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CASE REPORT

Rupture of a flow aneurysm secondary to spontaneous extracranial to intracranial revascularisation in the posterior fossa following radiation-induced vasculopathy for cerebellar tumour

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Abstract

Paediatric patients receiving cranial irradiation therapy for brain tumours are at increased risk of cerebrovascular complications. Radiation-induced moyamoya syndrome (MMS) is a well-recognised complication of this. We present a case of an 8-year-old boy with a history of medulloblastoma, who underwent surgical excision followed by post-operative adjuvant oncological treatment. Six years later, he developed cerebellar/intraventricular haemorrhage. He underwent an emergency external ventricular drain (EVD) insertion followed by posterior fossa suboccipital craniotomy. On dural opening, an abnormal vessel was visualised on the surface of the right cerebellar hemisphere, which was not disturbed. No obvious abnormalities were identified intra-operatively. Cerebral catheter angiography confirmed the presence of a right-sided occipital artery (OA) to posterior inferior cerebellar artery (PICA) extracranial to intracranial (EC-IC) bypass with a zone of the distal PICA territory supplied by this EC-IC bypass. A presumed flow aneurysm originated from the bypass in the distal PICA, identified as cause for the haemorrhage. We highlight a rare cause for intracranial haemorrhage in this cohort of patients. Children who have undergone radiotherapy may have exquisitely sensitive cerebral vasculature and need careful vigilance and evaluation for vasculopathic complications following spontaneous haemorrhage.

Keywords Moyamoya syndrome · Medulloblastoma · Radiotherapy · Cranial irradiation

Background

Paediatric patients receiving cranial irradiation therapy for brain tumours are at increased risk of cerebrovascular complications (e.g. stroke, cerebral haemorrhage, aneurysms, occlusive vasculopathies) [1]. Campen et al. report that incidence of such events is 100-fold higher in this population of patients in comparison to the general paediatric population [1].

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Moyamoya disease is an occlusive vasculopathy, characterised by progressive occlusion of the terminal portion of the internal carotid artery (ICA) and its main branches, resulting in cerebrovascular events [2]. The reduced blood flow leads to formation of compensatory spontaneous collateral neovascularisation, which classically arise from the ICA and more rarely from branches of the external carotid artery (ECA) [3]. Aneurysm formation as a result of collateral vasculature hypertrophy is well described in adult moyamoya disease [4].

When moyamoya vasculopathy occurs in the context of an underlying medical condition, it is known as moyamoya syndrome (MMS). Radiation-induced MMS is a well-recognised complication of cranial irradiation therapy [5].

We present the rare case of a child with history of medulloblastoma, who underwent surgical excision followed by post-operative adjuvant oncological treatment, and subsequently developed cerebellar/intraventricular haemorrhage from a spontaneous extracranial to intracranial (EC-IC) bypass-related flow aneurysm. To our knowledge, this is the only such case study reported in the literature.

Case

An 8-year-old boy presented in September 2016 with ataxia and was diagnosed with a posterior fossa tumour (Fig. 1A). He underwent craniotomy and macroscopic complete resection. Histology confirmed medulloblastoma—desmoplastic nodular subtype (stage M0, SHH activated, wildtype TP53 positive, MYCN/MYC non-amplified). Genetic screening was performed (normal *SUFU/PTCH1/TP53* genetics), and Gorlin syndrome was excluded. The cerebrospinal fluid (CSF) was negative for metastatic deposits. The oncological stage was M0. He received adjuvant chemotherapy (on good risk HIT-SKK protocol—3 cycles of vincristine, cyclophosphamide and intrathecal methotrexate, carboplatin, etoposide).

In August 2017, he had a local relapse in the tumour bed and underwent revision surgery for excision of recurrence. In September 2017, he underwent craniospinal radiotherapy with posterior fossa boost (whole CNS 23.4 Gy × 13 fractions with boost 30.6 Gy × 17 fractions for a total dose of 54 Gy). In December 2017, he underwent 4 cycles of topotecan and cyclophosphamide. After completion of adjuvant therapy in February 2018, serial radiological surveillance confirmed disease remission.

In March 2022, he presented with sudden onset headache associated with loss of consciousness. On arrival, he was in a coma with bilateral unequal pupils, hypertensive, bradycardic and required brief cardiopulmonary resuscitation. Following 2 boluses of hypertonic saline, he was intubated for neuroprotection. A CT of the brain confirmed a posterior fossa haemorrhage with its epicentre in the right cerebellar hemisphere, with rupture and extension into the 4th ventricle, 3rd and lateral ventricles (Fig. 1B, C). There was vermian haemorrhage with anterior effacement of brainstem and obliteration of the pre-medullary and pre-pontine CSF spaces.

He underwent an emergency right frontal external ventricular drain insertion followed by posterior fossa suboccipital craniotomy. On dural opening, an abnormal vessel was visualised on the surface of the right cerebellar hemisphere. This was not disturbed, and a telo-velar approach was utilised to evacuate the 4th ventricular, vermian and part of the cerebellar haemorrhage. Intra-operatively no obvious abnormalities were identified to suggest tumour recurrence or abnormal vascular malformation (Fig. 1D).

Post-operative MRI confirmed no recurrent tumour or vascular malformation. A CT angiogram raised the suspicion of a small hypertrophic vessel with possible aneurysmal dilatation and some fine vascular structures at the periphery of the haematoma, corresponding to the location where the abnormal vessel was visualised (Fig. 2A–D).

Cerebral catheter angiography confirmed the presence of a right-sided occipital artery (OA) to posterior inferior cerebellar artery (PICA) EC-IC bypass with a zone

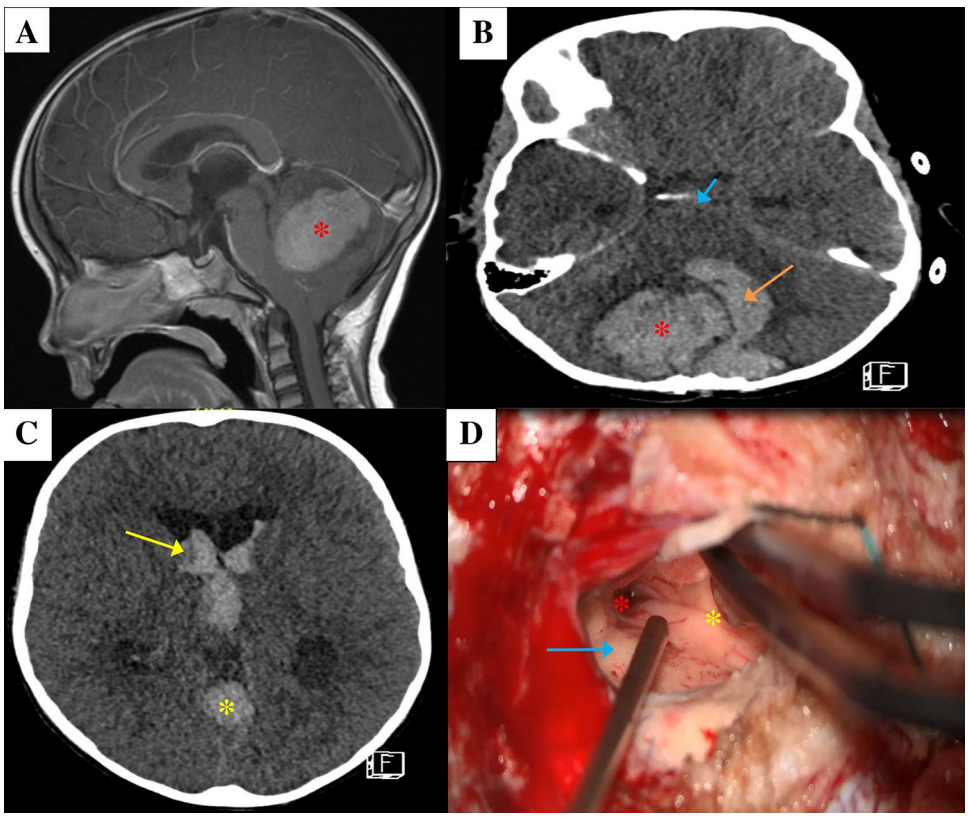

Fig. 1 **A** T1 weighted with gadolinium enhancement (T1+C) sagittal MRI (09/2016) demonstrating the initial presentation with posterior fossa medulloblastoma arising from the superior medullary velum (red star). **B** Presenting axial CT head (03/2022) with new right cerebellar haemorrhage (epicentre/red star), 4th ventricular haemorrhage (orange arrow) and anterior displacement of brainstem and pre-pontine/pre-medullary cisterns (blue arrow). **C** Upper slices of the presenting axial CT head (03/2022) with supra-cerebellar haemorrhage (yellow star) and lateral and 3rd ventricular extension of blood (yellow arrow). **D** Intraoperative visualisation of the 4th ventricular floor with cerebral aqueduct (red star), median sulcus (yellow star) and left superior cerebellar peduncle (blue arrow) following evacuation of intraventricular haemorrhage. No evidence of tumour recurrence or abnormal vascular pathology

of distal PICA territory supplied by this EC-IC bypass (Fig. 3A, B). A presumed flow aneurysm originated from the bypass in the distal PICA, identified as cause for the haemorrhage. It was treated with ONYX via the OA to occlude the aneurysm without distal embolisation into PICA territory (Fig. 3C, D).

The child's post-operative course was complicated by a right mesial occipital territory infarction secondary to cerebrovascular vasospasm (Fig. 4A, B). He was treated with a combination of nimodipine, volume replacement and hypertension and the vasospasm rapidly reversed (Fig. 4C). He was in the intensive care unit for a total of 13 days and at discharge was mobilising with assistance. He had a period of cerebellar mutism, which rapidly improved, and he was communicating with words and good comprehension at 3-month follow-up. MRI/MRA demonstrated completed resolution of global cerebral vasospasm (Fig. 4D).

Discussion

MMS is a well-recognised long-term complication of radiotherapy predominantly in children receiving treatment for suprasellar tumours, with the posterior cerebral circulation rarely involved [6, 7]. The interval between radiation exposure and development of MMS varies on a case-by-case basis, with a reported median interval of 5.4 years [5].

Radiation-induced MMS is usually managed with surgical revascularisation utilising ECA sources for flow augmentation. This includes direct, indirect and combined revascularisation techniques [5].

In our case, we hypothesise that the combination of previous surgeries and irradiation led to a local area of vascular insufficiency. This in combination with the presence of a previous craniotomy and apposition or proximity of extracranial tissues supplied by the OA led to development of a spontaneous OA to distal PICA EC-IC bypass with resultant flow aneurysm. The severity of vasospasm was not in keeping with the pattern or degree of haemorrhage. Our patient had isolated ventricular and cerebellar parenchymal haemorrhage with no subarachnoid haemorrhage. Classically vasospasm is reported to be highest in high Fisher grades of subarachnoid haemorrhage. Nonetheless, there was severe global vasospasm affecting the anterior and posterior circulations. This may be due to exquisitely sensitive cerebral vasculature related to previous radiotherapy.

We highlight a rare cause for intracranial haemorrhage in this cohort of patients. The development of a spontaneous EC-IC bypass and subsequent flow aneurysm in this patient raises interesting questions about the feasibility

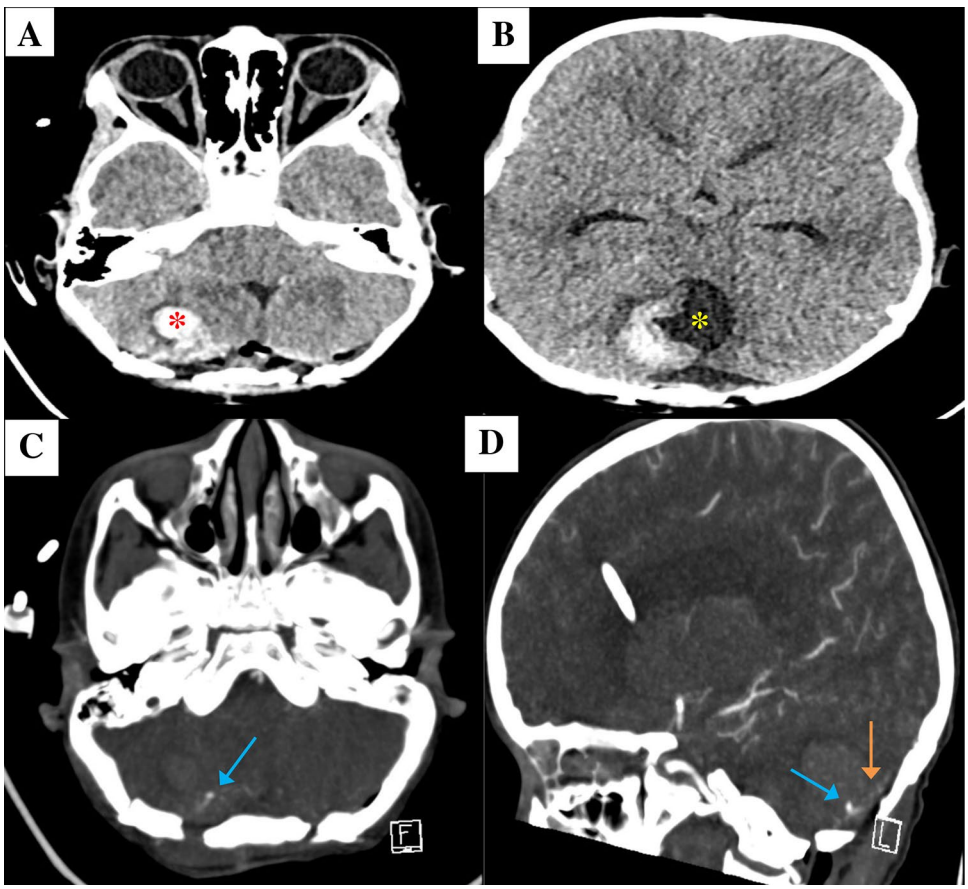

Fig. 2 **A** Axial post-operative CT Head with 4th ventricular haemorrhage evacuated and residual cerebellar hemispheric haemorrhage (epicentre = red star). **B** Upper slices of axial post-operative CT head demonstrating decompression of 4th ventricle (yellow star), near complete evacuation of vermian and 4th ventricular haemorrhage and reduction in ventricular calibre. **C** CT angiography with suspicion of hypertrophic vessel directly over the epicentre of the cerebellar hemispheric haemorrhage on the surface (blue arrow) corresponding where the abnormal vessel intra-operatively was visualised. **D** Sagittal maximal intensity projection (MIP) of the CT angiogram demonstrating the aneurysmal like dilatation (blue arrow) with a subsequent leash of vessels distally over the cerebellar hemispheric surface (orange arrow)

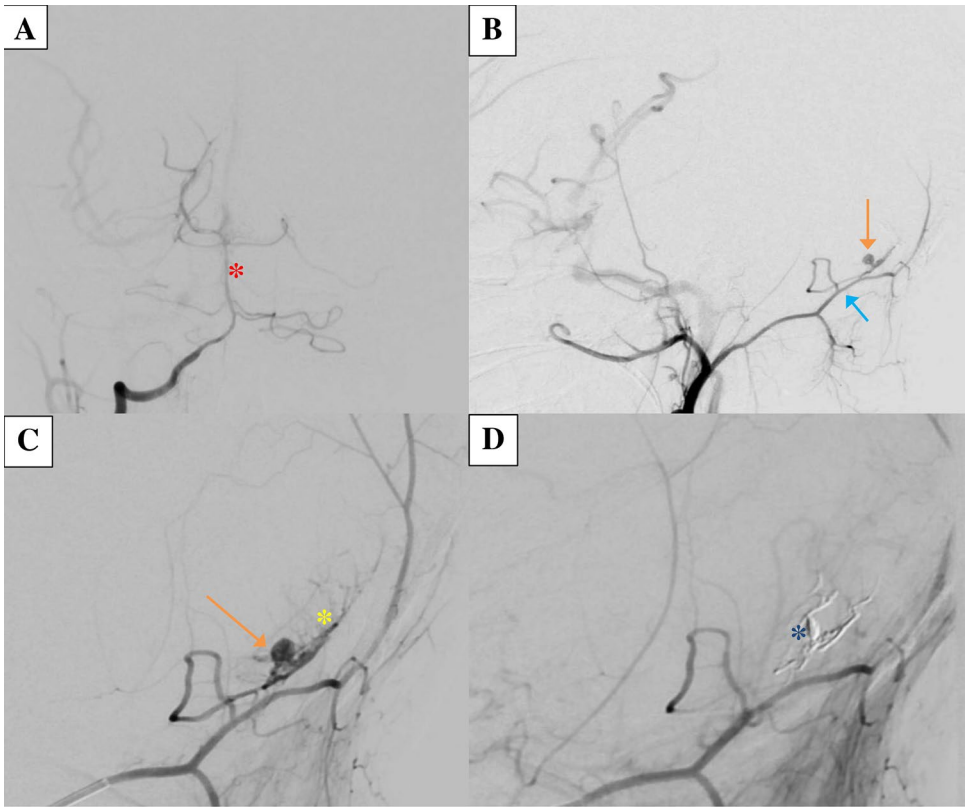

Fig. 3 **A** AP angiography with right vertebral artery injection. Severe vasospasm in vertebrobasilar trunk and posterior circulatory vasculature observed (red star). **B** Lateral angiography with right external carotid artery (ECA) injection (note there is some contamination with internal carotid artery filling). Branch from right occipital artery (blue arrow) visualised forming spontaneous EC-IC bypass to PICA territory with flow aneurysm (orange arrow). **C** Selective right occipital artery catheterisation better demonstrates flow aneurysm and collateralisation of distal PICA territory on hemispheric surface (yellow star). **D** Selective right occipital artery injection with ONYX occlusion of flow aneurysm (blue star)

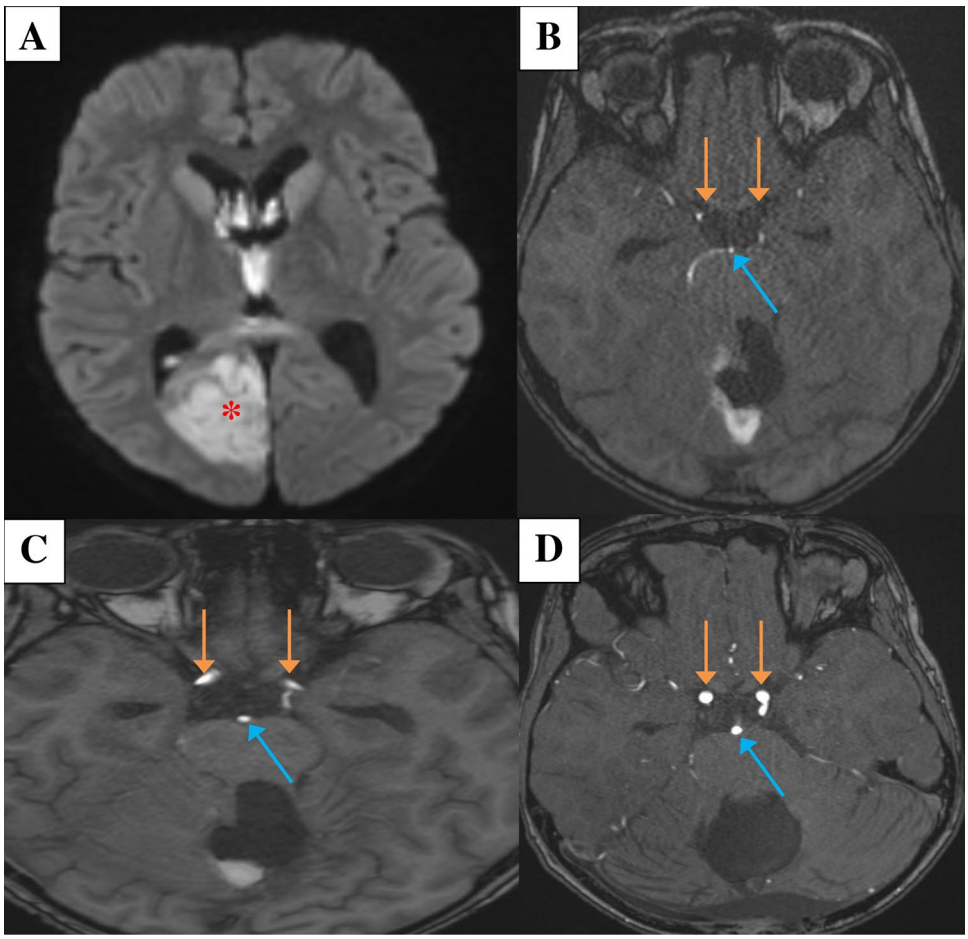

Fig. 4 **A** Diffusion weighted axial MRI confirming vasospasm-related mesial right occipital ischaemia. **B** Time of flight MRA at day 6 post ictus confirming severe vasospasm in basilar artery (blue arrow) and bilateral internal carotid arteries (orange arrows). **C** Time of flight MRA at day 16 post ictus confirming slight improvement in vasospasm in basilar artery (blue arrow) and bilateral internal carotid arteries (orange arrows) as evidenced by increasing calibre of vasculature. **D** Time of flight MRA at 3-month post ictus confirming complete resolution of vasospasm in basilar artery (blue arrow) and bilateral internal carotid arteries (orange arrows) as evidenced by normalisation of vascular calibre

for indirect revascularisation techniques in the posterior fossa utilising vascularised tissues supplied by the ECA. Children who have undergone radiotherapy may have exquisitely sensitive cerebral vasculature and need careful vigilance and evaluation for vasculopathic complications following spontaneous haemorrhage.

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Availability of data and material Data are available on request from the authors.

Declarations

Competing interests The authors declare no competing interests.

Ethics approval and consent to participate Not applicable.

Consent for publication Formal consent has been obtained from the patient's parents using the CNS consent form.

Conflict of interest The authors have no competing interest to declare that are relevant to the content of this article.

References

1. Campen CJ, Kranick SM, Kasner SE et al (2012) Cranial irradiation increases risk of stroke in pediatric brain tumor survivors. Stroke 43(11):3035–3040. https://doi.org/10.1161/STROKEAHA.112.661561
2. Scott RM, Smith ER (2009) Moyamoya disease and moyamoya syndrome. N Engl J Med 360(12):1226–1237. https://doi.org/10.1056/NEJMra0804622
3. Baltsavias G, Khan N, Valavanis A (2015) The collateral circulation in pediatric moyamoya disease. Child's Nervous System 31(3):389–398. https://doi.org/10.1007/s00381-014-2582-5
4. Kim JH, Kwon TH, Kim JH, Chong K, Yoon W (2018) Intracranial aneurysms in adult moyamoya disease. World Neurosurg 109:e175–e182. https://doi.org/10.1016/j.wneu.2017.09.127
5. Scala M, Fiaschi P, Cama A et al (2020) Radiation-induced moyamoya syndrome in children with brain tumors: case series and literature review. World Neurosurg 135:118–129. https://doi.org/10.1016/j.wneu.2019.11.155
6. Wu Y, Chang F, Liang M et al (2016) Incidence and long-term outcome of postradiotherapy moyamoya syndrome in pediatric patients with primary brain tumors: a single institute experience in Taiwan. Cancer Med 5(8):2155–2160. https://doi.org/10.1002/cam4.785
7. Ullrich NJ, Robertson R, Kinnamon DD et al (2007) Moyamoya following cranial irradiation for primary brain tumors in children. Neurology 68(12):932–938. https://doi.org/10.1212/01.wnl.0000257095.33125.48