Case Report

Anterior Interhemispheric Approach for Aggressive Rhabdoid Tuberculum Sellae Meningioma: Report of a Rare Case and Literature Review

Ahmad Faried1*, Muhammad Z. Arifin1, Roland Sidabutar1 and Bethy S. Hernowo2

1Department of Neurosurgery, Universitas Padjadjaran–Dr. Hasan Sadikin Hospital, Indonesia
2Department of Pathology Anatomy, Universitas Padjadjaran–Dr. Hasan Sadikin Hospital, Indonesia

Abstract

Tuberculum Sellae Meningioma (TSM) is one of the most challenging operations among neurosurgeons. Many approaches have been proposed in the attempt of total removal of the tumor. Rhabdoid meningioma is a rare subtype of meningiomas accounting for 2-3% of all intracranial meningiomas, an aggressive tumor classified as World Health Organization (WHO) grade III; it occurs mainly in the early childhood, but also rarely in teenagers and adults. It is an aggressive tumor and needs to be treated using both surgery and radiotherapy. We describe the case of a 16-year-old girl with bilateral progressive visual loss. Imaging studies show a tumor mass in the sellar region. Surgery using an anterior interhemispheric approach was performed, and gross total removal was achieved. Histopathological findings established a diagnosis of rhabdoid meningioma. The visual function was improved postoperatively within a week. Rhabdoid tuberculum sellae meningioma, a rare clinical entity in teenagers and adults, had been treated with excellent clinical outcome. This case report supports the suitability of the anterior interhemispheric approach for resection of rhabdoid TSM, followed by radiation therapy.

INTRODUCTION

The Tuberculum Sellae Meningioma (TSM) arises from the tuberculum sellae, chiasmatic sulcus, and limbus sphenoidale. They represent approximately 3-10% of all intracranial meningiomas [1]. The extent of visual deficit is the single most important reason for surgical treatment, and visual outcome is the major concern after surgery. TSM is one of the most challenging operations in neurosurgical field. The tumor has a dural attachment generally at the tuberculum sellae, which may extend anteriorly to the limbus sphenoidale, superiorly to chiasmatic sulcus, and posteriorly to diaphragm sellae. The tumor mass could expand to fill the sellar and suprasellar area, and could displace, stretch, or even encase vital structures; the optic chiasm could be displaced superiorly, the internal carotid arteries may shift laterally, and if the tumor extends posteriorly it may push on the pituitary stalk [2,3]. Based on an anatomical point of view, the safest, most direct surgical access can be achieved through the tumor’s anterior. The bifrontal, unilateral frontal, supraorbital keyhole, and pterional approaches were proposed to resect this tumor; we truly understand that the preferred surgical approach, surgical tactics, and strategies is every individual surgeon’s authority [2,4-6]. The anterior interhemispheric approach was originally developed for anterior communicating artery aneurysms [7], but with several technical modifications, this approach yields minimal morbidity and maximizes visualization around the tuberculum sellae.

From a histopathological point of view, meningiomas are tumors of meningothelial origin and are mostly slow growing tumors with a wide range of histological appearances. Some subtypes, however, are recognized in World Health Organization (WHO) classification as displaying aggressive behavior, including atypical, clear cell, and chordoid meningiomas (WHO grade II);
papillary and anaplastic meningioma variants as WHO grade III. Rhabdoid meningiomas were described for the first time in 1998 as an unusual variant with increased proliferative activity [8]. Later, in 2000, these tumors were included in the revised WHO classification of CNS tumors as an aggressive meningioma corresponding to WHO grade III [9]. These anaplastic meningiomas represent 2-3% of all meningiomas [10]. They are associated with rapid growth, parenchymal invasion, and high recurrence.

CASE PRESENTATION

We present the case of a 16-year-old girl who suffered from vision deterioration of her right eye followed by her left eye over the past 3 months, which worsened one month prior to admission. She also had progressive intermittent headaches for the last year. Neurological examination revealed a visual acuity of 1/> of both eyes (only able to distinguish light perception), and a funduscopic examination suggested papillary edema of both eyes. The patient was thought to have had multiple endocrine abnormalities, as her body size is very small compared to her peers, she had not developed secondary sexual characteristics, and had not experienced menarche. Other cranial nerves were normal, with no motoric or sensoric deficits. Routine laboratory examinations showed normal findings. Laboratory examination of endocrine functions was not performed in this patient because of budgetary constraints. Brain MRI scans exhibited an isohypointense mass at the sellae region on T1-weighted images, enhancing homogenously with a dural tail after gadolinium administration; and an isohyperintense mass without peritumoral edema on T2-weighted images (Figure 1). A preoperative diagnosis of TSM was made. A craniotomy tumor removal was then performed (see surgical technique).

Surgical Technique: Resection Using the Anterior Interhemispheric Approach

Under general anesthesia, the patient was positioned supine with her back elevated about 20 degrees, and the head in a straight line above the heart to facilitate venous drainage. A coronal skin incision behind the hairline was utilized (Figure 2A and B). The scalp was elevated, taking care to reserve the vascularized pericranium medial to the linea temporalis of each side, and preserving the 2 supraorbital nerves. Eight burr holes were used, with the two initial holes made on each side of the orbito temporal region, and the other four holes at the midline. A bifrontal craniotomy was performed (Figure 2C and D). The medial inferior osteotomy was made close to the nasofrontal suture; therefore the frontal sinus must be opened. The frontal sinus mucosa was removed as completely as possible and was packed using tiny pieces of abdominal fat grafts soaked in fibrin glue. A viable flap of pericranium was swung over the frontal sinus and stitched to the frontal base dura at the end of the procedure. A W-shaped dura opening was made, and was reversed anteriorly. The superior sagittal sinus was ligated and divided at its most anterior part (Figure 2E and F). We preserved the bridging veins from both frontal lobes during this maneuver.

Figure 1. T1-weighted triplanar MRI images after application of gadolinum, showing TSM with homogenous contrast enhancement and a dural tail extending from tuberculum sellae in axial (a), coronal (B), and sagittal plane (C).
Next, the interhemispheric fissure just above the knee portion of the anterior cerebral arteries (ACA) was dissected until both callosomarginal arteries were found, with a vertex-down position of the head to allow easy opening of the fissure. Dissection was continued towards the A2 portion of the ACAs and the posterior part of the pericallosal cistern was opened. Then, after elevating the head, the interhemispheric fissure was dissected anteriorly and inferiorly toward the planum sphenoidale. Prior to this procedure, dissection of the both olfactory nerves from the frontal lobe was performed to avoid traction injuries. The anterior part of the tumor was exposed during this step, and special attention was required to preserve the arachnoid plane between the tumor and both rectal gyri. The head was returned to horizontal position, the suprachiasmatic cistern was widely dissected and the TSM was now exposed (Figure 2G and H).

Tumor resection was performed according to the following steps. The tumor was first detached from its attachment with bipolar cautery and debulked. This procedure was started at the planum sphenoidale in the midline, freeing the tuberculum sellae and working straight back and continued just off the midline. During this step, the main tumor feeder arteries from the posterior ethmoidal artery and the superior hypophyseal artery were interrupted, and the tumor devascularized. Then, both optic nerves were identified at the optic canal, and the tumor was debulked to allow dissection of the arachnoid plane separating the nerve and tumor. The of the optic nerve with better visual function was first dissected because it had a better arachnoid plane. By following the arachnoid plane, the chiasm and the anterior communicating artery were gently dissected from the tumor. After sufficient debulking of the tumor, the compromised optic nerve was dissected while preserving the arachnoid plane and the vascular supplies (such as right internal carotid artery). The tumor was then dissected from the pituitary stalk and the interpeduncular cistern. Damage to the small superior hypophyseal branches of the optic apparatus and small perforators from the posterior communicating artery was avoided.

The macroscopic appearance of the tumor was multiple bits of firm, irregular sized grayish white tissue. The mass was firm to hard. Cut section was whitish and solid. Microscopic sections showed tumor cells arranged in sheets and small loose clusters (Figure 3A). Significant foci of hyperplastic large, round to polygonal cells, with abundant pale eosinophilic cytoplasm, central to eccentrically placed vesicular nuclei and prominent nucleoli with irregular mild atypia maligna (rhabdoid cells) were appreciated and intranuclear cytoplasmic pseudoinclusions were present at places; stromal tissues shown hyalinization and necrotic area appeared in some places with capiler dilatation and cluster of small bleeding (Figure 3B). The final histopathological report indicated rhabdoid meningioma. Gross total tumor removal through surgical intervention was achieved and the visual function was improved up to normal within a week postoperatively.

DISCUSSION

Tuberculum sellae meningiomas arise from the dural midline over the tuberculum sellae, chiasmatic sulcus, and limbus sphenoidale; accounting for 3-10% of all intracranial meningiomas [1]. The narrow anatomical relationship between this area and the optic tract explains the early visual disturbances secondary to the displacement of the optic chiasm and nerves by TSMs [11], the most common initial symptom. The resection of TSMs was commonly performed via a lateral subfrontal approach using pterional or unilateral frontal craniotomy [12], but an anterior subfrontal approach using free bifrontal flap craniotomy [13] has also been proposed.

There are many publications of surgical approaches to TSM since it was first reported. All of these approaches have their advantages and disadvantages, but the most commonly used approaches are the pterional, unilateral frontal, frontolateral, and bifrontal approaches with their modifications, and the
The final histopathological report revealed rhabdoid meningioma. Microsection showed tumor cells arranged in sheets and small loose clusters (A) (Hematoxylin and eosin, x50). Significant foci of hyper plastic large, round to polygonal cells, with abundant pale eosinophilic cytoplasm, central to eccentrically placed vesicular nuclei and prominent nucleoli with irregular mild atypia maligna (rhabdoid cells) were appreciated and intranuclear cytoplasmic pseudoinclusions were present at places; stromal tissues shown hyalinization and necrotic area appeared in some places with capillary dilatation and clots of small bleeding (B) (Hematoxylin and eosin, x400).

CONCLUSION

We are the first to report a case of rhabdoid TSM in a 16-year-old girl with a high grade meningioma that was seen to emerge as a recurrent growth during the process of malignant transition and have poor outcome. This clinical vignette justifies the use of surgery and aggressive adjuvant treatment such as radiation therapy, which we have performed using an anterior interhemispheric approach.

CONSENT

Informed consent was obtained from the patient for publication of this case report and any accompanying images. Her family was present at the time.

AUTHORS’ CONTRIBUTIONS

AF, MZA and RS had examined, treated, observed, and followed up the patient. AF and RS performed the operation on the patient. BSH carried out the histopathological studies and interpreted the results of the patient’s tissue. All authors participated in writing the manuscript. All authors have read and approved of the final manuscript.

ACKNOWLEDGEMENT

The authors would like to thank Dhira Atmanand Agung B. Sutono from Department of Neurosurgery, Faculty of Medicine Universitas Padjadjaran–Dr. Hasan Sadikin Hospital for their illustration and technical assistance.

REFERENCES


Cite this article