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(CASE REPORT)

Radiotherapy of epithelioid hemangioendothelioma of the posterior fossa

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Abstract

Epithelioid hemangioendothelioma is a widespread vascular cancer that is infrequently detected within the skull. Frequently seen in young individuals and typically treated with surgery at first. Interferon and radiotherapy are commonly used as treatments, especially during the adjuvant phase. Prognostic factors are decided by the initial treatment and the type of histology. We present a case of a patient who received adjuvant radiotherapy following a partial resection in our department. After 4 months of monitoring, we saw a significant improvement in the neurological symptoms, however the right central facial paralysis persisted primarily because the patient did not consistently attend motor physiotherapy sessions for facial re-education.

Keywords: Epithelioid hemangioendothelioma; Brain; Neurosurgery; Radiotherapy; Senegal.

1. Introduction

Hemangioendothelioma is a type of tumor found in soft tissues that has features that are in between those of hemangioma and angiosarcoma, both histologically and clinically. It could show the characteristics of a hemangioma, as well as showing a significant level of cellularity and mitotic activity [1]. Epithelioid hemangioendothelioma (EHE) is the most often seen among the four varieties of hemangioendothelioma [2]. Usually originating from a medium or large vein, a small soft-tissue tumor is commonly found in the subcutaneous tissue or muscle [3]. It happens in different organs like the lungs, liver, and bones. Intracranial EHE, a condition that can affect either the brain parenchyma or its coverings, is uncommon. Kepes et al [4] described the first intracranial lesion. We present a patient who had adjuvant radiotherapy for EHE in the posterior fossa.

2. Observation

The patient was 34 years old, with no specific pathological history, and had been treated for severe headaches associated with vertigo and jet vomiting. The initial clinical examination revealed a conscious, well-oriented, right-handed patient with right central facial paralysis, with no sensory disturbance and no cerebellar or pyramidal syndrome. Brain magnetic resonance imaging (MRI) revealed an intensely enhanced, heterogeneous right lesion process cerebello-vermian sub tentorial lesion compressing V4, responsible for active upstream hydrocephalus, associated with a cervical medullary centrifugal lesion.

No secondary lesions were found on extension. A ventriculocisternostomy was performed, followed by partial removal of the tumor. A post-operative CT scan revealed an oedemato-haemorrhagic right cerebellopontine remodeling, responsible for a 7.5 mm tonsillar involvement with tetra ventricular hydrocephalus with no signs of resorption.

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A follow-up MRI showed satisfactory resorption of the hydrocephalus despite persistent infracentimetric nodular contrast of the right edge of the cerebellar tent, remodeling of the right cerebellar hemisphere with heterogeneous filling of the

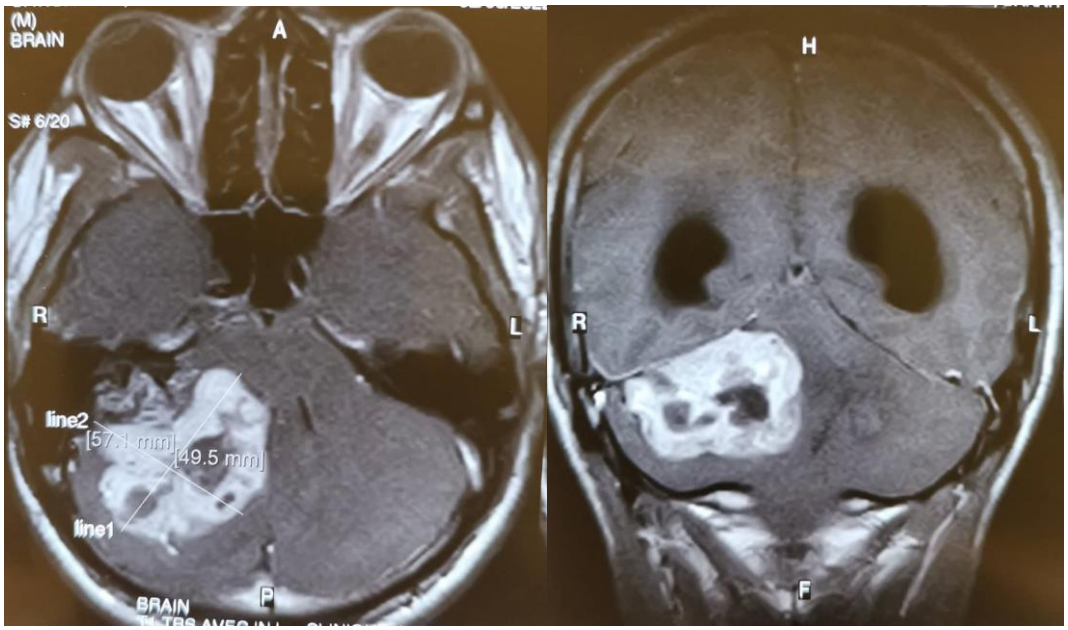

Figure 1 Axial and coronal sections of pre-operative brain MRI

Middle ear next to the acoustic-facial bundle and mastoid cells. The pathological report of the surgical specimen showed an epithelioid hemangioendothelioma. Following a multidisciplinary consultation meeting, the decision was taken to treat the patient with adjuvant radiotherapy.

The patient underwent radiotherapy over a period of 06 weeks. The technique used was 3D conformal without intensity modulation. He received 60 Gy to the residual tumor in 30 fractions of 2 Gy. The clinical anatomical target volume (CTV), receiving 50 Gy, included the residual tumor considered as the gross target volume (GTV) with isotropic margins of 20 mm, respecting anatomical barriers, particularly bone, and including the surgical approach. The planning target volume (PTV) was delineated with 10 mm margins around the CTV. Figure 2 shows the isodose curves and the ballistics used.

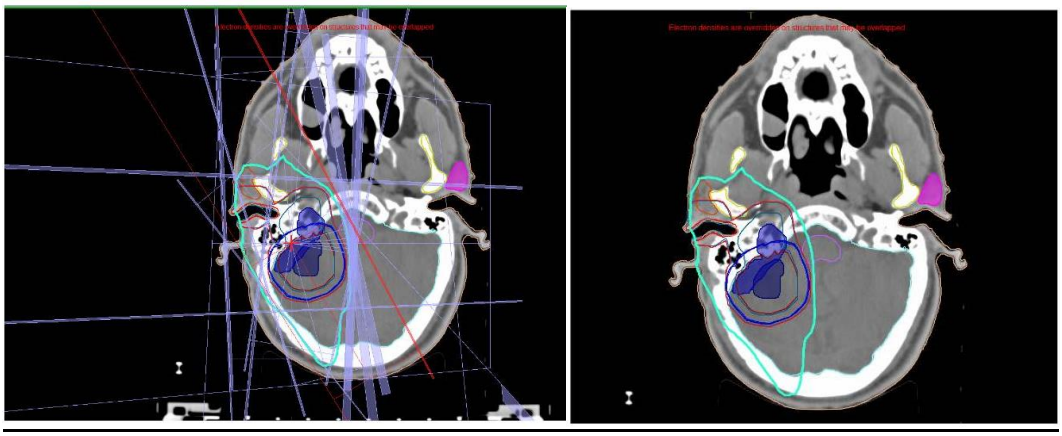

Figure 2 Ballistics and isodoses curves

Prophylactic corticosteroid therapy was administered during radiotherapy. Toxicity was marked by radiodermatitis of the right parieto-temporo-mandibular region and hemiface, radiomucositis of the oral cavity and dry eyes, all grade I according to CTCAE version 4.0.

One month (M1) after the end of radiotherapy, the patient noted a few episodes of simple vomiting but a clear regression of the initial headaches and radiomucositis. On clinical examination, the right central facial paralysis persisted. Motor physiotherapy was prescribed. The patient was seen again in consultation 4 months after the end of his irradiation. The clinical examination was like that conducted at M1, as the patient had not followed his physiotherapy sessions for facial rehabilitation.

3. Discussion

Epithelioid hemangioendothelioma (EHE) is a rare tumor making up 1% of all vascular tumors [5]. It is uncommonly found within the brain, typically in the skull base, dura mater, or brain tissue, making up less than 0.02% of all brain tumors [6]. EHE can be identified at any age, but it is typically diagnosed between the ages of 20 and 30, with no connection to gender [7].

According to a systematic review by Barger et al [8], the occurrence of EHE in the posterior fossa seems to be exceedingly rare. This study detailed 47 instances of intracranial EHE, with 37 occurring in adults, none of which were in the cerebellum. There were just 6 instances of EHE located in the posterior cranial fossa, not affecting the cerebellum.

Just like in the case of our patient, four individuals received more radiotherapy, with one of them experiencing a recurrence. One case showed stable tumor response for a period of 8 years [9].

Various approaches have been suggested for treating EHE, typically involving surgery followed by radiation therapy and possibly interferon therapy according to some sources. Nevertheless, it is unclear whether pre- and post-operative radiation therapy provide advantages for EHE when complete removal is possible. Yet, the close connection to crucial components and/or their irregular blood supply could hinder total removal.

Radiotherapy [13-15] is suggested for lesions that have been partially removed. In most cases reported, conventional radiotherapy was the main technique used, along with its side effects of toxicity.

The outlook for individuals with intracranial EHE is unknown. Recurrence is uncommon; Weiss and Enzinger [3] found that only 13% of patients experienced a recurrence of the disease. Even after complete removal, three intracranial EHE reappeared: two quickly and one after six years [16]. The quick return could have been caused by not removing everything. The outlook for EHE is still unclear, even for patients with seemingly typical locations [17]. While the general agreement is positive, prognostic information is more uncertain for intracranial locations.

4. Conclusion

EHE of the posterior fossa is a rare pathology in terms of histology and exceptional in terms of location. To our knowledge, this is the first publication on this location in Senegal. Surgery is still the mainstay of management, but adjuvant radiotherapy is still common, especially in cases of partial resection. The prognosis of these patients depends on the first management and the anatomopathological aspects of the tumor.

Compliance with ethical standards

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Disclosure of Conflict of interest

The authors have declared no conflicts of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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