopic resection, confirmed with CT	Neurologically intact postoperative. No follow-up mentioned
Prieto and Roda ⁷	73 years, M	Confusion, dysphasia	<i>CT</i> : Contrast-enhancing lesion <i>MRI</i> : Heterogeneous lesion with central areas of hypointensity. Homogeneous enhancement with contrast	Single, solid, reddish, vascular tumour	Multiple small vessels, slightly stained cytoplasm	Renal cell carcinoma	Surgery: middle temporal gyral approach	Total macroscopic resection, confirmed with CT	Transient mutism
Jaggi et al ⁹	30 years, M	Headaches	<i>MRI</i> : Hypointense (T1WI), hyperintense (T2WI), contrast-enhancing lesion	Single, solid, reddish, vascular tumour	Multiple small vessels, slightly stained cytoplasm	None	Surgery: superior parietal transsulcal approach	Total macroscopic resection, confirmed with CT	Transient receptive dysphasia
Takeuchi and Takasato ⁸	33 years, M	Headaches	<i>MRI</i> : Multiple lesions: 3rd and lateral ventricles and cerebellum.	Part solid, part cystic tumour	Multiple small vessels. Multiple stromal cells	VHL gene. Spinal haemangioblastomas	Surgery (two stage)	Unclear	Mild memory disturbance. Shunt for hydrocephalus
Al-Najar et al ¹⁰	70 years, M	Headaches, vomiting, dizziness	<i>CT</i> : Intraventricular haemorrhage <i>MRI</i> : Isointense (T1WI), hyperintense (T2WI), heterogeneously enhancing	Not described	Cells arranged in nests with fibrovascular septae	None	Initial biopsy followed by surgical resection	Complete excision, unclear if postoperative imaging performed	Unclear

F, female; M, male; NA, not available; T1WI, T1-weighted imaging; VHL, von Hippel-Lindau disease.

At the clinic review 6 months postoperatively, the patient was able to mobilise with a stick and walk independently for short distances. He could write with his right hand but had some ongoing difficulties with fine movements. The patient is now 3 years following surgery and has full power in the upper and lower limbs; he is walking unaided and has returned to full-time employment.

DISCUSSION

Haemangioblastomas of the central nervous system are benign, highly vascular neoplasms, accounting for 2% of all intracranial tumours. They are associated with the von Hippel-Lindau disease in as many as 38% of cases.¹ These tumours typically occur infratentorially but are occasionally seen in the supratentorial compartment. The first documented case of lateral ventricle haemangioblastoma was an incidental postmortem finding, reported in 1935 by Vecchi and Patrassi.² Lateral ventricle haemangioblastomas are extremely rare; this is the ninth case of lateral ventricle haemangioblastoma to be recorded in the world literature and the eighth to have been worked up in vivo.^{3–10} Details of the previously reported cases are summarised in table 1.

Resection of haemangioblastomas is not without operative risk, largely on account of their potential for intraoperative haemorrhage. Previous authors have described the great difficulty in achieving haemostasis in these cases; indeed, there is a published report of one patient who died from postoperative bleeding following the surgical resection of a third ventricle haemangioblastoma.^{11 12}

We were able to utilise CTP to differentiate radiologically between haemangioblastoma and pilocytic astrocytoma. This is because the rCBV in haemangioblastoma is much higher than it is in pilocytic astrocytoma. The mean rCBV reported for haemangioblastoma is typically 11–26 compared with pilocytic astrocytomas, which have a mean rCBV of between 1 and 3.^{4 13–15} Although the measurement of rCBV has been described previously in the diagnoses of these tumours, it was not performed in the previous cases that have occurred in this rare location.

Learning points

- ▶ Although intraventricular haemangioblastomas are rarely encountered, the consequences of inadequate preparation and workup of these cases can prove fatal.
- ▶ Perfusion scanning is a useful tool to distinguish haemangioblastomas from other intraventricular tumours where the diagnosis might otherwise have been discounted.
- ▶ When combined with embolisation, perfusion scans may also be used to map the devascularised tumour in order to plan the surgical approach.
- ▶ Preoperative embolisation should be seriously considered in order to reduce the risk of intraoperative bleeding when dealing with these lesions.

The CTP findings ratified the surgeon's own observations, and therefore, despite the provisional frozen section histology report of pilocytic astrocytoma, endovascular embolisation was performed. Adjuvant endovascular embolisation of haemangioblastomas elsewhere in the brain and spine has occasionally been performed where a tumour is fed by large vessels. Complete or almost complete embolisation has been shown to reduce operative complications and morbidity, even for large tumours, although this does not seem to be the case where only partial embolisation is achieved.⁸

Comparison of pre-embolisation and postembolisation CTP images not only illustrated the marked tumour devascularisation that embolisation had achieved but also highlighted residual areas of high tumour blood volume, enabling the surgical team to perform a safer surgical approach to the lesion without large-scale blood loss.

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Competing interests None.

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