

## Case report

### Sellar recurrent hemangiopericytoma without metastasis: a case report

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#### Abstract

Primitive hemangiopericytomas of the central nervous system are rare and represent less than 1% of intracranial tumors. Their nonspecific clinical and imaging can be misleading and bring to a mistaken diagnosis of meningioma. Radical surgery is the treatment of choice, but must be supplemented with postoperative radiotherapy. Hemangiopericytomas have a high potential for recurrence and metastasis. Sellar location is very rare and can simulate a pituitary adenoma. We report a case of a 24 year-old woman diagnosed with sellar recurrent HPC without metastasis.

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## Introduction

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Hemangiopericytoma (HPC) is a potentially malignant tumor originating from Zimmermann's pericytes around capillaries and postcapillary venules [1]. HPCs are located most often, in the musculoskeletal system and the skin [2]. Intracranial HPCs are uncommon tumors, accounting for less than 1% of primary central nervous system (CNS) tumors, and constitute 2% to 4% of meningeal tumors [3]. HPC occurred generally on supratentorial with most frequency in parasagittal and falx region mimicking meningioma [1]. These tumors have a high potential of recurrence and metastasis [4]. We report a case of a recurrent sellar HPC without metastasis.

## Patient and observation

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A 24-year-old woman was admitted with a 1-month history of headache, and bilateral visual loss. Physical examination found no neurological deficit, her visual field was 1/10, and bitemporal hemianopsia. The other cranial nerves were normal. She had no medical past history of hypertension, diabetes mellitus or pulmonary tuberculosis. Brain CT scan showed a sellar and supra sellar lesion intensely and homogeneously enhanced by contrast-agent. Brain magnetic resonance images (MRI) revealed an intrasellar and suprasellar mass with the compression of the chiasm. The lesion was isosignal on T1-weighted sequence and slightly hyperintense on T2-weighted sequence, with an intense and homogenous enhancement (Figure 1 (A)). Endocrinological studies showed hypopituitarism. The possible diagnosis included pituitary adenoma and meningioma. Patient underwent surgery using transsphenoidal approach. The tumor appears highly vascularized accommodated a subtotal resection (Figure 1 (B)). Histological diagnosis was hemangiopericytoma (Figure 2). She received a local radiotherapy (52 Gy) of the pituitary fossa. Two years later, she complained of headaches and visual loss. Brain MRI showed a contrast-enhancing soft tissue mass arising from the pituitary fossa (Figure 3). Partial resection of the tumor was done and pathologic findings were compatible with hemangiopericytoma. Thoracoabdominal CT scan was normal. The patient received a second radiotherapy with favorable outcome after one year follow-up, without recurrence of the tumor (Figure 4).

## Discussion

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Intracranial HPCs are uncommon tumors, accounting for less than 1% of primary CNS tumors, and constitute 2-4% of meningeal tumors [3]. They were previously classified as angioblastic sub-type meningiomas [5]. Since 1993, HPCs are classified in the group of "mesenchymal, non-meningothelial tumors" [6]. These tumors arise from smooth muscle perivascular pericytes of Zimmerman. They are more aggressive than meningiomas, with high frequency of recurrence and metastasis [4]. HPCs are commonly located in supratentorial region (parasagittal or falx) [1]. Sellar location is very rare. Few cases have been reported in the literature [1,4,7]. It occurs on male patients in 55% to 77% from 38 to 42 years [1]. In our case the patient was a young woman of 24 years old.

Brain imaging (CT and MRI) are important but couldn't make a difference with meningioma [7-9]. However, some elements could help to make a difference between HPC and meningioma. Intratumoral calcification or hyperostosis reaction in contact with the insertion zone are never viewed in HPCs; bone destruction

without piping osteosclerosis is a classic look, but nonspecific. The edges are often irregular and polylobed. Contrast enhancement after injection is often intense and heterogeneous [7,8]. In sellar locations, diagnostic of pituitary macroadenoma is most often carried, and transsphenoidal approach is proposed. The treatment of primitive HPCs central nervous system based on complete resection of the tumor, followed by radiation in order to prevent recurrence and metastasis [8-10]. Immunohistochemistry can differentiate HPC from meningioma: HPC stains positive for CD 34, factor VIII, and vimentin but negative for S-100 and epithelial membrane antigen (EMA) [3,11].

HPCs can recur locally or distantly in the neural axis or metastasize to extraneural sites [12]. Schiariti reported 28 cases of local recurrence, in a series of 39 patients. The recurrence rate at 1, 5, and 15 years was 3.5%, 46%, and 92%, respectively. Extraneural metastasis occurred in 8 patients (26%) at an average of 123 months after initial surgery [12]. In our case the recurrence appears after two years. According to Myung-Hwan [1], it is after 9 years. Recurrence is a late event in the natural history of HPC. Close follow-up for a long period after the first surgery is needed. Complete excision favorably affects recurrence and survival, as opposed to incomplete excision [13]. Because the hypervascular characteristics of HPC simply "radiosensitivity", postoperative adjuvant radiotherapy appears to significantly decrease the local recurrence rate and lengthen recurrence-free duration. However recurrence remains a common treatment outcome regardless of initial strategy [13]. Stereotactic radiosurgery is benefic in case of recurrent HPCs [14]. Extended analysis using lesion data available revealed that higher marginal dose ( $\geq 17$  Gy) was also significant ( $P = 0.028$ ). Kim and al., reported that Gamma Knife Stereotactic Radiosurgery provides an effective and safe adjuvant management option for patients with recurrent or residual HPCs [14]. The local tumor recurrence in our case could be assigned to incomplete excision at the first operation.

## Conclusion

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Sellar and suprasellar HPC can mimic a pituitary tumor. This case illustrates a difficulty to establish diagnostic on imaging and clinical exams. Diagnosis is made by histological studies. Unfortunately HPC has usually poor prognosis. Although rare in sellar location, HCP should be considered in differential diagnosis of pituitary tumors.

## Competing interests

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The authors declare no competing interests.

## Authors' contributions

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Doleagbenou Agbeko Komlan: substantial contributions to conception and design, acquisition of data, drafting the article and revising it critically for important intellectual content; Final approval of the version to be published. Habibou Lamine: substantial contributions to conception and design, acquisition of data, final approval of the version to be published. Djoubairou Ben Ousman : drafting the article and revising it critically for important intellectual content; final approval of the version to be published. Gana Rachid: final approval of the version to be published. Bellakhdar Fouad: final approval of the version to be published. All authors read and agreed

to the final version of this manuscript and equally contributed to its content and to the management of the case.

## Figures

**Figure 1:** (A): Preoperative magnetic resonance image showing a sellar and suprasellar solid mass; (B): postoperative magnetic resonance image showing a remnant

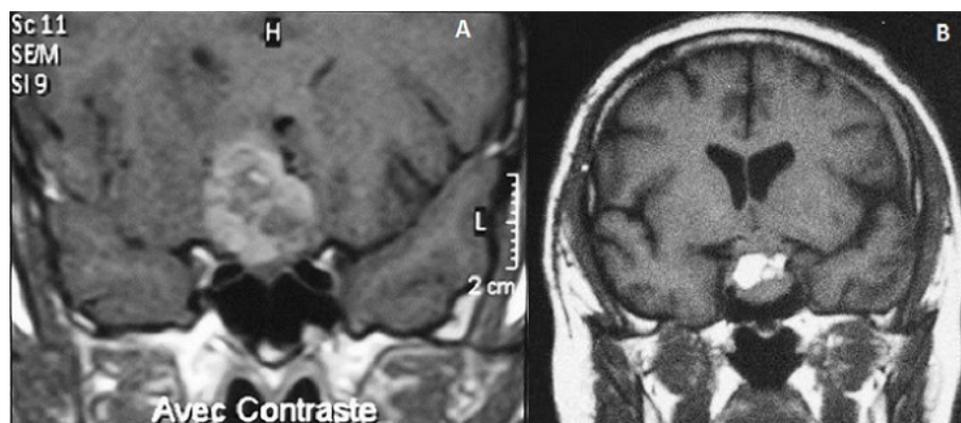
**Figure 2:** Histology of tumor, immunohistochemical staining revealing positive reaction for CD 34

**Figure 3:** Recurrent tumor arising from the pituitary fossa

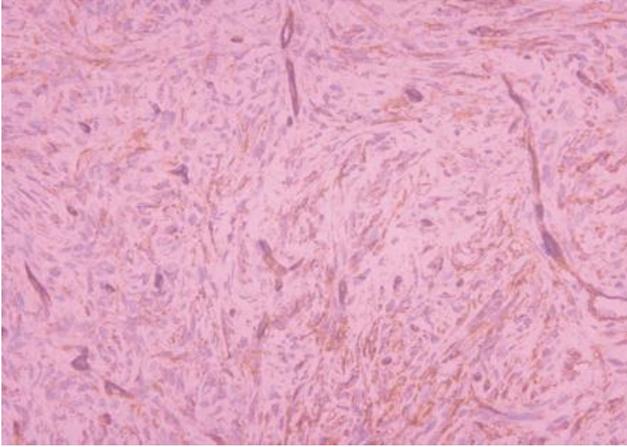
**Figure 4:** MRI one year after second surgery and radiotherapy

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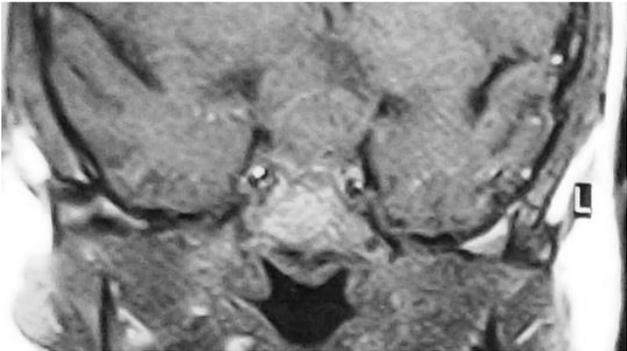
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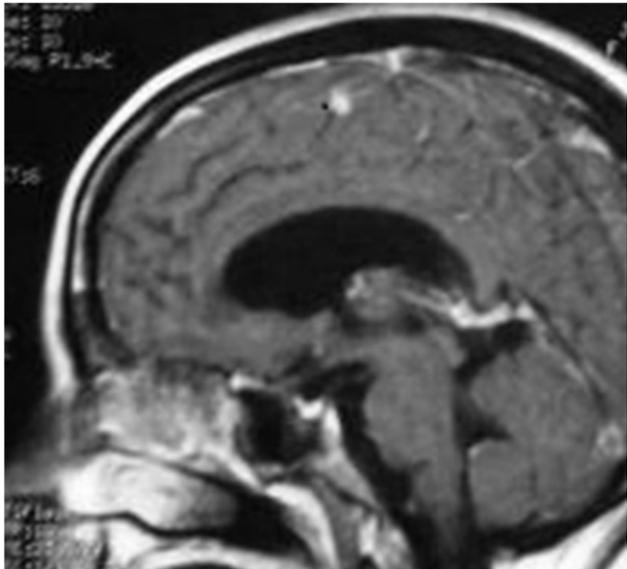
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**Figure 3:** Recurrent tumor arising from the pituitary fossa



**Figure 4:** MRI one year after second surgery and radiotherapy