Case Report

Giant Parasagittal Meningioma with Complete Visual Loss in Young Female: a Case Report

Meningioma Parasagital Berukuran Besar disertai Buta Kedua Mata pada Perempuan Muda: Sebuah Laporan Kasus

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ABSTRACT

Parasagittal meningioma is a benign extra-axial tumor from the arachnoid cap cell that fills the parasagittal angle. This case report presents a case of a 21-years old young female with a history of complete visual loss, left side paresthesia, and progressive blunt headache. Brain MRI and MRV revealed a giant enhancing tumor measuring 9.2 cm x 8.41 cm x 7.5 cm on the right parietooccipital lobe with obstruction of the posterior third of the superior sagittal sinus. Gross total removal was achieved. The pathology reports confirmed a WHO grade I meningioma. The paresthesia and headache were improved, but the loss of visions did not change. Giant parasagittal meningioma may invade the superior sagittal sinus leading to intracranial hypertension. The surgical strategy should target the survival and postoperative quality of life. Occlusion of posterior third superior sagittal sinus affects deficiency venous return resulting in visual loss.

Keyword: *Neuro-oncology, parasagittal meningioma, visual loss, young female*

ABSTRAK

Meningioma parasagital merupakan tumor jinak ekstra-aksial yang berasal dari konveksitas hemisfer yang melibatkan dinding dan lumen sinus sagitalis superior. Laporan kasus ini menunjukkan seorang perempuan muda usia 21 tahun dengan keluhan buta pada kedua pandangan mata, kebas sisi tubuh sebelah kiri, dan nyeri kepala progresif. MRI dan MVR *Brain* menunjukkan sebuah tumor besar yang menyangat kontras dengan ukuran 9,2cm x 8,41cm x 7,5cm pada regio parietal oksipital kanan disertai penyumbatan sinus sagitalis superior sepertiga belakang. Pengangkatan tumor total dilakukan. Hasil patologi anatomi menunjukkan tumor meningioma WHO grade I. Keluhan kebas dan nyeri kepala membaik, namun keluhan pandangan mata tidak berubah. Meningioma parasagital berukuran besar dapat melibatkan sinus sagitalis superior sehingga terjadi hipertensi intrakranial. Pemilihan tindakan bedah harus mempertimbangkan angka kelangsungan hidup dan kualitas hidup paska operasi. Penyumbatan sinus sagitalis superior sepertiga posterior menyebabkan aliran balik vena menurun sehingga menimbulkan kehilangan pandangan mata.

Kata Kunci: Buta, meningioma parasagital, neuroonkologi, perempuan muda

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INTRODUCTION

Meningioma is an extra-axial tumor from arachnoid cap cells in the leptomeningeal layer. The etiology of meningioma is still debated. Some neurosurgeons view that meningioma occurs sporadically. It may be associated with a familial syndrome such as Neurofibromatosis 2 (NF2), Meningioangiomatosis (MA), and Gorlin Syndrome. Several exogenous factors increase the risk of intracranial meningioma development, such as cranial ionizing radiation, hormones, breast cancer, head trauma, cell phone use, family history, occupational factor, diet, and allergy (1). Meningioma is more frequently 2.25 times in females than in males. The incidence is 8.44 per 100,000 in females and 3.76 per 100,000 in males (2).

Parasagittal tumors arise from the dura mater on the hemisphere's convexity involving the walls and the superior sagittal sinus (3). Cushing and Eisenhardt first described parasagittal meningioma coming from a parasagittal angle without brain tissue between the superior sagittal sinus and tumor in 1938 (4). Parasagittal meningiomas have a broad spectrum of clinical manifestations and surgical nuances. It develops diagnostic neuroradiology, clinical series, and surgical technique to preserve vascular flow in radical resection management (4).

The giant-sized meningioma may invade the vital neurovascular surrounding structure that makes surgery technically challenging. The tumor can grow to be a large size in area maximal compliance before it is symptomatic. Headache, papilledema, and visual deterioration are the manifestation of this tumor. Complete visual loss can be developed. Papilledema can be followed later by secondary optic atrophy. Giant meningioma is considered a tumor with more than 5 cm in size (5). The location of the tumor and the surrounding structure determined the prognosis of surgery management. This case report presents a giant posterior third parasagittal meningioma causing complete visual loss in a young female.

CASE REPORT

The case report presents a case of a 21-years old young female with a history of loss of both visions for one month before she presented at our facility. She had come to the ophthalmologist facility after she complained of progressively blurred vision for three months. She experienced paresthesia on the left side. She had a progressively dull headache for six months, which more severe in the morning and relieved for a short time after getting analgetic. She could walk as well as run. Her speech had not changed, and she could write well. Her menses were regular. Her first menstrual cycle was at 12 years old. Family history was unremarkable.

Physical examination, cranial nerve examination and the eye movement in six different directions were showed normal figures. Pupillary examination on both eyes showed 4 mm of pupil diameter, no pupil constriction to light in either eye. Fundusoscopic examination showed optic atrophy of both eyes. Ophthalmic examination was found decreasing of visual acuity in both eyes (VODS: No light perception). All routine laboratory investigations were within normal range.

Figure 1. Brain MRI: sagittal

Note: Sagittal sequence of brain MRI shows homogeneous enhancement of tumor.

Figure 2. Brain MRI: axial

Note: Axial sequence of brain MRI shows homogeneous enhancement of tumor.

Figure 3. Brain MRI: coronal

Note: Coronal sequence of brain MRI shows homogeneous enhancement of tumor.

Figure 4. Lateral MRV

Note: Lateral projection of Magnetic Resonance Venography documented occlusion of posterior third of superior sagittal sinus

Figure 5. Anteroposterior MRV

Note: Anteroposterior projection of Magnetic Resonance Venography documented occlusion of superior sagittal sinus

rain MRI with intravenous contrast revealed an extensive, significantly enhancing tumor measuring approximately 9.2cm x 8.41cm x 7.5cm on the right parietooccipital lobes with the sagittal sinus invasion (Figure 1, 2, and 3). This large tumor was considered as a giant right posterior third parasagittal tumor. The falx was shifted to the left. The parietooccipital gyrus depressed downwards. The left lateral ventricle was slightly dilated, indicating hydrocephalus. MR Venography documented occlusion of the posterior third of the superior sagittal sinus (Figure 4 and 5).

A working diagnosis of a giant meningioma with a posterior third of superior sagittal sinus infiltration was made, and the patient was scheduled for surgery. Gross total removal of the tumor with maximum function preservation of the sagittal sinus and eloquent areas of the brain is the goal of surgery to be achieved since the patient was a young adult. The position of the patient was prone, with the head fixed tightly with Mayfield three-pin fixators. After general anesthesia was done, the skin incision was prepared by shaving the hair, marking the skin, and then dropping with povidone-iodine.

Figure 6. Duraplasty

Note: Gross total resection of tumor was followed closure of duramater with dura graft.

Figure 7. Tumor

Note: Piecemeal resection of meningioma.

Figure 8. Bone

Note: Bone flap was infiltrated by tumor.

Figure 9. Microscopic examination

Note: Psammomatous meningioma (hematoxylin and eosin (HE): original magnification, x20). Red arrow shows psammoma body.

Figure 10. Microscopic examination

Note: Psammomatous meningioma (HE: original magnification, x100). The tumor was composed of spindle cells arranged in parallel. Fine chromatin spread smoothly. Tumor cells exhibited eosinophilic cytoplasm.

The skin incision was made, and hemostasis was applied with cauterization. Burr holes were made, and bone

osteotomy was cut by cutting drill bit. Tack-up suture of dura was followed by opening dura. The tumor was seen, and after debulking the tumor in a piecemeal fashion (Figure 6 and 7), total tumor resection was achieved by reserving the sagittal sinus and eloquent area. The tumor infiltrated the bone flap, so it was not replaced (Figure 8). The skin was closed in layers with placing a subgalea drain tube. The patient was further observed in the neurosurgical intensive care unit to reach recovery. The pathology reports confirmed a meningioma (WHO grade I) (Figure 9 and 10). Postoperative management was uneventful while the paresthesia was disappeared gradually, and the headache was improved, but the loss of both visions did not change.

DISCUSSION

This study presents a case of giant parasagittal meningioma grade I in a 21-years old young female who came with a history of complete visual loss, left side paresthesia, and progressive blunt headache. Meningioma is an extra-axial tumor from arachnoid cap cells in the leptomeningeal layer. The etiology of meningioma is still debated. The tumor may arise from anywhere arachnoid cells are found (1). Meningioma can grow into giant sizes with invading surrounding structures. This giant tumor may be found in early adulthood without early treatment because it is usually silent without significant clinical manifestation and benign tumor with slow-growing (1). The exclusiveness of our case is a young female with visual loss.

Studies indicated that mutation in chromosome 22 and multiple chromosomal copy number alteration might have a role in malignant tumor grades (1). Another study showed that loss of genomic region in 14q, 1p, 6q, and 18q are frequently found in meningioma (6). Another study demonstrated that upsurge with meningioma grade is associated with genetic aberration complexity (7). Ionizing radiation-exposed is one of the environmental risk factors of meningioma (1). A higher pathological grade may occur in radiation-induced meningioma (2). In this case report, the patient did not have any family member suffering from any tumor. She neither lives in an area with a high risk of radiation exposure nor gets treatment with radiation.

Several studies showed that alteration of meningioma size was observed in pregnancy and the menstrual cycle's luteal phase. However, the retrogresses of tumor size occur when estrogen agonist therapy is terminated (1). The interesting one is oral contraceptive and hormone replacement therapy increase the chance of initiating meningioma (1). In this case, the menses of the patient was regular as she had menarche at 12 years old.

Harvey Cushing considered head trauma as one of the risk factors for meningioma (1). Peritumoral edema in meningioma associates with the increase in aquaporins expression (AQP4) (8). Expression of Aquaporins (AQP4) is increased in traumatic brain injury (9). Several studies tried to find the link of cell phone use to meningioma, but it was limited to follow-up time since onset of cell phone use, and small sample-sized specific to meningioma (1). Parasagittal meningioma in posterior third of superior sagittal sinus can be undetected for a long time until the tumor mass effect symptoms occur. A study of 154 parasagittal meningiomas showed the distribution of

symptoms of posterior third tumors, such as headache (36%), visual symptoms (21%), focal seizure (21%), or mental status abnormalities (21%) (4).

Previous study reported that 20% of giant meningioma (>5cm) are WHO grade II or III. Furthermore, the tumor size is related to disease-free survival in meningioma that received radiotherapy (10). Grade II tumors grow more quickly than grade I tumors to reach a larger size. Microenvironment factor (hypoxia) may facilitate the progression of a slow-growing tumor to a more aggressive phenotype (11).

Intracranial hypertension can occur due to impaired venous drainage after a posterior third of parasagittal meningioma invading superior sagittal sinus. The manifestation of this type of tumor is headache, papilledema, and visual loss (12). The visual loss mechanism in intracranial tumors with high intracranial pressure is likely due to axoplasmic flow stasis. High CSF pressure surrounding the optic nerves disturbs the normal gradient between intraocular pressure and retrolaminar pressure leading to high tissue pressure within the nerves. The metabolic processes that mediated axoplasmic flow are interrupted by high pressure. Intracranial hypertension caused by space-occupying lesion leads to papilledema. Post-papilledema optic atrophy may occur later (13). Post-papilledema optic atrophy causes visual acuity unchanged after intracranial pressure is relieved. Otherwise, paresthesia disappeared gradually after mass effect in the parietal lobe by the tumor and peritumoral edema was relieved.

Head CT scan and MRI brain are routine imaging for parasagittal meningioma. Hyperostotic changes of the cranium can be evaluated by a non-contrast head CT scan (2). Brain MRI intravenous contrast with Magnetic Resonance Angiography (MRA) and Magnetic Resonance Venography (MRV) are considered as the gold standard diagnostic of meningioma in parasagittal meningioma (14). MR angiography can provide arterial and venous anatomy, sinus patency and invasion, and collateral venous drainage pattern. The advantage of MR angiography is its non-invasive nature over DSA (Digital Subtraction Angiography). MR Venography (MRV) can provide venous infiltration and collateral venous anastomoses (14).

Previous study showed a larger parasagittal meningioma associated with progressive neurological deficits resulting from brain compression underwent resection and planned second-stage radiosurgery soon after any residual tumor or neoplastic dura remnant (1). The pathology reports of this case confirmed a meningioma (WHO grade I). Histopathology estimates tumor potential growth using the Ki-67 labeling index, which quantifies the proliferating cells within a tumor, and MIB-1 monoclonal antibody is used to stain the Ki-67 antigen (1). Ki-67 antigen does not consider age as a factor of labeling indices of meningioma. It is postulated that increased Ki-67 labeling indices do not predict recurrence or prognosis in childhood meningioma (15).

Cortical veins should be preserved during surgery. Compromising venous outflow will be followed by venous infarction, resulting in brain swelling, hemorrhage, and neuronal death. Chronic venous occlusion is well endured. Otherwise, acute occlusion is uncompensated disastrously (16). The number and diameter of bridging veins in the posterior third of the superior sagittal sinus were changed

by invading parasagittal meningioma (14). Giant parasagittal meningioma with progressive neurological deficits from brain compression underwent tumor removal followed by radiosurgery for any residual tumor (1). Giant meningioma has a worse prognosis than non-giant meningioma. Survival and postoperative quality of life is the goal of the surgery. The giant meningioma adversely affects the extent of removal, recurrence rate, postoperative outcome, operative morbidity, and

mortality rates, as well as survival (17).

The conclusion of this case report is a complete visual loss in giant posterior third superior sagittal sinus meningioma can occur as secondary optic atrophy after increasing intracranial pressure due to mass effect and occlusion of venous outflow. The suggestion is that tumor removal can be done as soon as possible in intracranial tumors with visual deterioration.

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