A giant solid cavernous hemangioma mimicking sphenoid wing meningioma in an adolescent

A case report

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Abstract

Rationale: Central nervous system (CNS) solid cavernous hemangiomas are rare extra-axial anomalies that may sometimes resemble meningiomas. Due to their complex vascular nature, accurate preoperative diagnosis is important to avoid disastrous hemorrhage during operation. To the best of our knowledge this is the first case in an adolescent since all middle cranial fossa hemangioma cases reported in literature are adults in their 40s or 50s and all the pediatric cases are cystic.

Patient concerns: We present a case of a 14-year-old girl with headache and dizziness for 3 months. She occasionally experienced nausea and vomiting but denied visual disturbances and loss of smell.

Diagnoses: MRI revealed a lesion that extends to the greater wing of the sphenoid bone as well as the pituitary fossa. Our initial diagnosis was a sphenoid wing meningioma but interestingly, histopathology revealed solid cavernous hemangioma.

Interventions: The residual tumor was completely removed with 2 sessions of Gamma Knife radiotherapy after surgery.

Outcomes: We were confronted with excessive bleeding during surgery so we attained subtotal resection. However, the patient recovered well with no recurrence of the tumor.

Lessons: Our case shows that space occupying lesions involving the cavernous sinus and sphenoid ridged could be easily misdiagnosed as sphenoid wing meningiomas in children and adolescents and even adults therefore great care must be exercised when confronted with this kind of presentation.

Abbreviations: CH = cavernous hemangiomas, CN = cranial nerve, CNS = central nervous system, CT = computed tomography, DSA = digital subtracting angiography, ENT = ear, nose and throat, GCH = giant cavernous hemangiomas, GKS = Gamma Knife radiosurgery, ICA = internal carotid artery, ICP = intracranial pressure, ICU = intensive care unit, MCF = middle cranial fossa, MCFCH = middle cranial fossa cavernous hemangiomas, MRI = magnetic resonance imaging, MRS = magnetic resonance spectroscopy.

Keywords: adolescent, extra-axial, hemangioma, intra-axial, meningioma, mimicking

1. Introduction

Intracranial cavernous hemangiomas (CH) are very rare vascular malformation of unknown etiology. They are mostly limited to intracranial-intraaxial although some are intracranial-extraaxial and constitutes about 5% to 13% of all intracranial vascular malformations.[1–4] The prevalent rate of intracranial CHs is about 0.5% to 1% among general population.[1,5–7] Most intracranial-extraaxial CHs are situated at the cavernous sinus and extends to surrounding structures or at the cerebellopontine angle in the middle cranial fossa.[1,3,7,8] On the other hand, meningiomas arising from the sphenoid wing are benign and...
constituents about 14% to 18% of all intracranial meningiomas.\cite{9,10} The cavernous hemangiomas may develop gradually into a huge size. They become giant cavernous hemangiomas (GCH) when their diameter reaches 6 cm.\cite{11,12} CHs typically appear sporadically and arise as solitary mass with symptoms or asymptomatic.\cite{11,13,14} Familial type of the disorder has been reported and mostly linked to the formation of multiple masses and associated with mutations in numerous chromosomes.\cite{11,15}

CHs are made up of atypical cavernous endothelial-lined spaces lacking smooth muscle and involving neural and vascular structures.\cite{1,16} During their formation, static blood flow leads to thrombosis, hematoma formation, and ultimately calcification inside these channels.\cite{11} The diagnosis of GCHs are mostly problematic on computed tomography (CT) and magnetic resonance imaging (MRI) because many radiologists are unable to differentiate them from neoplastic conditions. Cavernous sinus CHs and for that matter sphenoid wing hemangiomas are frequently misdiagnosed as a meningioma since meningioma is a common lesion are around this location.\cite{1,2} Surgical resection of CH is generally difficult and has serious complications since they are highly vascularized lesions and mostly found very close to vital neurovascular structures.\cite{1,16} Alternative and most effective treatment option is Gamma Knife radiosurgery (GKS) since it has proven to shrink tumor size drastically.\cite{17} We present a rare case of giant solid CH mimicking sphenoid wing meningioma in an adolescent. To the best of our knowledge very little has been said about sphenoid wing CHs.

2. Case report

A 14-year-old girl presented with headache and dizziness for 3 months. She occasionally experienced nausea and vomiting but denied visual disturbances and loss of smell. She had no seizures or muscle twitching. All other systems were grossly normal. Her past medical history was unremarkable and her immunization was completed according to her age. Her parents denied family history of such illness. Physical examination was unremarkable. Neurological examination was also grossly normal. Ophthalmic examination as well as ear, nose and throat (ENT) examination were unremarkable. Routine laboratory investigations were all normal.

Preoperative CT demonstrated a large parasellar mass with slight bone erosion (Fig. 1A and B). MRI also revealed a huge mass measuring about $8.9 \times 6.8 \times 7.4$ cm. The mass was hypo-dense on T1 and hyper-dense on T2 images. The mass was enhanced and had clear boundary with a dura tail or rat tail sign (Fig. 1C–F). The lesion also had a wide base and extended to the greater wing of the sphenoid bone as well as the pituitary fossa. The lesion compressed the adjacent bone with slight infiltration of the bone. There was also significant compression of the adjacent brain matter as well as brain tissue edema. The 2-lateral ventricles plus the third ventricle were significantly compressed with a marked midline shift of the right lateral ventricle and hydrocephalus. There was cerebrospinal fluid extravasation all around. The sphenoid sinus mucosa was also thickened. An initial diagnosis of meningioma with obstructive hydrocephalus and sphenoid sinusitis was made based on the aforementioned radiological findings. The patient was booked for surgery after the family was taken through a series of education and counseling.

Intraoperatively, we did not see any abnormalities on the skull or dura but there was high intracranial pressure (ICP). CSF was decompressed to decrease the ICP after opening the dura. We saw a solid, blue color lesion measuring about $8.9 \times 6.8 \times 7.4$ cm in diameter, firm in constituency with rich blood supply. We were confronted with excessive bleeding during surgery so we could not remove the whole tumor (Fig. 2A). Histopathology revealed a solid cavernous hemangioma (Fig. 2B). Postoperative CT revealed subtotal resection of the lesion (Fig. 2C–E). We suspected left cavernous hemangioma because of the profuse bleeding during surgery. Intraoperative transfusion of 1300 ML red blood cell concentrate and plasma 1250 ML was done. The patient was managed at the intensive care unit (ICU) after the surgery. Postoperative recovery was uneventful. The residual tumor was totally resected via Gamma Knife. She was discharged home on 16 days after operation with scheduled visits every 6 months. Two years follow-up indicated no recurrence. She is well and goes about her daily activities.

3. Discussion

Intracranial CHs located at the middle cranial fossa (MCF) especially the cavernous sinus, sellar, and para sellar regions as well as the sphenoid wings present with subtle inception of gradually advancing symptoms and leads to compression of cranial nerves (CNs) in and around the cavernous sinus as well all neighboring retro-orbital structures.\cite{7,11} Some may even grow into giant sizes and extend into the middle cranial fossa.\cite{7,11} Though the disorder is benign in nature, its enlargement often causes distortion and compression of CN II to VI, with characteristic much like neoplastic disorders.\cite{17} Hemangiomas at MCF are most frequently seen in the fourth and fifth decades of life with a substantial female prevalence of about 7:1 and are more prevalent in the Asian population especially Japanese.\cite{11,21} It is therefore very puzzling to see this kind of presentation in a 14-year-old female adolescent. Estrogen has been implicated to play crucial role in the pathogenesis of this disease although the precise etiology of this disorder is still a matter of debate.\cite{17,18}

Clinical presentation of cavernous hemangiomas located at MCF may include headaches, retro-orbital pain, ophthalmoparesis, hypopituitarism (obesity, amenorrhea), and sensory changes in the face.\cite{19,20} Additional symptoms include proptosis, visual symptoms due to compression of the optic nerves or chiasm. An exacerbation of this disorder associated with pregnancy has been reported meaning that hormonal factors stimulates their pathogenic process.\cite{7,19,21} Focal neurologic deficit, seizure, and devastating life-threatening hemorrhage has also been reported in cases of hemangiomas located at the MCF.\cite{11,22} During surgery, we encountered life-threatening hemorrhage on account of which we did massive blood transfusion. Furthermore, giant lesion may present with increased intracranial pressure (ICP) as a result of obstructive hydrocephalus or as a result of a substantial intracranial neoplastic lesion.\cite{11} Our case also presented with ICP, compressive as well as obstructive symptoms. Some patients may present with acute hemiparesis and cerebral herniation syndromes.\cite{11}

Skull radiographs of CH located at the MCF may reveal expansion or erosion of the sella turcica, greater wing of sphenoid, petrous apex, or clinoïd process.\cite{11,12,19} Studies have indicated that the radiographic appearance of GCHs at the MCF is highly inconstant and comprises of entirely solid, predominately cystic, or heterogeneous masses made up of these 2 constituents.\cite{11,23} The appreciation of typical CHs at the MCF are frequently challenging on standard CT, nevertheless, when detectible, they are seen as hyper-dense parenchymal lesions. However, on MRI, T2-weighted and gradient-echo MRI
sequences show hemosiderin deposition and hematomas in different levels of maturation, leading to the practically pathognomonic form of a circumscribed, heterogeneous lesions with an irregular “popcorn” imaging appearance.\textsuperscript{[1,2,11]} Radiologic enhancement of the lesions is usually very rare, and frequently faint if at all present.\textsuperscript{[11,24]} In most cases, digital subtracting angiography (DSA) is usually normal but may reveal vascular lesions causing blockade of the cavernous sinus. It may reveal tumor staining with description of feeder arteries originating from the meningeal branches of the internal or external carotid arteries.\textsuperscript{[7]}

Magnetic resonance spectroscopy (MRS) may be beneficial since it is able to indicate deficiency of metabolites typically linked with meningioma such as elevated choline, alanine peaks.\textsuperscript{[7,25]} Furthermore, the presence of lipid peak in the non-necrotic area may be evidences against meningioma. On the other hand, middle cranial fossa cavernous hemangiomas (MCFCH) are seen as isodense on MRI usually adjacent to the brain.\textsuperscript{[7,26]} Other most significant CT and MRI discoveries are calcifications, fatty compositions, and flow void. Studies have indicated that with MRI signal intensity characteristic and clinical records, it is not challenging to differentiate MCFCH from most of these

Figure 1. (A and B) Preoperative axial CT showing a large parasellar mass with sphenoid wing and middle cranial fossa involvement. (C–F) Preoperative (axial, sagittal, and coronal) MRI showing the skull base involvement and dural tail sign of the mass. CT = computed tomography, MRI = magnetic resonance imaging.
Figure 1. Continued.

Figure 2. (A) Gross specimen of the resected tumor. (B) Pathological image of the tumor (HE x200). (C–E) Postoperative (axial, sagittal, and coronal) MRI showing the subtotal resection of the tumor. MRI = magnetic resonance imaging.
Solid cavernous hemangioma of the central nervous system is extremely rare and the clinical and radiographic presentation is fairly nonspecific; therefore, an accurate preoperative diagnosis is often difficult to make. The surgical resection of MCFCH in our case was difficult because of the abundant vascular network so we attained subtotal or partial resection. GKS was a very useful optional adjunctive therapy in our case since we could not attain total resection. Two years follow-ups with serially MRIs revealed no recurrence of the lesion.

**Author contributions**

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