A giant invasive parasagittal meningioma with recurrent seizures in a young female: A case report and review of literature

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ARTICLE INFO

Keywords:
Parasagittal meningiomas (PM)
Superior sagittal sinus (SSS)
Seizures
Young female
Angiography

ABSTRACT

Introduction: Parasagittal meningiomas (PM) are very common intracranial lesion but their occurrence in young adult is very rare. Most giant PM are characterized with the invasion of the superior sagittal sinus and form very huge collaterals which makes surgical resection very difficult. The origin of meningioma is still a matter of debate which make our case much more puzzling. We therefore reviewed detailed literature on the etiologies of meningioma as well.

Case presentation: We present a case of 24-years old young lady with history of seizures, blurred vision, intermittent tremors and occasional dropping of objects on the right arm. She had dizziness, headaches, tinnitus and paresthesia on the right side with loss of smell. She had a transient amaurosis fugax once before presenting to us. MRI done revealed a huge tumor compression on the left fronto-parietal gyrus and invading the superior sagittal sinus (SSS). The goal of surgery in this young lady was to preserve the SSS completely, stop frequent seizures and also preserve eloquent areas since the lesion was lying directly on the focal motor area. With a good preoperative evaluation and careful resection of the tumor, we achieved total resection without any further neurologic complications.

Conclusion: We believe the etiology of meningioma in our patients started at childhood and progress to this giant status. We therefore suggest that neurosurgeon's screen children early to detect meningiomas before they advance into giant stages. Two years follow-up after surgery showed no tumor recurrence.

1. Introduction

Meningioma is extremely frequent benign extra-axial idiopathic tumor that originate from arachnoid cap cells in the outer layer of the leptomeninges and may occur anywhere arachnoidal cells are found [1–8]. This tumor is the most universally seen intracranial tumor making up about 20–36% of all primary intracranial tumors with yearly incidence of about 1.8–13 per 100,000 in the general population worldwide [3,5,9–11]. Although the etiology of meningioma is still a matter of debate to many neurosurgeons, some are of the view that these tumors occur sporadically and maybe associated with familial syndrome such as neurofibromatosis 2 (NF2), meningioangiomatosis (MA) and Gorlin syndrome [1,5,12,13]. Wiemels et al. implicated clonal outgrowths from a single mutated cell trigger by chromosomal abnormalities as the origin of meningioma [1]. Studies have shown that deletion or inactivation of the NF2 gene on chromosome 22 is regularly observed on some types of meningioma, while 1p, 6q, 14q, and 18q deletions have been detected in other types [1,5,7,12–15]. Other exogenous influences such as cranial ionizing radiation, hormones, breast cancer, head trauma, cell phone use, family history as well as occupational, diet and allergy have been implicated as increased risk of intracranial meningioma development [1,5,6,12–16–18].

A sizable number of meningiomas originate from the supratentorial tumors usually beside the dura venous sinuses in the cerebral convexity, parasagitally, and in sphenoid wing areas [19,20]. Although cases have been reported at the optic nerve sheath, cerebellopontine angle, and choroid plexus, this location is infrequently affected by meningioma [2,19–23]. The striking features of meningioma at parasagittal region are the invasion of the superior sagittal sinus (SSS) and the enlargement of collateral veins into large ones that enter the sinus in the vicinity of these tumors [4,24–27]. Studies have indicated that these parasagittal meningioma (PM) as well as falcine meningiomas poses various challenges to neurosurgeons and are the second most frequent intracranial meningiomas [4,24,25]. The prognosis as well as surgical resectability are usually determined by the location of the tumor and the surrounding structure. We present a case of a young female with parasagittal meningioma infiltrating the SSS, extending to the falx and causing recurrent seizures who we successfully operated on...
with minimal complication. We further reviewed detailed literature on the etiologies of meningioma.

2. Case report

We present a case of 24-years old young lady with history of seizures and blurred vision which started three months before she presented at our facility (West China Hospital). Her illness started with a transient amaurosis fugax and subsequently seizures which occurs five times a day. She experiences intermittent tremors on the right arm. She was not able to hold firmly on objects with the right and occasional drops objects with this arm. She had dizziness, headaches, tinnitus and paresthesia on the right side with loss of smell. She however could walk as well as run. Her speech had not changed and she could write very well. Her menses were normal. Family history was unremarkable and vaccination was completed according to age. Her parents denied radiation or radioactive substances exposure or any serious viral infection. Systemic examination was unremarkable. Cranial nerve examinations were grossly normal. Ophthalmic as well as ear, nose and throat (ENT) evaluation was also unremarkable. We notice occasional tremors of the right arm. She did not have seizures on admission prior to surgery. All routine laboratory investigations were normal.

MRI done revealed a huge tumor measuring approximately 7.2 × 3.6 × 5.5 cm on the left frontal lobe with invasion of the sagittal sinus (Fig. 1A, B, C, D, F). The mass is significantly enhanced. The left ventricles were significantly under pressure and the falk obviously shifted to the right. The adjacent fronto-parietal gyrus also shifted more to the right. The left fronto-parietal gyrus depressed downwards. The right ventricle slightly dilated indicating a mild hydrocephalus. (Fig. 1A, B, C, D, F). MRI angiogram done confirmed infiltration of the tumor into the sagittal sinus (Fig. 2A, B). A working diagnosis of a giant meningioma with sagittal sinus infiltration was made and the patients scheduled for surgery. The patient as well her family were taken through a series of counseling sessions.

The goal of surgery in the patient was to achieve total tumor resection with maximum preservation of function by preserving the sagittal sinus as well as the eloquent areas of the brain since she was a young adult. Neuronavigation was used through the operation. The patient was put on a supine position with the head fixed tightly with Mayfield three keys fixators. After general anesthesia, the hair was shaved and skin incision site marked (Fig. 3) followed by draping with povidone iodine. Skin incision was made and bleeding secured by applying Raney clips. Initial bone drilling was done and bone osteotomy with high speed drill. Tack-up suture were laid and the dura opened. The tumor was seen attacked to the dura and after careful debulking of the tumor in piece meal fashion, we achieved total tumor resection reserving the sagittal sinus and the eloquent areas. Frozen section done intra-operatively confirmed meningioma. Total hemostasis was secured and bone flap replace and secured with plates and screws. A drain tube was place and skin closed in layers. The patient was sent to neurosurgical intensive care for recovery. Postoperative CT-Scan revealed total tumor resection (Fig. 4A, B). Immunohistochemistry revealed that the tumor cells expresses: EMA (+), S-100 (+), GFAP (−), Ki-67 (MIB-1+ in about 3% to 10%). cell hyperplasia with focal zone seen. MF < 4 cells/10 HPF (Fig. 5A, B). This confirms the diagnosis of meningioma (WHO1). Post-operative management was uneventful. She did not have seizures again and right arm tremor disappeared. She was discharged home two weeks after the operation and out-patient scheduled visited were arranged six months’ interval. Two years follow-up showed no tumor recurrence.

3. Discussion

Meningioma is extremely frequent benign extra-axial idiopathic tumors that originate from arachnoid cap cells in the outer layer of the
leptomeninges and may occur anywhere arachnoidal cells are found [1–7]. This tumor occurs rarely in children and adolescents [28] and have the tendencies of growing into giant sizes and invading surrounding structures in early adulthood if not diagnoses early and treated. This tumor can usually be silent with no obverse clinical manifestations since they are benign and slow growing. Our case is special because of the sex, age and associated seizures and intermittent tremors.

Meningioma has been linked with several familial cancer predisposition syndromes embracing those relating to genes with NF1, PTCH, CREBBP, VHL, PTEN, and CDKN2A [1,29]. Sporadic meningiomas are classically linked with focal chromosomal deletion(s). Uncommon and malignant grades tend to have multiple chromosomal copy number alterations coherent with the possession of “mutator” mutations which promote genomic instability [1,5,30]. While biallelic deletions are more familiar, deletion and inactivation of NF2 on chromosome 22 is a prevalent characteristic of sporadic meningiomas [1,5,31]. Ragel et al. and other authors have indicated that the loss of NF2 happens relatively in 1/3 of patients who demonstrate loss of heterozygosity of chromosome 22 although extra genes may be involved as well [1,32]. Lee et al. and other authors also demonstrated that 14q, 1p, 6q, and 18q are genomic regions that are frequently lost in meningiomas [1,5,33]. Riemenschneider and other authors proposed that the complexity of genetic aberrations in meningioma upsurge with tumor grade [1,18]. Encoding enzymes that alleviate mutilation from reactive oxygen species, metabolism and detoxification enzymes, cell-cycle control proteins, and genes linked with DNA repair mechanisms are similar genes that may affect meningioma. One of such enzymes is Superoxide dismutase (SOD) which scavenges reactive oxygen species to avert DNA damage [1,34]. Exposure to ionizing radiation (IR) is currently the primary environmental risk factor highly associated with meningioma [1,35–38]. Several studies have proven that direct cellular DNA damage or indirect free-radical mediated DNA damage caused by high energy
ionizing radiation [1,5,39]. The source of this ionizing radiation could be from the environment as a result of breakdown of facilities that release damaging radiation into the air, ground, or water [5].

Studies have shown that meningiomas alter in size throughout the luteal phase of the menstrual cycle and pregnancy, and the retrogresses subsequently when estrogen agonist therapy is terminated [1,40,41]. This makes the management of case more complex because of her age and early recurrence of the tumor. Studies have demonstrated that in meningioma, estrogen and progesterone receptors has prognostic connotation because their levels are increase during pregnancy and regresses at menopause [5,40–48]. Progesterone receptor which is good prognostic indicator is highly positivity in WHO grade one (1) tumors [5,42,44,45]. Furthermore, elevated levels of estrogen receptor secretion may be associated with chromosome 14 and 22 alterations as well as molecular markers recognized in meningothelial cell neoplastic modifications [5,42]. Vadivelu et al. and other authors observed that meningiomas may retrogress with termination of progesterone agonist medications [5,41], but there are nonetheless no justifications that exogenous hormone use predisposes women to meningiomas [40,46]. It is however interesting to note that some authors have proven that oral contraceptives and hormone replacement therapy have higher chance of initiating meningiomas [19,46,49–52]. Several studies have linked breast cancer and meningioma with the opinion that common risk factors like endogenous and exogenous hormones as well as mutual genetic predisposition such as variants in DNA repair polymorphisms are usually involved in their etiology [1,40,53,54].

Head trauma has been suggested as a risk factor for meningioma since the time of Harvey Cushing, although the results across studies are not consistent. Whereas the link between head trauma and risk of developing meningioma has been reported by some small case/control studies in both males and females [1,55,56], other studies disagree with such connection [1,57,58]. Although studies have tried to link the cell phone use to the development brain tumors, minute evidence available linking the two because sample sizes specific to meningiomas are comparatively small and follow-up time since onset of cell-phone use is quite short, and, in some occasions, the quantification of cell-phone use is rather crude [1,59–61]. Specific chemicals have used by Claus et al. to demonstrate the connection between meningiomas in occupationally or industrially exposed groups ended indecisively [1,15]. It also very important to indicate that no family based association or isolation studies of meningioma have been described [1].

Ransohoff II indicated that the clinical presentation of PM could be focal motor seizures of the contralateral leg followed in the postictal period by weakness or without seizures as a gradual hemiparesis mostly linked to increased intracranial pressure. He further indicated that while tumors exclusively anterior to the coronal suture, it may present with signs of diffuse cerebral dysfunction, i.e., the "frontal lobe" syndrome, or increased intracranial pressure without localizing signs, those most posteriorly located will apparently tend to present with hemisensory symptoms with or without visual field deficits [62]. The seizures and tremors in our patient could partly be due arteria compressing at the left fronto-parietal gyrus of a developing brain by the meningioma and partly because of intermittent obstruction of the SSS and collateral or both which leads to cerebral hypoxia and then to seizures. Digital subtraction angiography (DSA) and magnetic resonance (MR) angiography are the gold-standard for preoperative assessment of the vascular system [63,64].

Hakuba modified the initial classification of PM proposed by Bonnal and Brotni [65–67]. In this modified classification (Table 1), Type 1 PM adhered to the outer surface of the lateral wall of the SSS while type 2 PM’s situated at the corner of the SSS. In type 3, the PM invades one layer of the SSS while in type 4 the PM invade two layers of the SSS. Furthermore, in type 5, PMs invade one wall of the SSS and protruding outward causing stenotic while type 6 PMs invade the three walls of the SSS leading to severe stenotic. Also, in type 7, the PMs completely obliterated SSS but does not cross to the contralateral side while in type 8, the PM crosses to the contralateral side, invading as well as completely obstructing SSS [66,67].

Studies have indicated that the cortical veins that serve as the normal drainage pathway of the brain, as well as those that may offer important collateral drainage should well be preserved during surgery because when the venous outflow is compromised, venous infarction follows, resulting in brain swelling, hemorrhage, and neuronal death [68,69]. In principle, slow and chronic venous occlusion is well endured, whereas acute occlusion may have disastrous. Andrews et al. further stressed on the preserving collateral drainage since sudden loss of superficial venous drainage due to disruption or thrombosis of the draining veins after surgery

<table>
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<tr>
<th>Type</th>
<th>Location of PM and its effects on the SSS</th>
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<tr>
<td>1</td>
<td>The tumor adhered to the outer surface of the lateral wall of the SSS</td>
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<td>2</td>
<td>The tumor is situated at the corner of the SSS.</td>
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<td>The tumor invades one wall of the SSS and protruding outward causing stenotic</td>
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<td>The tumor invades the three walls of the SSS leading to severe stenotic</td>
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<tr>
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<td>The tumor completely obliterated SSS but does not cross to the contralateral side</td>
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<td>The tumor crosses to the contralateral side, invading as well as completely obstructing SSS</td>
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Fig. 5. A, B; Are HEX 10 and HEX40 immunohistochemical staining of the tumor.

Table 1 The modified classification of PM by Hakuba.
frequently leads to neurological deficits [68,70,71].

With regard of surgical and classification of PMs, studies shown that in types 1 and 2, the surgeon should try as much as possible to preserve the patency of the sinus without graft while in types 3, 4, 5, and 6, the surgeon should reconstruct sinus walls with an autogenous dura or vein graft. The further argue that in types 7 and 8, the resected segment of the sinus may be reconstructed using an autogenous prosthesis with side arms made from the external jugular vein [67]. Studies have proven that with the above suggestions, so many SSS will have to be reconstructed but in reality, sinus reconstruction is rarely practiced because most surgeons think the long patency rate of the autogenous prothetic SSS is still undetermined [67,72]. Studies have shown prudent resection of this kind of tumor and/or radiosurgery. Kondziolka et al. and other authors argue that patients with smaller PMs and patent SSS should have radiosurgery alone as the first surgical procedure while those with larger PM and associated with progressive neurological deficits resulting from brain compression undergo resection and planned second-stage radiosurgery soon after for any residual tumor or neoplasic dura remnant [67,73].

Histopathological estimates of potential growth of a tumor utilizes Ki-67 labeling index which quantities the proliferating cells within a tumor and MIB-1 monoclonal antibody is used to stain the Ki-67 antigen [5,74–76]. It’s an undeniable fact that higher Ki-67 labeling indices are associated with higher tumor grades and predictive of meningioma recurrence [5,74]. Studies have indicated elevated Ki-67 labeling indices in childhood and adolescent meningiomas but elevation does not predict recurrence or prognosis [5,75,79]. Furthermore, epithelial membrane antigen (EMA) is usually positive in 50–100% of Meningioma cells during immunohistochemical studies [3]. Also, GFAP is generally negative in most meningiomas but positive in filament-rich rhabdoid lesion [3,80]. The only exception is seen in variant of meningioma with whirling sclerosing characteristics with GFAP-positive cells although the variant is very rare [3,81,82]. Additionally, positive GFAP cells have been linked with invasive meningiomas that are attached to blood vessels [3,83]. S-100 protein may be helpful in distinguishing meningiomas from schwannomas although 90% of fibrous meningiomas are usually express S-100 protein [3,84].

4. Conclusion

It is very unusual to see giant meningiomas like our case in very young people. The cause of meningioma is still a matter of debate between neurosurgeons. We believe the etiology of meningioma in our patients started at childhood and progress to this giant status hence we suggest that neurosurgeon’s screen children early to detect meningiomas before they advance into giant stages. The goal of surgery in this young lady was to preserve the SSS completely, stop frequent seizures and also preserve eloquent areas since the lesion was lying on top focal motor area. With a good pre-operative evaluation and careful resection of the tumor, we achieved total resection without any further neurologic complication. Two years follow-up showed no tumor recurrence.

Declaration

Ethics approval and consent to participate

The ethical committee of West China Hospital fully approved our case study. The patient and her relatives were informed about our intention to involve her in a case study and she/they agreed to partake in the study. She/they signed the concern form before the operation was carried out according to all surgical protocols.

Consent for publication

The patient and her relatives were dually informed about our intention to publish her case and she/they fully concerted to the use of her information. The West China Hospital also concerted to the use of this information for publication.

Competing interests

All the authors have no competing interest to disclose.

Funding

None.

Authors’ contributions

All the authors contributed equally to the manuscript design and writing.

Disclosure

The authors have no conflict of interest to report.

Acknowledgments

Not applicable.

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