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Case Report

Successful preoperative embolization of a cystic-solid variant of cerebellopontine angle hemangioblastoma ☆

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ABSTRACT

Tumors of the cerebellopontine angle (CPA) represent an heterogeneous group which can arise extradural, intradural-extraaxial or intraaxial compartment.

Hemangioblastomas of the cerebellopontine angle (CPA) are extremely rare.

Computed tomography (CT) and magnetic resonance imaging (MRI) are often the gold-standard radiological imaging modalities used in characterizing the lesion's features, and its relationship with the surrounding structures.

They are vascular lesions and may cause profuse bleeding intraoperatively, that is why angiography remains a crucial diagnostic and therapeutic tool, by reducing both the presurgical differential diagnosis, as well as the intraoperative bleeding by providing capability of embolization of this vascular tumor.

We present the case of a 65 year old patient with a cystic-solid variety of HMB at the right CPA, which was successfully treated by a combination of an endovascular preoperative embolization and surgery without major complications or neurological deficits.

Introduction

Hemangioblastomas are benign neoplasms of blood vessel lineage, accounting for 1.5%-2.5% of all intracranial tumors and 7-12% of posterior fossa tumors in adults [1]. Those originating from the cerebellopontine angle (CPA) are extremely uncommon [2,3]. HMBs can be identified on radiological images as solid, solid-cystic, or predominantly cystic tumor with a small mural nodule. The cystic-solid form of this lesion at the above location is even rarer, composed of a large cystic portion as well as a large solid portion [4]. Computed tomogra-

Abbreviations: HGB, hemangioblastoma; CPA, cerebellopontine angle; CT, Computed Tomography; MRI, Magnetic Resonance Imaging; ICA, internal carotid artery; ECA, external carotid artery; PVA, Polyvinyl alcohol; NBCA, N-butyl-2-cyanoacrylate.

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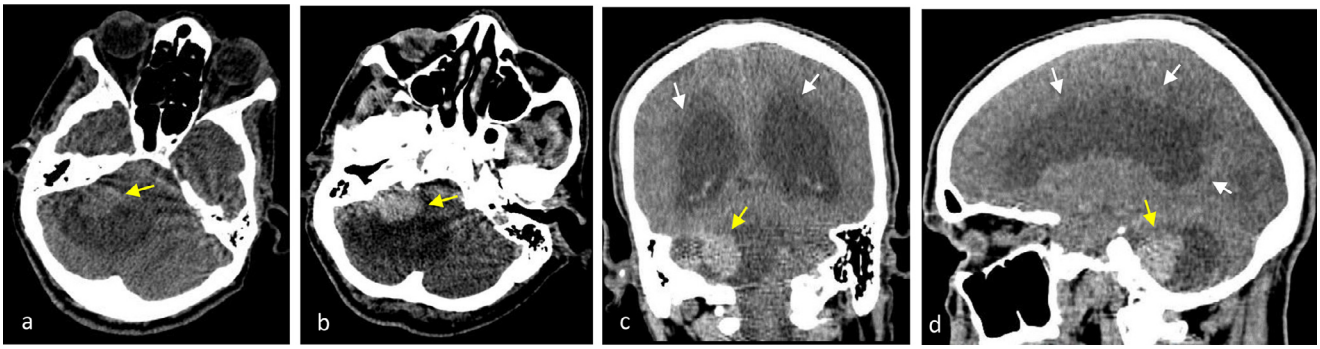

Fig. 1 – CT scan of the brain : axial section non enhanced (a), postcontrast axial (b), sagittal (c), and coronal reconstructions (d) showing a cyst-solid lesion with mural nodule (yellow arrows) of the right cerebellopontine angle measuring 65 x 55 x 50mm, strongly enhanced after contrast injection. Note the obstructive upstream hydrocephalus (withe arrows).

phy (CT) and magnetic resonance imaging (MRI) are often the gold-standard radiological imaging modalities used in identifying HMBs although they are often mistaken for other CPA neoplasms [3–5].

This highly vascular lesions carry a significant risk of hemorrhage during surgery, leading to increased rates of perioperative morbidity and mortality as well as higher rates of incomplete resections [6]. Preoperative embolization improves safety and efficacy, allowing to achieve a cleaner surgical bed and unobstructed view, resulting in shorter durations of surgery and less perioperative morbidities [7].

We present a rare case of cystic-solid HBM of the CPA, in which complete resection was achieved without morbidity, 24 hours after a preoperative endovascular embolization.

Case report

This is a case of a 65 year-old patient, with high blood pressure, presented with vertigo persistent headache associated with vomiting, balance disorder with ataxia. He denied otorrhea, history of otologic disease, or prior ear surgery. The neurologic examination revealed a cerebellar syndrome. The biological assessment was unremarkable. He firstly underwent a CT scan of the brain that revealed a large cyst with mural nodule of the right cerebellopontine angle, measuring about 65 × 55 × 50 mm, exerting mass effect upon fourth ventricle and the adjacent structures with dilatation of upstream ventricles; the evoked diagnosis was a hemangioblastoma the cerebellopontine angle with moderate obstructive hydrocephalus (Fig. 1). We completed the exploration by a cerebral MRI for a better lesion characterization; the latter confirmed our hypothesis by showing a cystic-solid lesion at the level of the right internal auditory canal. The cystic component was hypointense on T1-weighted images and hyperintense on T2-weighted images. The nodule was isointense on both T1 and T2 weighted images and enhances strongly and homogeneously after contrast media injection (Fig. 2). General management options for CPA lesions were discussed, including stereotactic irradiation, and surgical excision. Given the hypervascular appearance of the lesion, its size and mass effect on the brain, surgical resection with pre operative embolization was recommended. To determine the extent of tumor blush and the feeding arteries; a pre-embolization angiogram was performed by a 5F Vertebral catheter (Optitorque, Terumo, Japan). Super selective catheterization of the main feeding arteries, ascending pharyngeal artery branches in this case, was carried out using a suitable Microcatheter (Progreat 2.4F, Terumo, Japan) and embolization were done by 2 mL of Polyethylene Glycol (PEG) microspheres (Hydropearl 400 ± 75 μm, Terumo, Japan) and completed with a sterile absorbable gelatin sponge (Curaspon; Fig. 3). The patient underwent a total resection with minor bleeding during the procedure and the histopathological examination confirmed the diagnosis of hemangioblastoma of the CPA (Fig. 4).

Discussion

Tumors of the cerebellopontine angle (CPA) comprise 6%-10% of all intracranial neoplasms. Most of them are vestibular schwannomas (VSs) and meningiomas, which account respectively for 70%-80% and 10%-15% of all CPA lesions; however, a large variety of unusual lesions, can also be encountered in the CPA [8].

Hemangioblastomas (HGBs), which are a highly vascular and benign tumor of the central nervous system (CNS), accounts for 1.5%-2.5% of all intracranial tumors and 7%-12% of posterior fossa tumors in adults. Compared to other CPA lesions, the prevalence of HGB is extremely low [2], with a single reported case out of a cohort of 1354 CPA tumors [9].

These lesions are more common in adults aged 40-50 years, and twice more common in male than in female patients [10].

Hemangioblastomas can be solid, solid-cystic, or cystic with a tiny mural, with a vascularized nidus composed of stromal cells and many capillaries. HGB can develop spontaneously or as part of von Hippel-Lindau disease (vHL) syndrome. This condition affects around 25% of HGBs [11,12].

For long periods due to their slow growth, the femangioblastoma often remain asymptomatic. The symptomatol-

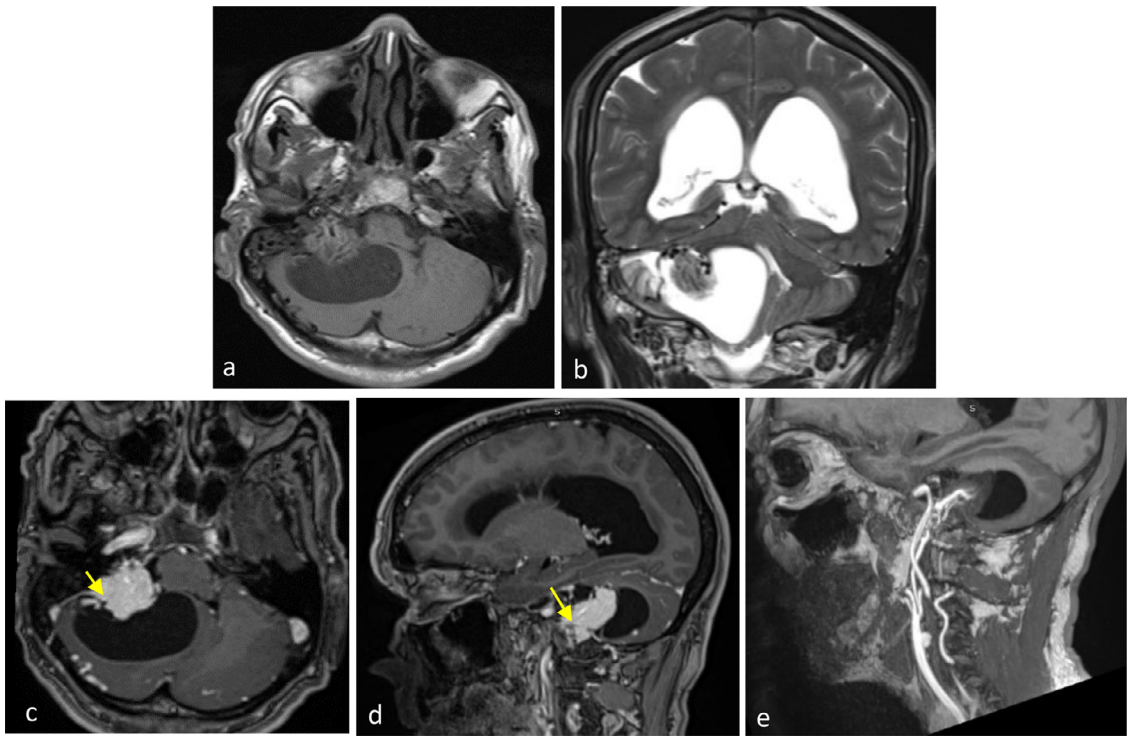

Fig. 2 – MRI of the brain. (a,b) Large cyst-solid tumor of the right cerebellopontine angle. The cystic component appears hypointense on T1 and hyperintense on T2 weighted images (WI). The solid part is isointense on both T1 and T2 WI sequences. (c,d) Axial and Sagittal T1 sequences after gadolinium administration showing the intense and homogeneous enhancement of the solid part of the lesion (Yellow arrows). (e) Sagittal T1 sequence with MIP (Maximal Intensity Projection) reconstruction demonstrating the vasculature of the lesion. (Right ascending pharyngeal artery: White arrows).

ogy depends on the size and location of the lesion. At the CPA, hemangioblastoma (HMG) may present with symptoms secondary to compression of the cranial nerves [8].

Vertigo, ataxia, hearing loss as well as tinnitus, are the usual symptomatology in patients with HMGs. This tumor is often misdiagnosed because these signs and symptoms are typical presentation of most CPA tumors [8].

In our case, the patient presented with vertigo, persistent headache associated with vomiting, balance disorder with ataxia, and neurologic examination revealed a cerebellar syndrome.

Morphologically, HGB can appear as cystic (60%-90%), solid or a combination of the two [8].

CT and MRI are very useful in the definitive diagnoses of this tumor [13]. On both CT and MRI, HMGs are frequently seen as a cystic mass with a small hypervascular mural nodule [13].

On MRI, the cystic component usually appears as hypointense on T1-weighted images and hyperintense on T2-weighted images. The nodule is isointense on both T1 and T2 and enhances intensely and homogeneously after contrast media injection [8].

The vascular nature of HGBs is manifested by high-flow vessels and dilated feeding arteries, which are demonstrated as flow voids on conventional MRI and as a prolonged tumor blush on angiography [3].

In most cases, high-flow vessels can be seen as flow voids at the periphery of the mass due to the hypervascularity of the tumor. Edema is mostly insignificant or nonexistent around lesion in all the tumor variants [8].

The advantages to perform angiography (DSA) before surgery for hemangioblastoma of CPA are the distinction of the arterial feeders of the lesion for surgical planning; the embolization of such feeders for a bloodless operation; and the establishment of a more accurate pre-surgical diagnosis by demonstrating the site of origin (intaaxial vs extraaxial) [7].

However, it was also demonstrated that the risk of complications is higher if the prominent feeders arise from the internal carotid artery (ICA) rather than the external carotid artery (ECA) [14]. In our case, the lesion was supplied by the right ascending pharyngeal artery branches of the ispsilateral external carotid artery.

Preoperative embolization allows occlusion of deep feeders and more importantly filling the tumor with embolic materials to induce its degeneration. This facilitates complete resection of the tumor with minimal risk of major blood loss [15].

Hemodynamic factors and vascular anatomy influence the choice of the optimal embolic agent. Coils can be used to cushion the blood flow of high flow fistulas before the use of other materials. For an effective and successful embolization the penetration of smaller tumor vessels is required. This penetration can be achieved with small particles like PVA and Embospheres and liquid agents like NBCA and Onyx. However, small particles can run away and reach pulmonary ves-

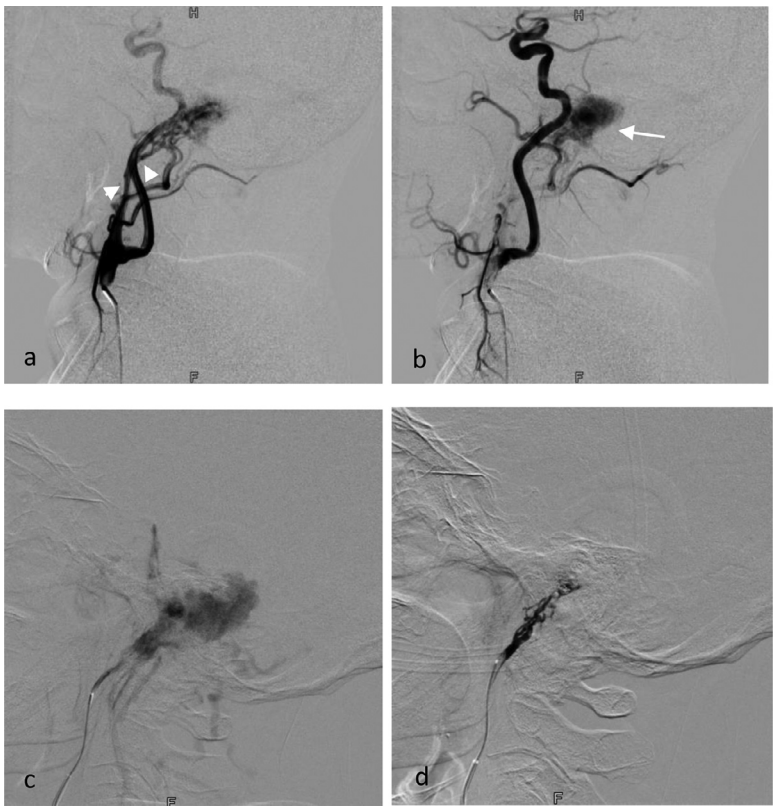

Fig. 3 – (a,b) Pre-embolization angiogram showing the tumoral blush (white arrow), arising from the right ascending pharyngeal artery (white head arrows). (c) Super-selective catheterization of the right ascending pharyngeal artery with a 2.4F Microcatheter, demonstrating the tumoral blush before embolization. (d) Post embolization angiogram demonstrating complete devascularisation of the tumor.

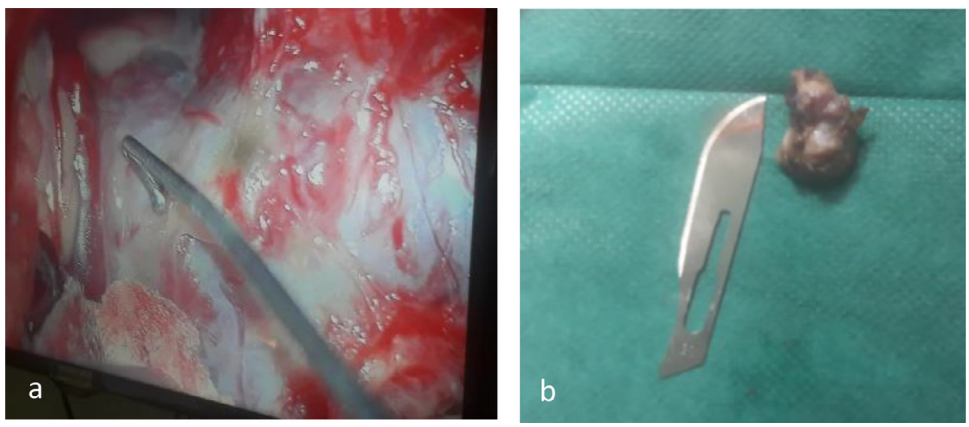

Fig. 4 – (a) Per operative view of the tumor. (b) Operative specimen demonstrating the macroscopic aspect of the solid portion of the tumor.

sels. Some authors have observed high morbidity and mortality rates with particle embolization. Although, other authors, such as Eskridge et al. outlined safe embolization using PVA in nine cases, with entire surgical resection [16].

Corneliusc et al. observed poor prognosis, acute tumor hemorrhage as well as death as cardinal complication associated with preoperative embolization for HMGs [17]. Furthermore, tumor engorgement, as well as vessel occlusion resulting in acute infarction, has been associated with preoperative embolization for HMGs [18,19].

Surgery via the middle fossa, retro-sigmoid or transtemporal approach is still the most preferred treatment modality for CPA HMGs [10,20]. In our patient a total resection of the tumor was achieved via the retro-sigmoid approach.

Massive bleeding with significant blood loss can occur during attempts at surgical resection, which resulted in ending the procedure in many cases [15,19]. In their study, Liu et al. reported that severe intraoperative bleeding impeded complete resection in eight cases of the control group, with blood loss reaching 3240mL in certain cases [15].

In surgically treated patients, Cheng et al noted facial paresis, lower cranial nerve deficits, abducens nerve paralysis, facial hypesthesia, cerebellar hemorrhage, CSF leakage, and pseudo-meningocele as postoperative complications [4].

We did not observe any such complication after embolization or surgical resection of this lesion in our patient.

Macroscopically, Hemangioblastomas can be either solid or cystic. The majority, 60%-90% of them are cystic. They usually present as a reddish or yellow mural nodule with a cystic component, and classified histologically into cellular and reticular subtypes [21].

Conclusion

The cystic-solid variant of cerebellopontine angle hemangioblastoma is a highly vascular tumor, which had feeders mainly from neuromeningeal division of ascending pharyngeal branch of external carotid artery, suggesting true extra-axial origin. Computed tomography (CT) and magnetic resonance imaging (MRI) are the gold-standard radiological imaging modalities used in characterizing of this lesion. Preoperative embolization allows the surgeon to achieve a total surgical excision without significant blood loss or morbidity.

Patient consent

We confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

The consent to publish potentially identifying information, such as details or the case, was obtained from the patient.

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