Long and Meaningful Survival of a Case with Disseminated Hemangiopericytoma - Treated with Radiation Alone

Bhargavi Ilangovan*, Janos Stumpf, Rathna Devi R, Subathira Balasundaramanjay
Department of Radiotherapy, Apollo Cancer Institute, Chennai, India


© Author(s), 2024, Publisher and License: THB. Open Access. This article is distributed under the terms of the Creative Commons Attribution 4.0 International License, which permits unrestricted use, distribution, and non-commercial reproduction in any medium, provided you give appropriate credit to the original author(s) and the source.

*Corresponding Author: Bhargavi Ilangovan, Department of Radiotherapy, Apollo Cancer Institute, Chennai, India. E-mail: bhargavi.ilangovan@gmail.com

Keywords: Hemangiopericytomas; Zimmermann; Radiosurgery

Abstract
Hemangiopericytomas are tumours arising from the pericytes of Zimmerman. They arise in the central nervous system and have a locally recurrent behaviour with occasional distant metastases too. This case highlights the careful and judicious use of different radiotherapy techniques used in tandem to ensure a long survival for a patient. Emphasis is made on the quality of life as he was able to function to his maximum till the end.

Introduction
Hemangiopericytomas (HPCs) are rare tumours of the central nervous system. They arise from the pericytes of Zimmermann [1]. They are notorious for their local recurrence and distant metastases. Known for their recurrent nature, radiotherapy has been indicated in the post-operative setting and at times of recurrence. To date, no systemic therapy has been found to be effective in hemangiopericytomas, so the patients are usually given the option of systemic therapy in the metastatic setting. Given their limited efficiency, the outlook of such patients is very dismal. We are discussing a patient who has been treated with radiotherapy for over 15 years. Radiotherapy has been used for him in the post-operative, recurrent, and metastatic setting. He has a good quality of life, though nearly 20 locations needed radiotherapy. Impressive progress in the precision delivery of radiation is an important factor in his treatment.
Case presentation

The patient was initially evaluated in 2004 and was diagnosed with posterior fossa hemangiopericytoma. He was 38 years old then, he underwent surgery in 2004. It recurred in 2006 and was operated again. He was given post-operative radiotherapy of 60gy/30# by IMRT from 9/1/2007 to 21/2/2007. In May 2012, he was found to have a recurrent occipital lesion for which he underwent fractionated stereotactic radiotherapy with Cyberknife. A total dose of 35gy/5# was delivered from 23/5/2012 to 28/5/2012 [Figure 1].

In April 2013, MRI brain showed an enhancing mass in the right inferior cerebellopontine angle cistern eroding clivus and right lateral mass of C1 vertebrae. The occipital lesion had responded well to radiotherapy. PET CT showed multiple lung lesions. The biopsy from the left lung mass showed features of hemangiopericytoma. Surgery was deferred due to the multiple oligometastatic status (less than 5 in a single organ). He was given 30Gy/5# to the brain lesion and 24gy/3# to the lung lesion from 24/8/13 to 27/8/13 as it was oligometastatic [Figure 2].

Figure 1: The target and the dose distribution
In December 2014, he was evaluated with complaints of speech disturbances for a week. PET CT (31/12/2014) revealed a stable lesion in the left occipital region and the inferior margin of the clivus. Nevertheless, he had developed another lesion in the right Petro occipital fissure cerebellopontine angle cistern, eroding the clivus and the jugular foramen. CT chest (18/12/2014) showed an increase in the size of pulmonary nodules. At this point, the options of systemic therapy were discussed with the patient. He was not keen on it as he had concerns about the side effects and quality of life. So far, he has been able to manage his day-to-day affairs with minimal assistance. After discussing the risks associated with repeated radiotherapy in the brain, he was treated with 30gy/5# to the lung and 32.5gy/5# to the brain lesion from 22/12/14 to 27/12/14 by Cyberknife radiosurgery.

In October 2016, PET CT (5/10/2016) showed a new hypermetabolic expansile lytic lesion in the third rib along with an interval increase in size and number of lung nodules scattered in both lungs. He was treated with 35Gy/5# to the lung lesion from 8/10/16 to 14/10/16.

In March 2018, he was evaluated for neck pain. MRI brain (20/3/2018) showed an increase in the size of the extra-axial mass in the occipital region at the level of confluence of venous sinuses infiltrating the C2 vertebrae and abuts vertebral artery. PET-CT (20/3/2018) showed a new mass involving the right mastoid, an increase in the size of the mass in the mid-occipital region, right third rib, and pulmonary metastases. Hence, he was given 35gy/10# to the recurrent brain lesion, 32.5gy/5# to the new right mastoid region, 24gy/3# to the anterior lung lesion, 35gy/10# to the left lung lesion near the pericardium, and 40gy/10# to the right rib [Figure 3].

Figure 2: The Stereotactic radiosurgery plan for the lung metastases
In February 2019, he was evaluated with complaints of neck pain and restricted movement of limbs. PET-CT (18/2/2019) showed a new sub-centimetric lesion in segment IV of the liver, and a dural deposit in the spinal canal extending from C3-C6 vertebrae levels with spinal cord compression. He was treated with 30Gy/10# to the spinal lesion and 21Gy/3# to the right lesion by Cyberknife radiosurgery from 4/4/2019 to 17/4/2019 [Figure 4].

He was still alive and well until our last follow-up in 2020 and had been put on steroids by his local physician for unsteady gait.
Discussion

Hemangiopericytomas are tumours that arise from the pericytes of Zimmerman. They were first described by Stout and Murray [1]. They are classified as non-meningothelial mesenchymal tumours. They are graded between 1 and 3 based on their soft tissue pattern [2]. They are characterised by local recurrences and late distant metastases. Complete surgical excision with or without postoperative radiotherapy has been the standard of care. Complete excision is often not possible due to their location close to critical structures [3]. Radiotherapy and radiosurgery are known to arrest the growth of the tumour. Considering the aggressive nature of the disease, arrest of further growth itself might be a reasonable goal.

True to the nature of the disease, our patient recurred locally within 2 years of the first surgery and received radiotherapy in the post-operative setting. When it recurred, he was given fractionated stereotactic radiotherapy (FSRT). Radio surgery is an option for treating recurrent, unresectable, or metastatic tumours [4]. They aim at delivering a large dose of radiation per fraction, with significant sparing of adjacent normal structures. The possibility of repeat irradiation is possible due to this feature, as in our case. Some have reported response rates as high as 90% with radiosurgery [5]. In our patient, there was a reduction in the size of the occipital lesion with radiosurgery.

When he developed lung metastases 9 years after his first diagnosis, the options of chemotherapy, surgery, and radiotherapy were discussed in detail. Due to the scant literature available on the role of chemotherapy, it was declined by the patient. Since the disease was still oligometastatic, further local treatment was offered. In our case, surgery for the lung lesion when there was progression in the primary site was not appropriate.

Three years later, when he came back with multiple lung lesions, chemotherapy was contemplated. The patient was still apprehensive about chemotherapy. Systemic chemotherapy has not been found to be effective in these tumours. Some combination regimens have been shown to produce a partial response, despite the toxicity. Recently Pazopanib has been used in metastatic Hemangiopericytomas, with arrest of tumour growth [6]. With the not-so-promising results of chemotherapy, and not to mention the added toxicity, we decided to explore radiation again. He responded well and had a disease-free interval of 2 years this time.

After the initial treatment, our patient has survival for 17 years. He was treated for recurrent and metastatic disease with radiation for 15 years. There have been very few cases reported to have such a prolonged survival after metastases [7]. Due to the precise radiation, he did not have any adverse radiation-related side effects. Multiple locations needed irradiation and there was an overlap of doses in these locations.
Conclusion

Our patient has received FSRT for numerous lesions. Some of them disappeared and some became stable. New lesions were observed after an interval of 1-2 years. We considered reporting this case to highlight the fact that radiotherapy is an effective tool for these tumours, even in the metastatic setting. The decision to not use systemic therapy was a combined decision of both the patient and the treating team. He was able to understand the outcome and was keen on his quality of life. This might not be possible at all, but in those with access to stereotactic irradiation, it must be thought of.

In general, the role of radiotherapy and radiosurgery should be considered with the present quality of precision radiation oncology. With better imaging, we can localise the target with precise delivery, while the sparing of normal tissues is ensured. This has facilitated aggressive management of this cancer with repeated radiation sessions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of Interest: Nil

Financial Disclosure: None

References

