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Original Article

Intracranial solitary fibrous tumor/hemangiopericytoma – A case series

Chi-Man Yip, Shu-Shong Hsu, Wei-Chuan Liao, Szu-Hao Liu, Yung-Shang Lin, Yu-Hone Hsu, Huang-I Hsu, Yu-Wen Cheng, Yu-Lun Wu

Division of Neurosurgery, Kaohsiung Veterans General Hospital, Zuoying, Kaohsiung, Taiwan.

E-mail: *Chi-Man Yip - yip_chiman@yahoo.com; Shu-Shong Hsu - sshsu@vghks.gov.tw; Wei-Chuan Liao - wcliao@vghks.gov.tw; Szu-Hao Liu - shliu@vghks.gov.tw; Yung-Shang Lin - yslin@vghks.gov.tw; Yu-Hone Hsu - yhhsu1971@vghks.gov.tw; Huang-I Hsu - hihsu@vghks.gov.tw; Yu-Wen Cheng - ywcheng1015@vghks.gov.tw; Yu-Lun Wu - ylwu@vghks.gov.tw

***Corresponding author:**
Chi-Man Yip,
Division of Neurosurgery, Kaohsiung Veterans General Hospital, Zuoying, Kaohsiung, Taiwan.

yip_chiman@yahoo.com

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ABSTRACT

Background: Intracranial solitary fibrous tumor/hemangiopericytoma (HPC) is a rare and aggressive tumor. We conducted this retrospective study to investigate the outcome of patients after treatment, the efficacy of postoperative adjuvant radiotherapy, and the factors not conducive to total resection.

Methods: We conducted a retrospective review of the medical records of patients harboring fresh intracranial solitary fibrous tumor/HPC treated from January 2009 to December 2019 in our hospital. We reviewed their clinical presentations, radiologic appearances, tumor size and location, extent of resection, estimate intraoperative blood loss, treatment modalities and results, and duration of follow-up.

Results: There were seven consecutive patients (three males and four females). The ages of the patients at the time of diagnosis ranged from 35 to 77 years (mean: 52.86 years). Five patients (71.43%) got tumor bigger than 5 cm in dimension and only 1 patient (14.29%) underwent gross total tumor resection in the first operation without complication. Five patients (71.43%) underwent postoperative adjuvant radiotherapy. Follow-up period ranged from 4.24 to 123.55 months and the median follow-up period was 91.36 months. Three patients had favorable outcome with Glasgow Outcome Scale (GOS) equal to 4; four patients had unfavorable outcome with GOS equal to 2 or 3. No mortality was happened.

Conclusion: Gross total tumor resection in the initial surgery is very important to achieve a better outcome. Massive intraoperative bleeding and venous sinus or major vessels adjoining are factors not conducive to total resection. Radiotherapy can be administered as adjuvant therapy for cases showing an aggressive phenotype or not treated with gross total resection.

Keywords: Intracranial solitary fibrous tumor/hemangiopericytoma, Massive intraoperative bleeding, Postoperative adjuvant radiotherapy

INTRODUCTION

Before the establishment of 2016 WHO classification of central nervous system (CNS) tumors, solitary fibrous tumor (SFT) and hemangiopericytoma (HPC) represented separate entities. The 2016 WHO classification of CNS tumors combined intracranial SFT and HPC into a single disease entity because of the discovery of NAB2-STAT6 fusion using whole-exome sequencing.[5,10] Intracranial HPC was first reported by Begg and Garret in 1954, which accounts less than 1%

of all primary CNS tumors.[4] HPC is a highly vascularized mesenchymal tumor that develops from pericytes which comprise the walls of capillaries and postcapillary venules with aggressive behavior.[4] We would like to review our cases of intracranial SFT/HPC based on the 2016 WHO classification of CNS tumors to investigate the outcome of patients after treatment, the efficacy of postoperative adjuvant radiotherapy, and the factors not conducive to total resection of the tumor.

MATERIALS AND METHODS

We conducted a retrospective review of the medical records of patients having newly diagnosed intracranial SFT/HPC who were treated in our hospital between January 2009 and December 2019. Their age at diagnosis, gender, clinical presentation, radiologic appearances, tumor location, tumor size, extent of resection, estimate blood loss in the first tumor resection surgery, procedure-related complication, WHO grading of the tumor, duration of follow-up, adjuvant therapy, and outcome were reviewed. The extent of resection was deduced from the operative records and the reports of the early postoperative imaging. We used Glasgow Outcome Scale (GOS) to score the outcome of our patients.

RESULTS

There were seven consecutive patients (three males and four females) having newly diagnosed intracranial SFT/HPC treated in our hospital between January 2009 and December 2019. The age of the patients at the time of diagnosis ranged from 35 to 77 years (mean age was 52.86 years). Their initial clinical presentations included focal neurological deficits as well as neuropsychological decline that were related to the tumor location and signs of increased intracranial pressure. The locations of tumors were as follows: convexity = 1; falcine/parasagittal = 2; sphenoidal ridge = 1; cerebellopontine angle = 1; and tentorium cerebelli =2. All tumors had closed attachment or invasion to venous sinus or major vessels. Superior sagittal sinus was involved by the two parasagittal located tumors; transverse sinus was involved by the two tentorium cerebelli located tumors; for the tumor located at the sphenoid ridge, the cavernous sinus, middle cerebral artery, and supraclinoid internal carotid artery were invaded and encased. The cerebellopontine angle located tumor attached the vertebral artery and basilar artery. The convexity located tumor coexisted an unruptured aneurysm of the right middle cerebral artery that had attachment to the tumor [Table 1]. This particular case underwent total tumor resection and complete surgical clipping of the aneurysm in the initial surgery which had been reported in detail.[12]

All of these seven patients had performed brain magnetic resonance imaging (MRI) to study the intracranial lesions before management. On T1-weighted image, the lesion was isointense with cortical gray matter in six cases and hypointense in one case. On T2-weighted image, the lesion was hyperintense in four cases and isointense in three cases. After gadolinium injection, six cases showed heterogeneous enhancement and one case showed homogeneous enhancement [Figures 1-3]. We had 5 patients (71.43%) got tumor bigger than 5 cm in dimension [Table 1].

A total of 14 operations were performed on these seven patients for the resection of either primary or residual or recurrent intracranial SFT/HPC. Six patients (85.71%) underwent partial tumor resection in the initial surgery and only 1 patient (14.29%) (case 7) underwent gross total tumor resection in the first operation without complication; the detail of this patient had been reported previously.[12] The recorded estimate blood loss in the first tumor resection surgery ranged from 300 ml to 3000 ml and the mean estimate blood loss was about 1543 ml. One patient (Case 2) performed preoperative embolization before the first tumor resection surgery, but the recorded estimate blood loss in the first tumor resection operation was 3000 ml. Five patients (71.43%) underwent postoperative adjuvant radiotherapy. One patient (case 6) refused further postoperative adjuvant radiotherapy after the

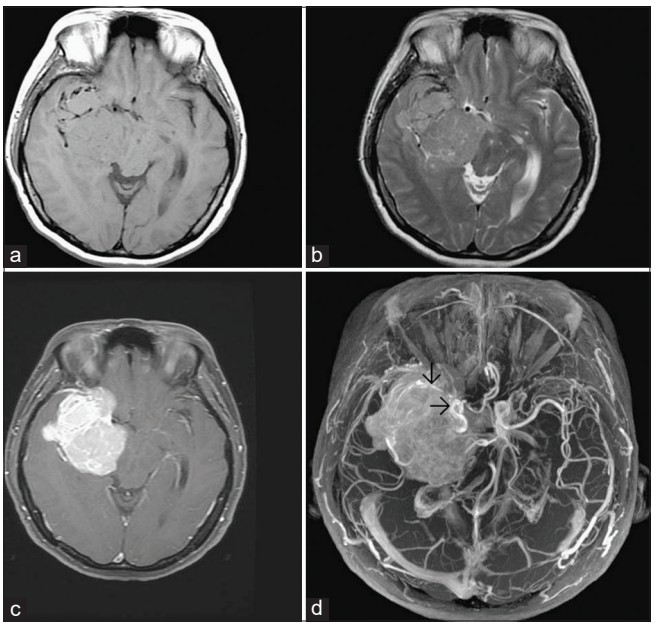

Figure 1: Preoperative brain MRI of case 2. Axial T1-weighted image (a) and axial T2-weighted image (b) showing a large tumor in the right side frontotemporal region, abutting the sphenoid ridge and right cavernous sinus, extension to the right basal ganglion and right side periventricular region with relative isosignal intensity on T1WI and T2WI. Axial T1-weighted image postgadolinium enhancement (c) showing good enhancement. MRA (d) partial encasement of the right middle cerebral artery and right supraclinoid internal carotid artery (black arrows).

Table 1: Demographic and clinical characteristics of patients.

Case No.	Age/ gender	Clinical features	Tumor location/size (cm)	MR feature (T1/T2/ enhancement)	Venous sinus or major vessel attachment	Estimate blood loss in the 1st op (ml)	WHO grade	Treatment modality	Follow-up period (mo)	GOS
1	45/M	Lt lower limb weakness	Rt high parietal parasagittal/6.5	Iso/hyper/ heterogeneous	Superior sagittal sinus	2100	2	Tumor resection x 2	123.55	4
2	51/F	Neuropsychological decline & IICP	Rt sphenoid ridge/6.0	Iso/iso/ heterogeneous	Rt cavernous sinus, Rt MCA, Rt supraclinoid ICA	3000	2	Tumor resection x 3 + R/T 60 Gy/30Fx	91.36	2
3	36/F	Blurred vision and IICP	Rt tentorium cerebella/7.8	Iso/hyper/ heterogeneous	right transverse venous sinus	3000	3	Tumor resection x 4 + R/T 60 Gy/30Fx+GKRS	111.19	3
4	35/F	Unsteady gait	Rt CP angle/3.0	Hypo/hyper/ homogeneous	Rt VA, basilar artery	350	2	Tumor resection x 2 + Cyberknife +GKRS	98.1	3
5	52/M	Rt homonymous hemianopia and IICP	Lt tentorium cerebella/6.2	Iso/hyper/ heterogeneous	Left transverse sigmoid sinus	850	2	Tumor resection x 1 + R/T 60 Gy/30Fx	74.37	4
6	77/F	Lt limbs weakness	Right parietal parasagittal/4.0	Iso/iso/ heterogeneous	Superior sagittal sinus	300	2	Tumor resection x 1	35.47	3
7	74/M	Unsteady gait, neuropsychological decline	Rt frontal-temporal convexity/7.7	Iso/iso/ heterogeneous	Rt MCA	1200	2	Tumor resection x 1 + R/T 54 Gy/27Fx	4.24	4

M: Male, F: Female, Rt: Right, Lt: Left, IICP: Increased intracranial pressure, CP angle: Cerebellopontine angle, ICA: Internal carotid artery, MCA: Middle cerebral artery, VA: Vertebral artery, R/T: Radiotherapy, GKRS: Gamma knife radiosurgery, Fx: Fraction, mo: Months, GOS: Glasgow Outcome Scale

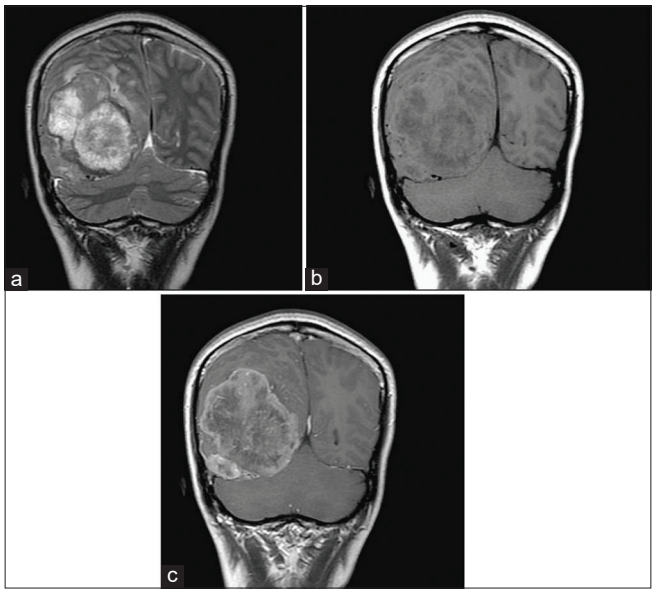

Figure 2: Preoperative brain MRI of case 3. Coronal axial T2-weighted image (a) and coronal T1-weighted image (b) showing a large tumor in right fronto-occipital-parietal region, abutting the posterior cerebral falx and right-sided tentorium and invading right occipital bone, isointense with cortical gray matter on T1-weighted image but hyperintense on T2-weighted image. Coronal T1-weighted image postgadolinium enhancement (c) showing heterogeneous enhancement.

initial partial tumor resection. One patient (case 1) suspended the postoperative adjuvant radiotherapy because the follow-up imaging showed no obvious recurrence after two tumor resection procedures [Table 1].

The histopathology of these seven patients was reviewed [Table 1], six of them were intracranial SFT/HPC WHO Grade 2, but one of them (case 3) was intracranial SFT/HPC WHO Grade 3 (anaplastic HPC), who had left femur metastasis 98 months after the diagnosis. For those patients having intracranial SFT/HPC WHO Grade 2 did not have metastasis.

Within these seven patients, 3 patients (42.86%) had favorable outcome with GOS equal to 4; 4 patients (57.14%) had unfavorable outcome with GOS equal to 2 or 3. Two patients (case 2 and case 3) (28.57%) got procedure-related complications including postoperative intracranial bleeding and cerebrospinal fluid leakage, respectively, and needed surgical intervention. No mortality was happened. The follow-up period of these seven patients ranged from 4.24 to 123.55 months and the median follow-up period was 91.36 months [Table 1].

DISCUSSION

HPC was described in 1942 by Stout and Murray, which accounts less than 1% of all primary CNS tumors.[6]

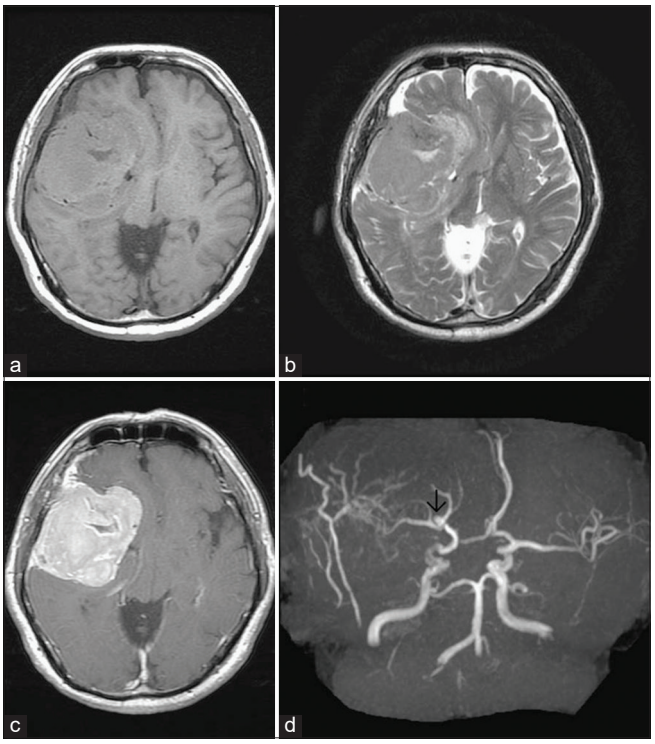

Figure 3: Preoperative brain MRI of case 7. Axial T1-weighted image (a) and axial T2-weighted image (b) showing a large extra-axial well-circumscribed mass lesion in the right frontal-temporal convexity, abutting the skull bone, isointense with cortical gray matter on both T1-weighted and T2-weighted image. Axial T1-weighted image postgadolinium enhancement (c) showing good enhancement. MRA (d) showing a small aneurysm in bifurcation of the right middle cerebral artery (black arrow).

Intracranial HPC was first reported by Begg and Garret in 1954.[7] HPC is a highly vascularized mesenchymal tumor that develops from pericytes which comprise the walls of capillaries and postcapillary venules with aggressive behavior.[6,9] It has high recurrence rate and can metastasize extracranially to bones, liver, lungs, abdominal cavity, lymph nodes, skeletal muscle, kidney, pancreas, skin and subcutaneous tissue, breast, adrenal glands, gallbladder, diaphragm, retroperitoneum, and heart.[4,8] SFT was first described in the pleura in 1931. SFT is spindle cell mesenchymal tumors which was reported more frequently in the visceral pleura and liver, skin, orbits, and paranasal sinuses.[3] SFT in the CNS initially reported by Carneiro *et al*. in 1996.[5]

Imaging features of HPC include the following: broad-based attachment to the dura, lack calcifications and hyperostosis, multilobulated tumors, heterogeneously hyperdense tumors with focal areas of hypodensity on unenhanced brain computed tomography (CT), heterogeneous or homogeneous enhancement on enhanced brain CT, and isointense with cortical gray matter on T1- and T2-weighted brain MRI and

show heterogeneous enhancement on gadolinium-enhanced T1-weighted brain MRI.[2] CT features of intracranial SFT seem similar to those of extracranial SFT, with isoattenuation and intense enhancement after intravenous iodinated contrast injection. Brain MRI features of SFT demonstrating an extra-axial, multilobulated, and heterogeneous signal intensity on T2-weighted image, hypointense T2 areas with good enhancement after gadolinium administration, but the adjacent meninges have no enhancement.[3]

Tihan *et al*. found that both SFT and HPC had the same microscopic features including spindle to oval cells, "staghorn" vascular pattern, biphasic architecture, and hyalinized vessels. In the field of immunohistochemistry, both tumors showed positive to CD34, Bcl-2, Factor XIIIa, and vimentin, but negative to epithelial membrane antigen and S100 protein.[11] Detection of NAB2-STAT6 fusion using whole-exome sequencing in both HPC and SFT supports that they are two variants of a single tumor entity.[5,10] Symptoms of these two tumors depend on the location of the tumor, so SFT and HPC have overlapping features clinically and radiologically.[1] From 2016, the WHO classified SFT and HPC as one entity and named SFT/HPC, which has three grades. Grade 1 most often corresponds to highly collagenous, relatively low cellularity spindle cell lesion previously diagnosed as SFT. Grade 2 typically corresponds to more cellular, less collagenous tumor with plump cells and staghorn vasculature that was previously diagnosed as hemangiopericytoma in the CNS. Grade 3 corresponds to what was termed "anaplastic hemangiopericytoma" in the past; diagnosed on the basis of ≧ mitoses/10 high-power field.[5]

Our case number and clinical experience of this unusual tumor are limited. It is hard for us to do any statistical analysis due to limited case number. As compared to other studies,[4,6,7] our gross total resection rate in the first surgical intervention was low; large tumor size, massive intraoperative bleeding, and major blood vessels involvement are factors leading to difficult gross total resection of the tumor. Stage operation and preoperative feeding artery embolization have been proposed to reduce blood loss during surgery,[4,7] however, the only one patient (case 2) who performed preoperative embolization before the first tumor resection surgery still had massive intraoperative bleeding. We had applied stage operation strategy to case 1 who underwent two tumor resection procedures to achieve gross total removal of the tumor. Postoperative adjuvant radiotherapy showed positive effect on tumor control.[5-7] We provided postoperative adjuvant radiotherapy to all of our patients except case 6 whose family refused any further therapy and case 1 who preferred to undergo radiotherapy if tumor recurrence detected on regular imaging follow-up.

CONCLUSION

Gross total tumor resection in the initial surgery is very important to achieve a better outcome. From our limited cases, we notice that massive intraoperative bleeding and venous sinus or major vessels adjoining are factors not conducive to total tumor resection. Radiotherapy including conventional radiotherapy or stereotactic gamma knife radiosurgery can be administered as adjuvant therapy for cases showing an aggressive phenotype or not treated with gross total resection. Improving the handling of intraoperative bleeding including the improvement of surgical strategy and hemostatic agents, further basic research of the nature of this particular tumor, especially the biomolecular field, will help us to advance our treatment.

Declaration of patient consent

Institutional Review Board permission obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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