CASE REPORT

Rare presentation of intracranial vascular blowout after tumor resection and radiation therapy

Ali Alaraj,¹ Mandana Behbahani,¹ Tibor Valyi-Nagy,² Nathan Aardsma,² Victor A Aletich¹

¹Department of Neurosurgery, University of Illinois at Chicago, Chicago, Illinois, USA

²Department of Pathology, University of Illinois at Chicago, Chicago, Illinois, USA

Correspondence to

Dr Ali Alaraj, Department of Neurosurgery, University of Illinois at Chicago, 912 S. Wood St, MC-799, Chicago, IL 60612, USA; alaraj@uic.edu

Accepted 2 April 2014

SUMMARY

A middle-aged patient presented with a rapidly growing right dural-based extra-axial posterior clinoid mass extending to the right cavernous sinus that was surgically resected. Histological examination showed solid growth of primitive neuroectodermal tumor arising from the third nerve. Following surgical resection, the patient was further managed by radiation and chemotherapy. Two years later the patient developed new intracranial hemorrhage in the area adjacent to the previous surgical cavity. A cerebral angiogram showed contrast extravasation at the junction of the posterior communicating artery (Pcom) and the right posterior cerebral artery (PCA), with an expanding pseudoaneurysm. This was managed with N-butyl cyanoacrylate embolization. Autopsy showed microscopic recurrence of tumor into the PCA/PCom region with invasion of the wall of the Pcom. This case report illustrates the concept of vascular blowout in intracranial cerebral vasculature. It appears that, in the presence of risk factors that contribute to weakening of vessel walls (surgery, radiation, tumor recurrence), a blowout can occur intracranially.

BACKGROUND

Vascular complications associated with intracranial surgical intervention, specifically in relation to tumor resection, are well known. Arterial blowout was first described in 1961. To date, it has only been described in carotid blowout syndrome (CBS) and has been attributed to factors associated with head and neck neoplasia, radical resection, and a history of irradiation.^{1–3}

The illustrated case report presents symptoms, risk factors, and pathology consistent with that of arterial blowout which presented in the posterior cerebral artery/posterior communicating artery junction (PCA/Pcom), extending the etiology of vascular blowout to include cerebral vessels. The concept of arterial blowout has been limited to CBS. We hypothesize that blowouts can occur in the cerebral vessels in association with radical surgical resection, irradiation of tissue, and tumor invasion of the vessels.

CASE PRESENTATION

A middle-aged patient with a history of congenital deafness presented initially with painless progressive right third nerve palsy. Initial investigation with MRI showed a dural-based extra-axial mass adjacent to the right posterior clinoid process of the dorsum sella which extended to the right

cavernous sinus along the course of the right oculomotor nerve. The size of the mass had increased notably during a period of 2 months on two sequential MRIs (figure 1A, B). The tumor was surgically resected. Intraoperatively, the tumor appeared to originate from the oculomotor cranial nerve; gross resection of the tumor was achieved without injuring any vital structures that were adherent to the mass, and both the PCA and Pcom were visualized with no gross abnormalities. Postoperative MRI showed no residual tumor (figure 1C). Histopathologic examination of the lesion revealed a primitive neuroepithelial tumor (PNET) involving the third cranial nerve and featuring high cellularity, round to oval to spindle-shaped nuclei with scant cytoplasm and brisk mitotic activity. These findings were most consistent with a CNS-type PNET involving the oculomotor nerve.

TREATMENT

Because of the risk of neuroaxial recurrence, the patient was treated with radiation therapy, receiving 36 Gy each to the spine and brain and 50.4 Gy to the tumor bed. The patient tolerated the radiation without significant toxicity and was later started on a chemotherapy regimen; a total of six cycles of Ewing's chemotherapy regimen were administered. At the 2-year follow-up MRI of the brain and spine, there was no gross sign of tumor recurrence; enhancement locally was thought to be related to post-radiation changes (figure 1D). After surgery for subglottic stenosis (related to prolonged intubation from chemotherapy-related sepsis 18 months after surgery), the patient did not awake from the procedure. A CT brain scan showed massive subarachnoid hemorrhage with intraventricular extension as well as extension into the thalamus (figure 1E). This was treated with placement of bilateral frontal external ventricular drains. The patient later underwent angiography, which showed active contrast extravasation from the right Pcom/PCA junction with an expanding hematoma (figure 2A, B). Using a Prowler-14 microcatheter (Cordis-Codman; Raynham, Massachusetts, USA), the PCA was super-selected and N-butyl cyanoacrylate (n-BCA; Cordis-Codman) 2:1 n-BCA/ethadiol oil was used for embolization. This resulted in complete occlusion of the right P1 segment and a portion of the proximal P2 segment distal to the junction with the Pcom (figure 2C, D). Because of persistent extravasation from the Pcom, the right Pcom was also also

CrossMark

To cite: Alaraj A, Behbahani M, Valyi-Nagy T, et al. *BMJ Case Rep*. Published online: [please include Day Month Year] doi:10.1136/bcr-2014-011192

Figure 1 (A) Baseline axial T1-weighted image with contrast of the brain showing a lesion along the trajectory of the right third nerve. The tumor enhances inhomogeneously (arrow). (B) Coronal T2-weighted image demonstrating the lesion adjacent to the posterior clinoid process, anterolateral to the brain stem (arrow). (C) Postoperative axial T1-weighted image with contrast showing no residual tumor within the surgical bed. (D) Two-year follow-up MRI axial images with contrast of the brain showing enhancement within the surgical cavity (arrow) in the vicinity of the posterior communicating artery. This was initially interpreted as enhancement related to the radiation therapy rather than tumor recurrence. (E) Axial CT scan of the brain performed at the time of presentation of the intracranial blowout showing massive intraventricular hemorrhage in addition to the intraparenchymal component. (F) Right internal carotid artery (lateral projection) showing a patent right posterior communicating artery (arrow); no acute extravasation was seen on this injection.

embolized with 2:1 n-BCA using a Prowler-14 microcatheter (figure 2E, F). No contrast extravasation was seen at the end of the procedure. Despite controlling the active bleeding, the patient progressed to brain death secondary to the massive intracranial hemorrhage and died a few days later. The last MRI of the brain performed 2 months earlier had failed to show gross tumor recurrence; local enhancement was thought to be related to radiation changes. Autopsy examination showed no evidence of tumor involvement of the brain and no gross evidence of tumor recurrence in the prior surgical bed. However, the autopsy detected microscopic recurrence of tumor into the PCA/PCom region with invasion of the wall of the Pcom (figure 3A, B). There was also evidence of focal dystrophic calcification of the vessel wall possibly related to previous radiation.

DISCUSSION

Vascular blowout syndrome (the term referring to rupture of an artery), known as blowout syndrome, has been described in the carotid vasculature in association with neoplasia involving the head and neck. The pathophysiology underlying carotid artery blowout involves several aspects including (1) tumor invasion of the carotid vessel causing weakening in the vessel wall and/or pseudoaneurysm formation; (2) extensive surgical resection of the tissue overlying the carotid vessel leading to loss of supportive connective tissue; and (3) a history of irradiation, with free radicals causing inflammation and increasing the risk of thrombosis of the vasa vasorum leading to breakdown of the vessel walls and fibrosis of the vessels targeted. Multiple other risk

factors such as smoking, age, a history of vasculitis, and hypertension have also been reported to play a role in CBS.^{1 4 5} The occurrence of vascular blowout in the intracranial vessels has not previously been reported, and it seems that the same pathophysiological mechanism for CBS applies to that of the intracranial vessels.

In the illustrated case the patient had a rare form of PNET originating from the third cranial nerve and received maximal therapy including surgical resection, radiation, and follow-up chemotherapy with no sign of gross tumor recurrence over 2 years, as followed by radiographic studies. At the time of the clinical presentation, the patient developed acute subarachnoid, intraventricular and intracranial hemorrhage for which he was treated with endovascular embolization of PCA/Pcom vascular blowout. Retrospective pathological analysis of the injured vascular tissue showed microscopic traces of primary tumor invasion of the vascular wall without any gross presence of tumor. This corresponds to what appears to be a grade 3 CBS, with evidence of extravasation from the ruptured artery.⁶ The microscopic analysis of our patient matches that of cases of CBS published in the literature, including weakening of the arterial wall, microscopic tumor invasion, and destruction of the artery itself.^{7 8} The combination of a lack of the adventitial layer and direct destruction of the wall with tumor invasion are all possible pathophysiological factors that can contribute to this pathology. Hemorrhage as a clinical presentation has been associated with tumor emboli (like myxoma) to the brain,⁹ yet it appears that the pathophysiology of hemorrhage in the tumor emboli

Figure 2 (A,B) Left vertebral artery angiograms (A, anteroposterior projection; B, lateral projection) showing active extravasation of contrast from the right posterior communicating (Pcom) artery with pseudoaneurysm formation adjacent to the site of bleeding (arrow). (C, D) Left vertebral artery angiograms (C, anteroposterior projection; D, lateral projection) after endovascular embolization of the proximal right posterior cerebral artery (PCA) with n-butyl cyanoacrylate. After embolization there is no filling of the right PCA (arrow). (E) Right internal carotid artery (ICA) angiogram (lateral projection) after embolization of the right PCA showing a patent right Pcom artery (arrow) with acute extravasation into the pseudoaneurysm. (F) Right ICA angiogram (lateral projection) after embolization of the right Pcom artery; no filling of the Pcom artery or contrast extravasation is seen.

group is related to the formation of pseudoaneurysms,^{9–11} probably related to tumor invasion. The angiographic features seen in our patient are more consistent with the pathology of vascular blowout, which is a local process related to surgery and radiation therapy.

As has been described with cervical CBS,^{12–14} it appears that endovascular treatment should be considered as a first option for the treatment of such pathologies. While bleeding from a cervical CBS might be massive, patients can undergo timely resuscitation with fluid prior to endovascular treatment; intracranial blowout is less forgiving because of the impending brain herniation with a relatively smaller amount of bleeding, as was the case in our patient. For patients with sentinel type of bleeding, endovascular vessel sacrifice with coils or liquid embolic agents (eg, glue or Onyx) would be the primary treatment.

The presentation of this case is consistent with the concept of vascular blowout syndrome and should be considered in the differential diagnosis of patients presenting with intracranial bleeding following tumor resection and radiation therapy.

Learning points

- ▶ A rare origin of primitive neuroepithelial tumor coming from the third nerve is described.
- ▶ This is a rare presentation of intracranial blowout syndrome after surgery and radiation.
- ▶ Treatment options for intracranial blowout should be learnt.

Figure 3 (A) Primitive neuroepithelial tumor (PNET) adjacent to (black arrow) and invading the wall (white arrow) of the artery at the base of the brain in autopsy specimen (H&E, 50× magnification). (B) PNET invading the wall (white arrow) and present in the lumen (black arrow) of an artery at the base of the brain in autopsy specimen. The vessel wall also exhibits intimal calcifications (H&E, 100× magnification).

Contributors All authors participated in the conception, design, drafting of the article and final approval of the version to be published. AA: patient data collection, interpretation and analysis, primary manuscript preparation and editing, critical review of manuscript approval of final version. MB: patient data collection, design of the article, manuscript editing. TV-N: patient data collection, data analysis, critical review and editing. NA: patient data collection, manuscript editing and preparation. VAA: data analysis, critical review and intellectual contribution, approval of final version.

Competing interests None.

Ethics approval An institutional review board approved the current study.

Provenance and peer review Not commissioned; externally peer reviewed.

Data sharing statement The department maintains the original record of the data presented in this manuscript; data that are not relevant to this study are available from the primary treating team.

REFERENCES

- McDonald MW, Moore MG, Johnstone PA. Risk of carotid blowout after reirradiation of the head and neck: a systematic review. *Int J Radiat Oncol Biol Phys* 2012;82:1083–9.
- Bogdasarian RS, Kwyer TA, Dauser RC, et al. Internal carotid artery blowout as a complication of sphenoid sinus and skull-base surgery. *Otolaryngol Head Neck Surg* 1983;91:308–12.
- Thawley SE. Complications of combined radiation therapy and surgery for carcinoma of the larynx and inferior hypopharynx. *Laryngoscope* 1981;91:677–700.
- Carmody BJ, Arora S, Avena R, et al. Accelerated carotid artery disease after high-dose head and neck radiotherapy: is there a role for routine carotid duplex surveillance? *J Vasc Surg* 1999;30:1045–51.
- Abayomi OK. Neck irradiation, carotid injury and its consequences. *Oral Oncol* 2004;40:872–8.
- Chang FC, Lirng JF, Luo CB, et al. Patients with head and neck cancers and associated postirradiated carotid blowout syndrome: endovascular therapeutic methods and outcomes. *J Vasc Surg* 2008;47:936–45.
- Swain RE, Biller HF, Ogura JH, et al. An experimental analysis of causative factors and protective methods in carotid artery rupture. *Arch Otolaryngol* 1974;99:235–41.
- Kane KK. Carotid artery rupture in advanced head and neck cancer patients. *Oncol Nurs Forum* 1983;10:14–18.
- Herbst M, Wattjes MP, Urbach H, et al. Cerebral embolism from left atrial myxoma leading to cerebral and retinal aneurysms: a case report. *AJNR Am J Neuroradiol* 2005;26:666–9.
- Chen HJ, Liou CW, Chen L. Metastatic atrial myxoma presenting as intracranial aneurysms with hemorrhage: case report. *Surg Neurol* 1993;40:61–4.
- Jean WC, Walski-Easton SM, Nussbaum ES. Multiple intracranial aneurysms as delayed complications of an atrial myxoma: case report. *Neurosurgery* 2001;49:200–2; discussion 202–3.
- Alaraj A, Wallace A, Amin-Hanjani S, et al. Endovascular implantation of covered stents in the extracranial carotid and vertebral arteries: case series and review of the literature. *Surg Neurol Int* 2011;2:67.
- Warren FM, Cohen JL, Nesbit GM, et al. Management of carotid 'blowout' with endovascular stent grafts. *Laryngoscope* 2002;112:428–33.
- Zussman B, Gonzalez LF, Dumont A, et al. Endovascular management of carotid blowout. *World Neurosurg* 2012;78:109–14.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow