

Intracranial Hemangiopericytoma with Right-Sided Aortic Arch: A Case Report and Comprehensive Diagnosis and Treatment Analysis

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1. Abstract

Hemangiopericytoma are clinically rare, and easily confused with meningioma. Right-sided aortic arch is also an infrequent disease of congenital cardiovascular variability. The intracranial hemangiopericytoma associated with right-sided aortic arch has not been reported in the literature. We are here to report a case of right aortic arch concurrent with intracranial hemangiopericytoma, the patient was initially thought to be a simple meningioma, but was eventually diagnosed as a hemangiopericytoma with right aortic arch and multiple intracranial vascular stenosis and malformation. Whether there is a genetic correlation between the two provides a potential research direction for us. Therefore, the diagnosis and treatment of some rare diseases should be more comprehensive analysis and evaluation. Adequate examination and accurate diagnosis are crucial, which will directly affect the efficacy of surgery and the prognosis of patients.

2. Introduction

Hemangiopericytoma (HPC) is a rare mesenchymal tumor generally occurring in adults and originating from the pericytes, clinically rare, and easily confused with meningioma [1,2]. Right-sided aortic arch (RAA) is also an infrequent disease of congenital cardiovascular variability with incidence is only 0.1% [3]. RAA is often associated with many secondary vascular mutations. However, the intracranial hemangiopericytoma associated with right-sided aortic arch has not been reported in the literature. We are here

to report a case of right aortic arch concurrent with intracranial hemangiopericytoma, the patient was initially thought to be a simple meningioma, but was eventually diagnosed as a hemangiopericytoma with right aortic arch and multiple intracranial vascular stenosis and malformation. The diagnostic and therapeutic challenges strongly emphasize the importance of this topic for future investigations.

3. Case Presentation

The patient was a 34-year-old female who was admitted with reduced binocular vision accompanied by headache and dizziness for 3 months. The patient began to have headache and dizziness 3 months ago, which appeared intermittently. Since the patient had just completed pregnancy and combined with high blood pressure, the other hospital initially considered to give antihypertensive treatment for pregnancy combined with hypertension. Due to the progressive aggravation of the symptoms, combined with binocular vision loss, nausea and vomiting, walking instability and other symptoms, the patient went to our hospital for further diagnosis and treatment. Our CT showed cerebellar space-occupying lesions, combined with mild hydrocephalus. Physical examination: State of consciousness: sanity, bilateral pupil diameter was about 4.5mm, sensitive to light reflection, papilledema, limb muscle strength was level 5. Enhanced MRI indicate that there was space-occupying lesion in the right cerebellum, growth in the tentorial lobes of the cerebellum, the size of which was about 3.5*2.9*4.3cm, compres-

sion of the fourth ventricle, enlargement of the third ventricle and bilateral lateral ventricles. According to this, a high possibility of meningioma was considered (Figure1). At this time, we all considered that the patients were simple meningioma complicated with hydrocephalus. In order to clarify the blood supply of the tumor, we do the CTA found that: the right aortic arch and left sub-clavicular artery were thickened, there was an irregular mass in the right cerebellum, left cerebellar vein malformation, the left anterior cerebral artery A1 segment was slender, and the left vertebral artery was slender. Further DSA revealed that the tumor was abnormally rich in blood supply, and the right posterior cerebral artery branch was more likely to be the blood supply artery for the tumor (Figure 2). The initial diagnosis was still in favor of meningioma, so no tumor embolization was performed. Postcranial fossa occupying resection was performed under general anesthesia. Considering that the patient had obstructive hydrocephalus and

high intracranial pressure, we performed lateral ventricle puncture and extra-ventricular drainage during the operation to slowly extract cerebrospinal fluid to reduce intracranial pressure. The bone flap was removed and brain tissue was separated. the tumor was exposed above and below the tentorium cerebellum, which showed dark purple, soft texture, rich blood circulation, difficulty in hemostasis and large blood loss, and the operation time was far beyond the expectation. The tumor was completely removed from the subtentorial and supratentorial parts, respectively (Figure 3).

Postoperative pathology indicated hemangiopericytoma. Immunohistochemical: CD34 (+), CK (-), EMA (-), Ki67 (3-5%), S100 (-), Vim (+) (Figure 4). The patient recovered well after the operation, the mass effect was relieved, the fourth ventricle was not significantly compressed, the symptoms of headache and dizziness were improved, and the vision gradually recovered.

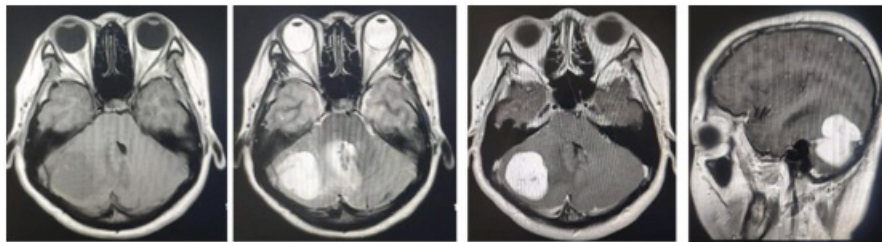


Figure 1: Radiological findings: there was an irregular mass in the right cerebellum which oppresses the fourth ventricle. Axial T1-weighted image showing an isointense mass in the right cerebellum (a), and Axial T2-weighted image showing that the mass is hyperintense (b). After gadolinium administration, the tumor has more enhancement. (c,d) showing a flow void effect (red arrow) in the part of the mass.

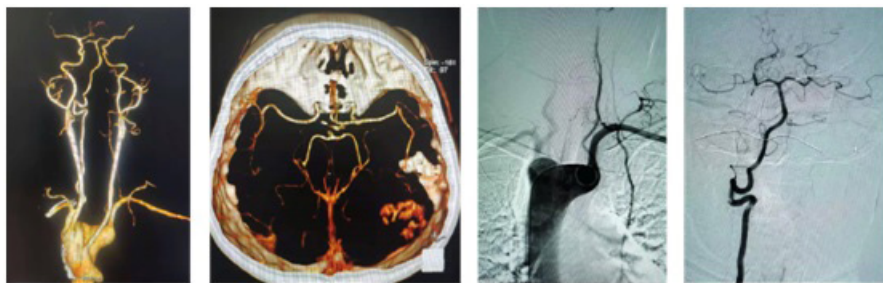


Figure 2 : CTA found that: the right aortic arch and left sub-clavicular artery were thickened, the mass in the right occipital lobe and cerebellum, the left cerebellar vein was malformed, the tumor was abnormally rich in blood supply (a,b), DSA showing that: the right aortic arch and the right posterior cerebral artery branch was more likely to be the blood supply artery for the tumor (c,d).

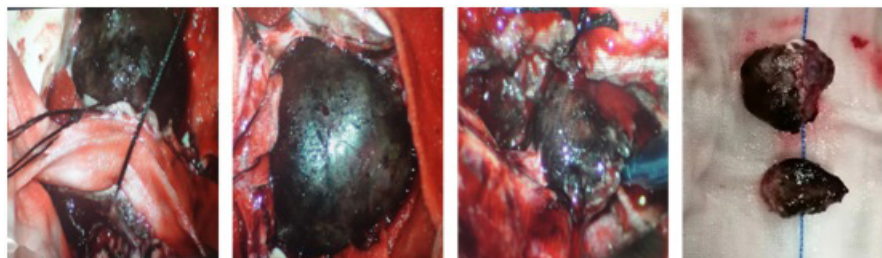


Figure 3: The tumor was bestriding above and below the tentorium cerebellum, which showed dark purple, soft texture, rich blood circulation (a,b,c). The tumor was completely removed from the subtentorial and supratentorial parts respectively (d).

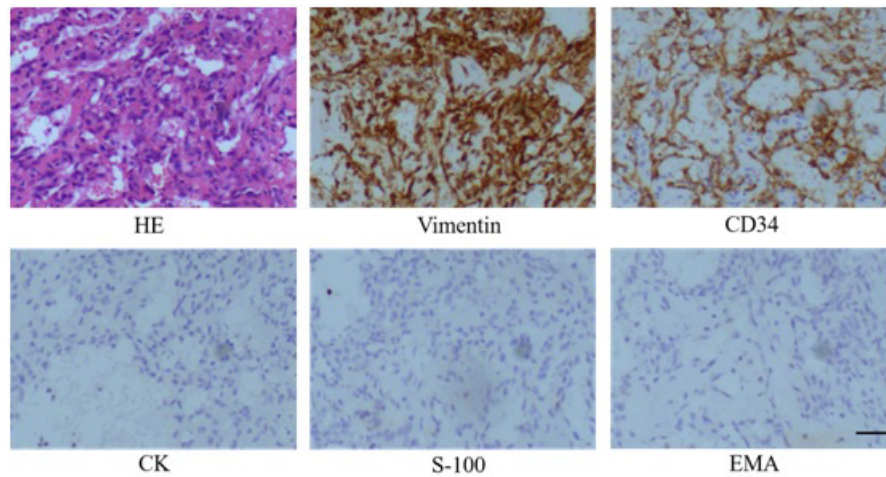


Figure 4. Histopathological examination of the resected tissue taken for biopsy after the operation (scale bar, 50 μ m). Immunohistochemical analysis further indicated that the tumor was immunopositive for Vimentin (+), CD34(+), CK(-), S-100 (-), and EMA (-).

4. Follow-Up

After 3 months of follow-up, the patient showed no obvious symptoms of headache, dizziness, nausea and vomiting, steady walking, improved vision compared with preoperative, normal blood pressure and no need for antihypertensive treatment, as well, no obvious recurrence was observed in imaging examination.

5. Discussion

HPC is a rare intracranial tumor and the differential diagnosis of HPC and meningioma is very easy to be confused. First, the common sites of HPC and meningioma are very similar, both of which tend to occur in the brain's convex surface, next to the falx cerebri and basis cranii. Both have abundant blood supply, so MRI enhancement shows significant enhancement effect on tumor entities, which is more difficult to distinguish when meningiomas tend to be malignant. At present, some clinical identification methods have been summarized, such as tumor shape HPC is mostly lobulated change, meningioma edge is relatively smooth, lobulated rare, mostly circular. In terms of imaging findings, cystic degeneration and necrosis of HPC were common, and T2WI signals and DWI signals of HPC were usually mixed. The signal intensity of DWI and the apparent diffusion coefficient within the tumor are one of the important factors for differentiating HPC from meningioma. HPC mostly presents high signal changes on DWI, while meningioma mostly presents low signal or iso-high signal changes. At the same time, the blood vessels of HPC were more abundant than those of meningioma, and the blood vessels in HPC were often left empty shadow [2,4]. In this case, we learned the following experiences: ① More comprehensive and perfect preoperative examination, such as adding MRI-DWI images ; ② The possibility of partial embolization of the tumor's supplying artery should be considered before surgery ; ③ The possibility of rare diseases

should be fully considered, especially in patients with congenital vascular dysplasia ; .RAA is a congenital aortic arch variation, which can exist in isolation, or can be combined with intracardiac and extracardiac malformations [5], such as ventricular septal defect, aortic straddle, endocardial pad defect, permanent left superior vena cava, transposition of the great artery, tetralogy of Fallot, cleft lip and palate, nasal bone dysplasia, etc. However, there are a few reports on RAA complicated with neurosurgical diseases. In addition to RAA and HPC, this patient also had multiple intracranial vascular abnormalities. First, RAA leads to intracranial hemodynamic changes, which may cause multiple variations of intracranial blood vessels in patients, such as vascular thinning and vascular malformation. It has been reported that the pathogenesis of RAA is related to the chromosomal 22q11.2 microdeletion to some degree [6, 7]. We also found that the pathogenesis of HPC is related to the complementary translocation of 13q22 and 22q11, which may involve the oncogene and fragile site on chromosome 22* [8]. Whether there is a genetic correlation between the two provides a potential research direction for us on if patients with RAA should be highly alert to the possibility of coexistence of intracranial lesions.

In conclusion, the diagnosis and treatment of some rare diseases should be more comprehensive analysis and evaluation. Hence, perfect examination and accurate diagnosis are crucial, which will directly affect the efficacy of surgery and the prognosis of patients.

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