American Journal of Clinical Neurology and Neurosurgery
Vol. 6, No. 1, 2021, pp. 7-10
http://www.aiscience.org/journal/ajcnn
ISSN: 2471-7231 (Print); ISSN: 2471-724X (Online)

A Rare Case of Recurrent Hemangiopericytoma Pituitary Fossa Treatment

Sujata Sarkar*, Irfan Bashir, Roopesh Reddy Yotham

Department of Radiotherapy, Batra Hospital and Medical Research Centre (BHMRC), New Delhi, India

Abstract

Hemangiopericytoma (HPC) is a tumor that arises from the pericytes of capillaries. It is a variety of soft tissue sarcoma. HPC is often asymptomatic in early stages. HPCs can be either benign or malignant. Malignant HPCs can metastasize or spread to other areas in the body, primarily the lungs and bones. HPCs are one of the least common intracranial tumors, accounting for less than 1% of cases reported. Pituitary fossa is one of the rarest sites among all cases reported. HPCs are positive for vimentin, negative for desmin, CD31, cytokeratin, S-100. Recently, STAT6 positivity has been shown to be very specific of HPC. Even with appropriate treatment, 80% of HPCs recur and 23% metastasize, making it rare yet very aggressive tumor. The high recurrence rate makes it difficult for patient to undergo repeated surgeries, especially in HPCs of central nervous system (CNS). Here, we are reporting a case of Hemangiopericytoma in pituitary fossa in a 55 year old male, which recurred to a large size of > 4 cm within a year of previous surgery. Due to its large size and close proximity to optic chiasm, both surgery and radiotherapy were critical for this patient. Patient was treated with radiotherapy by volumetric modulated arc technique (VMAT). VMAT has high efficacy in treating tumors that are in close proximity to critical tissues with least toxicity. Radiotherapy improves local control and increases overall survival irrespective of previous surgical outcome. Following radiotherapy, his symptoms subsided and no disease progression is seen on 3 months follow up.

Keywords

Hemangiopericytoma, Pituitary Fossa, Radiotherapy, VMAT

Received: June 28, 2021 / Accepted: August 3, 2021 / Published online: August 20, 2021

1. Introduction

Hemangiopericytomas(HPCs) arise from Zimmermann pericytes found in the endothelium of capillaries and venules; they are mesenchymal in origin, aggressive in behaviour and well-vascularised tumors [1]. They have a very low incidence rate, 0.060 per 100,000 population in 2016 [2]. They are classified under mesenchymal, non-meningothelial tumors (Grade I, II or III) in the 2016 WHO classification of CNS tumors, accounting for less than 1% of all intracranial tumors [3]. Despite of the best treatments, recurrence rate of HPC is 80% and 23% will metastasize [4]. The tumor cells in hemangiopericytoma are positive for vimentin, STAT6 [5]. Few HPCs are focally positive for actins; nearly half of HPCs are positive for CD34 and CD57. HPCs are always negative for desmin, CD31, cytokeratin, and S-100 [6].

Here, we are presenting a case of Hemangiopericytoma in pituitary fossa, which recurred within a year of previous surgery. Patient had received adjuvant radiotherapy, followed which his symptoms subsided and no disease progression was seen on 3 months follow up.

2. Case Report

A 55 year old male, Russian, had history of headache and vomiting in December 2019. MRI brain showed a well

* Corresponding author
E-mail address: sujatasarkar1989@gmail.com (S. Sarkar), maildrirfan@gmail.com (I. Bashir), roopesh6666@gmail.com (R. R. Yotham)

defined space occupying lesion in pituitary fossa. He underwent surgery in Tazikistan in January 2020. Histopathology report showed high grade mesenchymal neoplasm, features favouring Hemangiopericytoma(WHO grade III) in view of STAT-6, bcl2 and CD99 positivity. Approximately after 1 year in December 2020, he again had symptoms of headache and vomiting. CEMRI brain showed a large well-defined lobulated mass of 3.1x4.4x2.6cms in sella, supra-sellar region, extending inferiorly to sphenoid sinus, lower part of 3rd ventricle, abutting optic chiasm(L>R), partially encasing cavernous part of bilateral internal carotid arteries, extending into bilateral cavernous sinuses. The pituitary gland and infundibulum weren't separately visualized. PET-CT was also done which didn't reveal any disease in any other part of the body.

Trans-sphenoidal biopsy was done. Microscopic section showed cellular mesenchymal tumor arranged in diffuse sheets with interspersed dilated blood vessels. Tumor cells display marked nuclear pleomorphism, round to ovoid in shape, with round to ovoid nuclei, dense chromatin, rare conspicous nucleoli. Many pleomorphic and multinucleated tumor giant cells were seen. No necrosis, mitotic count 7-8/ 10 hpf. Immunohistochemistry was positive for STAT6, CD99, bcl2 in few cells, Vimentin, Retic, CD31 in vessels; negative for TTF1, S100, PR, EMA, synaptophysin, desmin; Ki67 7-8% (Figure 1).

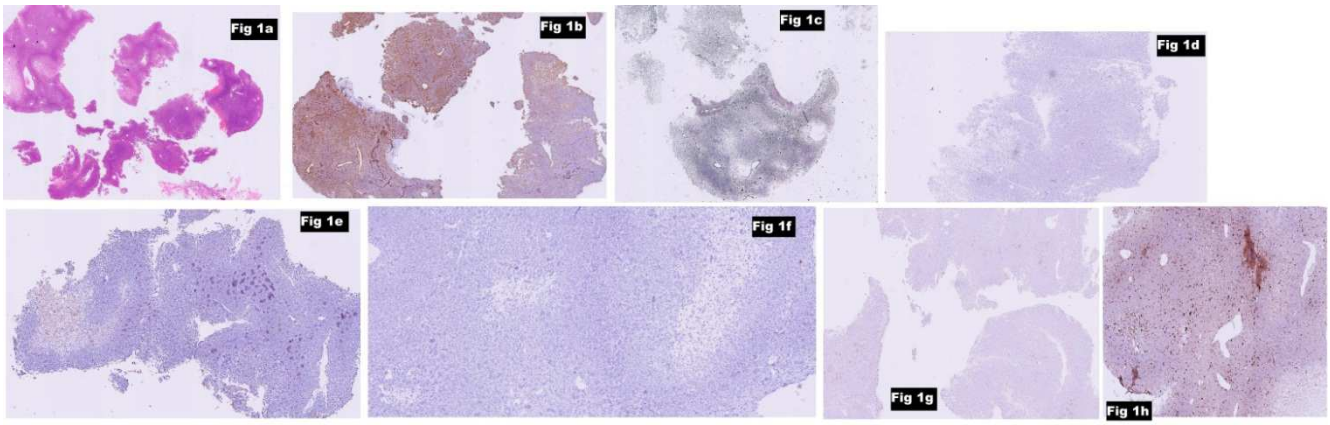

Figure 1. Figure 1a. Photomicrograph showing cellular mesenchymal tumor arranged in diffuse sheets with interspersed dilated blood vessels (H&E 4x). Figure 1b. Photomicrograph of Vimentin +ve, Figure 1c. Photomicrograph of Retic +ve around tumor cells, Figure 1d. Photomicrograph of S100 negative, Fig1e. Photomicrograph of EMA negative, Figure 1f. Photomicrograph of synaptophysin negative, Figure 1g. Photomicrograph of desmin negative, Figure 1h. Photomicrograph of Ki67 7-8%.

Patient visited our OPD in February 2021. Systemic examination showed no evidence of metastasis. Pituitary hormone levels were within normal limit. He had multiple comorbidities including hypertension and is on cardiac medications.

Patient underwent radiotherapy in March 2021 to a dose of 54Gy/30 Fractions with VMAT (Figure 2) which ensures least toxicity to surrounding critical structures called "Organ at risks (OAR)". OARs doses were within limits (Table 1). Patient tolerated radiation well with no complication during or post radiation. CEMRI brain after 3 months showed no disease progression. Patient is currently asymptomatic and is on follow up.

Table 1. Organ at risks (OARs) contoured with dose constraints to be achieved as per literature and dosesachieved.

OARs	DOSE CONSTRAINT	DOSE ACHIEVED
Brainstem	Max <54Gy (QUANTEC)	54 Gy
	1% <60Gy	51.3Gy
Lt Optic Nerve	Max <54Gy	44 Gy
Rt Optic Nerve	Max <54Gy	46 Gy
Lt Eye Lens	Max <7Gy(RTOG 0539)	5.7 Gy
Rt Eye Lens	Max <7Gy(RTOG 0539)	5.3 Gy
Lt Eye	Mean <35Gy (RTOG 0225)	8.7 Gy
Rt Eye	Mean <35Gy (RTOG 0225)	9.6 Gy
Lt Cochlea	Mean <45Gy (QUANTEC)	17 Gy
Rt Cochlea	Mean <45Gy (QUANTEC)	17 Gy

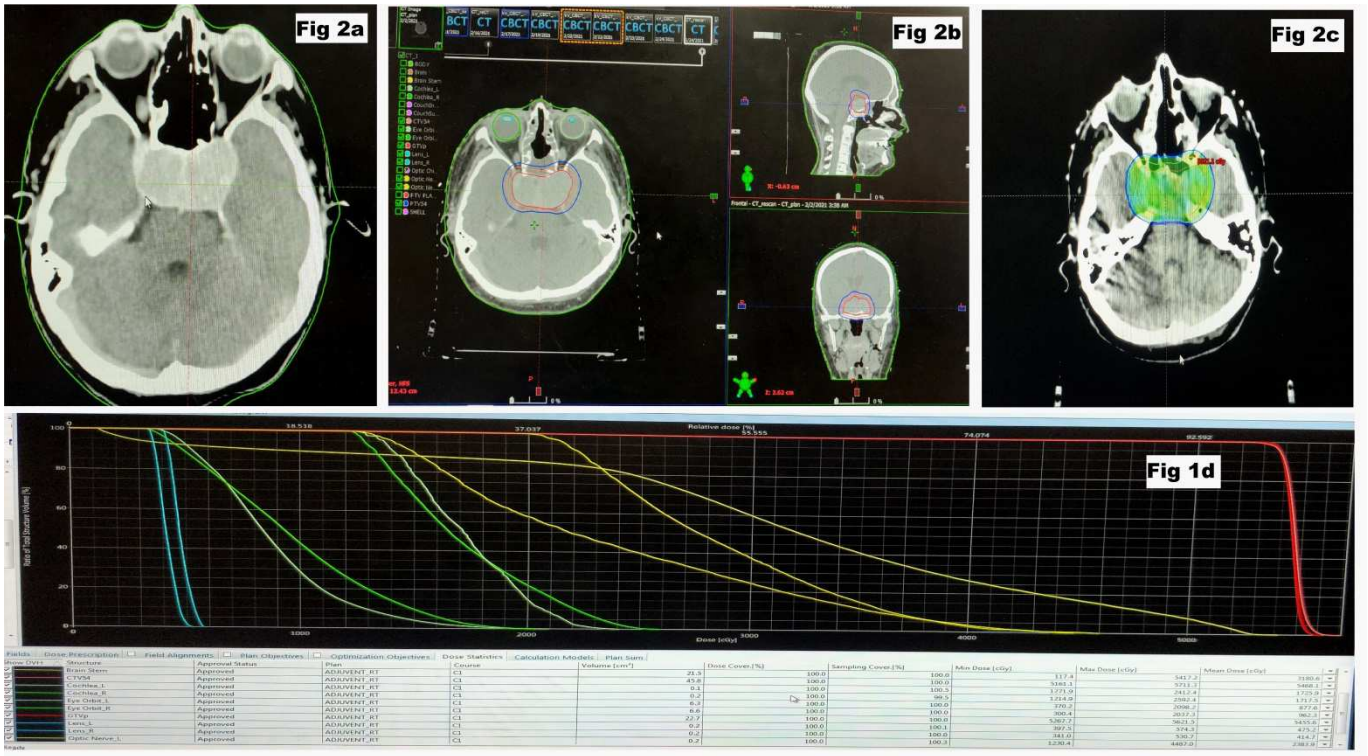

Figure 2. Figure 2a. CECT brain showing HPC. Figure 2b. Contouring HPC with GTV(red), CTV(pink), PTV(blue). Figure 2c. Color wash showing 95% coverage of PTV by target dose and no hot spot created. Figure 2d. Dose Volume Histogram (DVH) showing OAR and PTV coverage. **Note:** GTV- Gross Tumor Volume, CTV- Clinical Target Volume, PTV- Planning Target Volume

3. Discussion

HPCs are rare and aggressive tumors. Among all reported cases, HPCs are commonly seen in middle-aged individuals [7]. HPCs are more frequent in lower limbs, meninges, lungs, retroperitoneum, pelvis, but rare in bones, visceral organs of abdomino-pelvis [8]. Till now, only 13 cases HPC in pituitary fossa are reported as per published reports [9]. Patients with HPC are usually asymptomatic due to their slow growth. This necessitates proper imaging modalities and pathological examination for early diagnosis [10]. Radiologically, HPCs are homogenous, lobulated masses with irregular margins, in some cases CECT shows well-circumscribed mass with tissue necrosis or calcification. However, as CT or MRI cannot provide proper diagnosis, the confirmation of diagnosis largely rely on histopathology and immunohistochemistry report [11].

The standard treatment for HPC is surgery. Surgery improves overall survival (OS) and cancer specific survival (CSS) in HPC, but post-operative tumor recurrence rate is more than 30% [8], most commonly seen in HPCs of retroperitoneum and pelvis.

Radiotherapy is an alternative to surgery, especially when gross total resection is not achievable or patient is inoperable, like in our case. Radiotherapy decreases recurrence rate and improves survival [12]. Radiotherapy is indictaed in large tumors, particularly of >5cm or close/positive margin [13, 14]. Guthrie et al [15] reported that adjuvant radiotherapy decreases recurrence rate from 86% to 52%. Also, the relapse free survival was 75 months in those receiving adjuvant radiotherapy compared to 34 months in those who did not [15, 16]. Post-operative radiotherapy improves local control (LC) irrespective of the type of surgery, i.e, gross total resection or sub-total resection. Recommended dose is 50-60Gy. Dose more than 60 Gy improves LC [16].

However, in our case, the tumor resides in pituitary fossa. There is close proximity of organ at risk (OAR) like optic chiasm which has a dose constraint of 54Gy. So, dose of >54Gy would have compromised optic chiasm. Chemotherapy could be effective in metastatic HPC [17, 18]. Anthracyclines and Ifosfamide are clinically effective in metastatic HPCs [19, 20].

4. Conclusion

As HPC of pituitary fossa is very rare and it requires long term treatment, it is quite difficult for any institution to have a large series of cases to study regarding the epidemiology, behaviour and efficacy of treatment of HPC. More studies are required to study the epidemiology, basic molecular biology and various treatment modalities for HPCs.

References

[1] Soyuer S, Chang EL, Selek U, McCutcheon IE, Maor MH. Intracranial meningeal hemangiopericytoma: the role of radiotherapy: report of 29 cases and review of the literature. Cancer. 2004; 100: 1491-1497.

[2] Kewei Wang, Fei Mei, Sisi Wu, et al. Hemangiopericytoma: Incidence, Treatment, and Prognosis Analysis Based on SEER Database BioMed Research International / 2020 / Article ID 2468320 | https://doi.org/10.1155/2020/2468320

[3] Louis D, Ohgaki H, Wiestler O, Cavanee W. World Health Organization (WHO) classification of tumours of the central nervous system. Vol. 1. Revised. 4th ed. Geneva, Switzerland: WHO press; 2016.

[4] McKeever PE. Immunohistology of the nervous system. In: Dabbs D, ed. Diagnostic Immunohistochemistry. 2nd ed. Philadelphia: Churchill Livingstone; 2006: 746-816.

[5] E. Tani, J. Wejde, K. Astrom, I. L. Wingmo, O. Larsson, and F. Haglund, "FNA cytology of solitary fibrous tumors and the diagnostic value of STAT6 immunocytochemistry," Cancer Cytopathology, vol. 126, no. 1, pp. 36–43, 2018.

[6] Hanau CA, Miettinen M. Solitary fibrous tumor: histological and immunohistochemical spectrum of benign and malignant variants presenting at different sites. Hum Pathol. 1995; 26:440–449. [PubMed] [Google Scholar]

[7] S. Ohba, K. Murayama, Y. Nishiyama et al., "Clinical and radiographic features for differentiating solitary fibrous tumor/hemangiopericytoma from meningioma," World Neurosurgery, vol. 130, pp. e383–e392, 2019. View at: Publisher Site | Google Scholar

[8] M. Koch, G. P. Nielsen, and S. S. Yoon, "Malignant tumors of blood vessels: angiosarcomas, hemangioendotheliomas, and hemangiopericytomas," Journal of Surgical Oncology, vol. 97, no. 4, pp. 321–329, 2008.

[9] Matthias E Ernst, Aimée Hiller, Regina Reimann, Carlo Serra et al, "Hemangiopericytoma mimicking a pituitary adenoma: a case report" *Endocrine Abstracts* (2019) 63 P 738 | DOI: 10.1530/endoabs. 63. P 738.

[10] J. S. Brooks and S. Lee, "Contemporary diagnostics: sarcoma pathology update," Journal of Surgical Oncology, vol. 111, no. 5, pp. 513–519, 2015.

[11] M. Krengli, T. Cena, T. Zilli et al., "Radiotherapy in the treatment of extracranial hemangiopericytoma/solitary fibrous tumor: study from the Rare Cancer Network," Radiotherapy and Oncology, vol. 144, pp. 114–120, 2020.

[12] Ecker RD, Marsh WR, Pollock BE, Kurtkaya-Yapicier O, McClelland R, Scheithauer BW, et al. Hemangiopericytoma in the central nervous system: treatment, pathological features, and long-term follow up in 38 patients. J Neurosurg. 2003; 98: 1182–1187. [PubMed] [Google Scholar]

[13] S. H. Jeon, S. H. Park, J. W. Kim, C. K. Park, S. H. Paek, and I. H. Kim, "Efficacy of adjuvant radiotherapy in the intracranial hemangiopericytoma," Journal of Neuro-Oncology, vol. 137, no. 3, pp. 567–573, 2018.

[14] A. M. Stessin, C. Sison, J. Nieto, M. Raifu, and B. Li, "The role of postoperative radiation therapy in the treatment of meningeal hemangiopericytoma-experience from the SEER database," International Journal of Radiation Oncology • Biology • Physics, vol. 85, no. 3, pp. 784–790, 2013.

[15] Guthrie BL, Ebersold MJ, Scheithauer BW, Shaw EG. Meningeal hemangiopericytoma: histopathological features, treatment, and long-term follow-up of 44 cases. Neurosurgery. 1989; 25: 514–522. [PubMed] [Google Scholar]

[16] Ghia AJ, Chang EL, Allen PK, Mahajan A, Penas-Prado M et al. "Intracranial Hemangiopericytoma: Patterns of Failure and the Role of Radiation Therapy". Neurosurgery. 2013 Oct; 73 (4): 624-30; discussion 630-1.

[17] T. Akman, A. Alacacioglu, D. Dolek et al., "Malign recurrence of primary chest wall hemangiopericytoma in the lung after four years: a case report and review of the literature," Case Reports in Oncological Medicine, vol. 2014, Article ID 470268, 4 pages, 2014.

[18] S. J. Lee, S. T. Kim, S. H. Park et al., "Successful use of pazopanib for treatment of refractory metastatic hemangiopericytoma," Clinical Sarcoma Research, vol. 4, no. 1, p. 13, 2014.

[19] M. Delgado, E. Perez-Ruiz, J. Alcalde, D. Perez, R. Villatoro, and A. Rueda, "Anti-angiogenic treatment (sunitinib) for disseminated malignant haemangiopericytoma: a case study and review of the literature," Case Reports in Oncology, vol. 4, no. 1, pp. 55–59, 2011.

[20] S. Stacchiotti, N. Simeone, S. Lo Vullo et al., "Activity of axitinib in progressive advanced solitary fibrous tumour: results from an exploratory, investigator-driven phase 2 clinical study," European Journal of Cancer, vol. 106, pp. 225–233, 2019.