Emergency Extracranial-Intracranial Bypass during Surgery for Anterior Clinoid Process Hemangiopericytoma with ICA Invasion—Case Report

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Abstract

Hemangiopericytoma (HPC) is a rare tumor, accounting for less than 1% of intracranial tumors. Though it’s notoriously difficult to distinguish HPC from meningioma in pre-op images, HPC presents bone and vascular invasion, high local recurrence and distal metastasis clinically, as a surgically challenging malignancy.

High-flow extracranial-intracranial (EC-IC) bypass is a skill demanding surgery and is usually performed as a well-prepared elective surgery. However, emergency EC-IC bypass is indicated for unplanned major artery loss during surgery and sometimes as the final rescue for the patient.

We present a case of a 46 year-old man, who was diagnosed of a left anterior clinoid process (ACP) tumor with presentation of gradually visual loss. As the dura tail sign and MRI signal, we performed left pterional approach tumor excision for the impression of ACP meningioma. However, due to the vascular invasion of tumor, left ICA perforation was noted intraoperatively and finally the bleeding was controlled with ICA clipping. Emergency EC-IC bypass from common carotid artery to middle cerebral artery was performed. The patient recovered to oriented consciousness, independent ambulation but mild aphasia in our outpatient department follow-up.

Keywords: Hemangiopericytoma; Extracranial-intracranial bypass; ICA invasion

Introduction

Hemangiopericytoma (HPC) is a rare tumor accounting less than 1% of all intracranial tumors [1]. It originates from capillary pericyte (Zimmerman’s pericyte) [2,3]. Due to the vascular origin, hypervascularity is an important character of HPC. Besides, it is classified to be WHO grade II or even grade III (anaplastic HPC) and presents aggressive characters, such as local invasion, local recurrence and even distal metastasis [3]. However, it is very difficult to differentiate HPC and meningioma in pre-operative images. Therefore, owing to the high surgical risk and easily misdiagnosis as a meningioma, HPC is notoriously described as “a wolf in sheep’s clothing” [4].

Though high-flow EC-IC bypass was first reported in 1960 by Woringer, it is still a highly surgically demanding procedure even in modern Neurosurgery. It is mostly performed in a well-prepared setting unless in some emergency situations. Therefore, reports about urgent high-flow EC-IC bypass are few [5,6]. In this report, we performed emergency high-flow EC-IC bypass from left common carotid artery to middle cerebral artery in order to rescue the territory of the perforated internal carotid artery. Even added on post-op cerebellar ICH, after rehabilitation, he finally recovered to lead a minimally-dependent life.

Case

We present a 46 year-old man without family history of neoplasm. The symptom developed as left blurred vision for 3 years. The symptom progressed gradually. And in recent 1 year, his right vision also deteriorated. Initially, he went to a local hospital. The visual field examination disclosed nearly total vision loss in his left eye and temporal hemianopsia in his right eye. The brain CT disclosed a 2–3 cm lesion at his left suprasellar area. He was then referred to our hospital.

The consequent brain MRI with contrast presented a 3.6 x 2.5 x 2.8 cm tumor in sellar and suprasellar area extending forward to frontal base with suspected dura tail sign. His left sphenoid sinus, anterior clinoid process (ACP), left ICA, bilateral optic nerve and optic chiasm were also involved in the tumor. However, the pre-op MRA didn’t disclose stenosis or filling defect, which could impress us of the ICA invasion. The tumor presented heterogeneous hyper-iso intensity in T2, isointensity in T1 and strong enhancement with gadolinium contrast. Initially, ACP meningioma was impress based on the radiological characters. However, out of our pre-op evaluation, the lobulated tumor shape, left optic canal extension and evident hypervascularity could give some hints of the more aggressiveness and possible “an unusual diagnosis” rather than meningioma (Figure 1).

We performed left pterional approach with anterior clinoidecotomy to excise this tumor. A pinkish hypervascular tumor was exposed (Figure 2). After opening the capsule we started to remove the lesion piece by piece with tumor forceps. The internal decompression was started away from the ICA. The vital structures, such as optic nerve and ICA were gradually identified. However, suddenly fresh blood gushed out from the tumor. We struggled to expose the bleeding as an ICA defect suspected resulted from tumor local invasion. Finally, to achieve hemostasis, we trapped the ICA with aneurysm clips to directly cover the ICA defect and ligate the proximal segment (Figure 3). Cardiovascular surgeon was immediately consulted. The residual tumor was soon removed and the cardiovascular surgeon harvested left great saphenous vein as a vessel graft. The graft was Anastomosed to left common carotid artery and placed to the craniotomy field through subcutaneous tunnel. We then performed anastomosis of

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Received December 29, 2014; Accepted February 23, 2015; Published February 25, 2015


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the vessel graft and left middle cerebral artery M1 segment with 9-o prolene. The total ischemic time from ICA clipping to perfusion from bypass is 140 mins. During the anastomosis, we had infused 5000U heparin and mildly diffuse oozing was noted in the surgical field. However, the post-op brain CT disclosed a 35 ml cerebellar intraparenchymal hematoma with obstructive hydrocephalus (Figure 4). The hematoma was emergently evacuated. After the condition of the patient was stabilized, the consequent angiography disclosed patent bypass and well-perfused ICA territory (Figure 5). After several-month rehabilitation, he could lead daily life with moderate assistance. We implanted lumbar peritoneal shunt for the hydrocephalus and his gait and speech improved furthermore. 1 year after surgery, he could walk without assistance, speak slowly and reach things by himself.

In pathology, the tumor cells present monomorphic oval- to spindle-shaped cells with mild cellular atypia. There are abundant thin-walled, gaping vessels with “staghorn configurations”, which is the typical pattern of HPC. The mitotic count is 5-8/10 HPF. With CD34, the immunostain for vessels, the staghorn-patterned sinusoid is more prominent. The special stain of bcl-2 is also positive (Figure 6). Therefore, the final diagnosis is WHO grade II Hemangiopericytoma.

Discussion

Hemangiopericytoma (HPC) is an important differential diagnosis of meningioma. It is a rare tumor accounting for only 0.4% of all intracranial tumors and the incidence of meningiomas is about 50 times to HPC [2]. HPC was first described in 1954 by Begg and Garret [2,A]. The origin of HPC is capillary pericyte, which is an important structure of blood brain barrier. HPC is classified to WHO grade II and presents relatively high local invasion and recurrence rate [3], (5-year disease progression free survival rate: 51%, 10-year survival rate: 72%). Even extracranial metastasis rate had been reported as 11.6% [7]. The hypervascularity and invasion to adjacent structure increase the surgical risk and difficulty. The grossly total resection rate of all intracranial HPC is therefore only 56.89% [8]. Though HPC is well known as an important and dangerous differential diagnosis of meningioma, it's still very difficult to recognize it in preoperative images. Similar to meningiomas, MRI of HPCs discloses hypo- or iso- intense in T1, hyperintense in T2 and well enhancement with gadolinium contrast. Besides, though less common, HPC presents dura tail sign like this case [9]. Reviewing the pre-operative MRI in detail, only lobulation and hypervascularity, which was more common in HPC than meningioma, could give us some hints.

Besides the aggressive characters, in this case, the tumor located at anterior clinoid process, which also increases the surgical difficulty.
ACP tumors (meningioma in majority) have been surgical challenging lesions for decades since Cushing reported an ACP meningioma in 1938. Even with the decades of development of Neurosurgery, the grossly total resection rate is still usually less than 50% and the surgical mortality rate is about 5% [10]. Due to the rarity, no reliable complication rate of ACP HPC is reported, but it is logical to assume it more surgically risky than ACP meningioma.

Even highly challenging, surgery is still the treatment of choice to preserve vision and prevent disease progression for ACP tumors. The post-op vision improvement rate was reported about 66% in ACP meningiomas [10]. In HPC, surgical resection increases overall survival (OS), especially with grossly total resection. The mean overall survival times are 235 months with GTR and 175 months in STR, respectively (P=0.047) [7]. Though intraoperative great vessel loss occurred in our case, we still achieved grossly total resection in the tough condition. Adjuvant radiotherapy reduces the recurrence rate [7]. However, in our case, it was not performed due to the unstable post-op condition and the potential injury to adjacent vascular anastomosis.

To manage an ACP tumor, anterior clinoidectomy [11], is a favorable maneuver to increase the intradural exposure of tumor and adjacent vital structures, including optic nerve, internal carotid artery and cavernous sinus [12]. In this case, we also performed this procedure. The intraoperative picture disclosed well-exposed ICA. Nevertheless, totally encased and even invaded by the tumor, ICA loss still occurred.

Extracranial-intracranial bypass had been developed since 1960 and could be classified into “low-flow” and “high-flow” two types. The latter is commonly performed as anastomosis from carotid artery to ICA or MCA. Though it’s still in some debates, the current indications of EC-IC bypass are concluded to be 1. Planned vascular sacrifice for tumor, aneurysm or nontraumatic vessel dissection. 2. Emergency revascularization for unplanned vessel loss or stroke progression and 3. Augmentation of cerebral blood flow for chronic hemodynamic ischemia, which is not amenable for conventional surgery [5]. In this case, emergency EC-IC bypass was performed for unplanned intraoperative ICA loss. Though this situation is concluded to be an indication of bypass, few cases have been presented with the emergency procedure due to the surgical difficulty. Leonardo Rangel-Castilla et al. [6] reported 8 cases from 2007 to 2014 underwent urgent bypass for intraoperative skull base ICA injury [6]. Notably, only 2 of the 8 cases received emergency intraoperative EC-IC bypass after ICA injury as this patient. The others underwent urgent bypass surgery in 48hours after endoscopic surgery and angiography disclosed pseudoaneurysm or thrombosis, which were not feasible for endovascular intervention [6]. All of them achieved excellent outcome with none or minimal neurological deficit.

Unfortunately, this case was also complicated with massive post-op cerebellar ICH. The remote cerebellar hemorrhage was renowned as “Zebra hemorrhage” and was postulated to be resulted from ruptured cerebellar cortical veins due to CSF over-drainage and caudal migration of cerebellum [13]. However, in our case, no typical “Zebra”-pattern SAH was noted. We assumed that coagulopathy due to intraoperative heparin (5000U) infusion also contributed to the ICH.

We chose great saphenous vein as the graft for vascular bypass surgery due to its accessibility for the patient aseptically draped around head. Though artificial graft could save more ischemic time, GSV grafts have higher long-term patent rate and cost acceptable time to harvest. In addition, the operator had more experience with GSV as the graft in EC-IC bypass and therefore was more confident in this emergency situation. Heparin use was proved to increase the GSV graft patent rate [14]. However, to our knowledge, no definite heparin dose was proved to be both safe and efficient in EC-IC bypass.

**Figure 5:** Angiography disclosed patent bypass and well-perfused ICA territory.

**Figure 6:** Immunostaining of the tumor cells present monomorphic oval- to spindle-shaped cells with mild cellular atypia, which are abundant thin-walled, gaping vessels with “staghorn configurations”, which is the typical pattern of Hemangiopericytoma.
Conclusion

We illustrate a complicated case of skull base invasive hemangiopericytoma with anterior clinoid process and vital structures, including optic nerve and left ICA involvement. Emergency EC-IC bypass was performed for intraoperative left ICA loss. Good recovery was achieved after even intraoperative temporary left ICA territory ischemia and post-op cerebellar ICH evacuation. This case reminds us of hemangiopericytoma as a dangerous differential diagnosis of meningioma and the importance of emergency EC-IC bypass as possible the final rescue of intraoperative great vessel loss.

References


