# Central Nervous System Hemangiopericytomas/Solitary Fibrous Tumors: a case series.

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## Introduction

We describe treatment strategies and outcomes of 4 patients diagnosed with hemangiopericytoma (HPC), an uncommon mesenchymal tumour exhibiting high local recurrence and distant metastasis rates (1,2).

## Methods

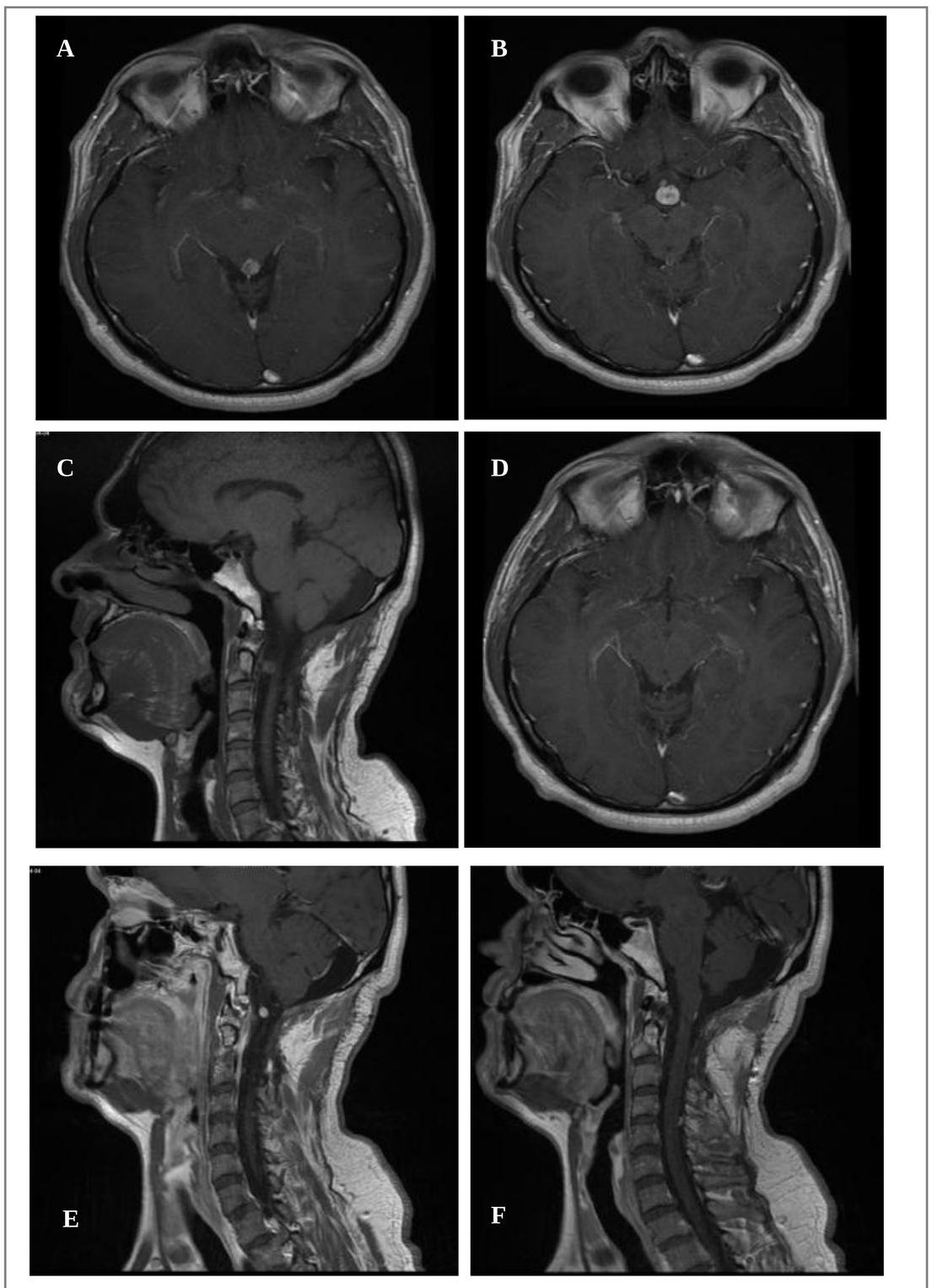
We retrospectively collected four cases of HPCs followed up from 2012 to May 2017. We assessed patients'age at diagnosis, symptoms at onset, tumor characteristics on brain or spinal MRI, and histopathological data. Moreover, we collected data concerning treatment of primary tumor and recurrencies.

## Results

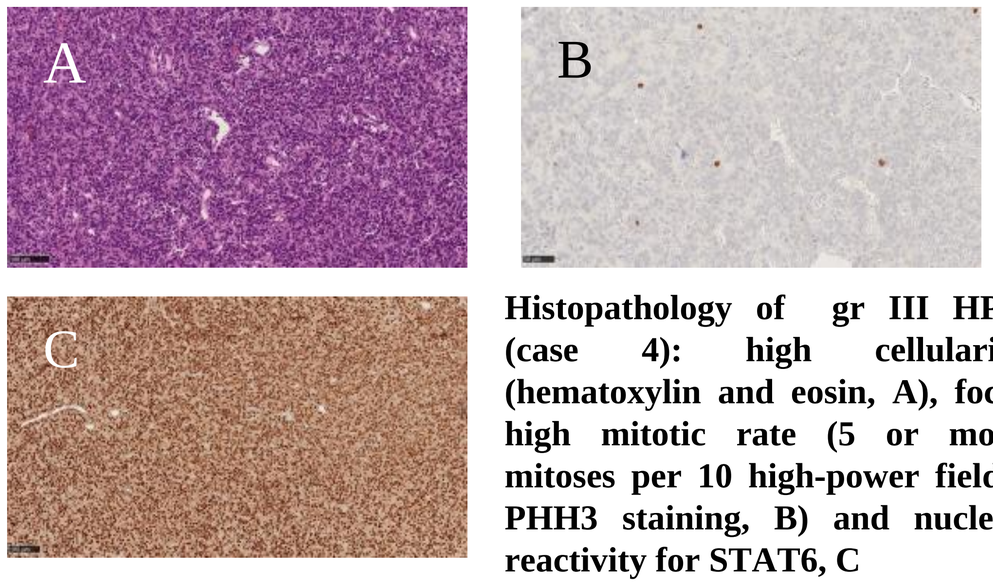
Mean age at diagnosis was 41,25 (SD 15,2); symptoms at presentation included seizures, aphasia, headache and cervical pain. In one case a subgaleal mass was recognized, due to HPC's local invasion. In three cases, MRI demonstrated a single T1 iso-hypointense nodular lesion, with marked contrast enhancement and T2 hyperintense appearance. In one case, tumor showed skull invasion and brain temporo-parietal infiltration; in the other two cases HPC was localized in the middle cranial fossa, with mass effect on right temporal lobe, or arised from cerebral falx, with right frontal lobe infiltration. The fourth case was misdiagnosed in 1999 (C2 spinal lesion pathologically diagnosed as malignal peripheral nerve sheat tumor) and in 2012 exhibited multiple spinal and cerebral nodular lesions, involving retrosellar area, middle brain, and cervical spinal cord. Some lesions showed necrotic areas and mass effect. PET-CT was performed in three cases for tumor staging. A gross total resection (GTR) was performed in patients with solitary HPC, a subtotal resection (STR) on C2 recurrent lesion of patient 4. Histological diagnosis was HPC grade II in three case, gr III in case 4. Adiuvant radiotherapy was prescribed in one case with GTR. In the fourth patient stereotactic radiotherapy was performed on cervical spinal recurrences and on cerebral HPCs in 2013. In 2017 a new cervical spinal progression was demonstrated and the patient was treated with surgery (STR) and adrotherapy; last systemic staging (PET-CT) showed a lung lesion suggestive of new disease localization. Patients with solitary HPC had no recurrences.

## Conclusions

Hemangiopericytomas can have an aggressive course and require a systemic staging given the possibility of distant metastases. HPC are challenging to treat despite multimodality therapies. Treatment options include primary resection, RT (namely stereotactic RT), embolization and chemotherapy, even if an effective chemotherapeutic regimen has not been reliably demonstrated (3). It is proved that adjuvant RT following STR prolongs Progression Free Survival (PFS) and Overall Survival (OS), while the role of RT following GTR is still debated (1). In our case series better outcome correlated with histology (gr II) and a radical surgical treatment.




**A, B, C: patient 4, 2012: multiple HPCs involving suprasellar cisterna, lamina quadrigemina, cervical spinal cord (C2) before C2 lesion STR and RT**
**D, E, F: patient 4, 2016: no recurrence of brain HPCs; evidence of new cervical HPCs**




**Histopathology of gr III HPC (case 4): high cellularity (hematoxylin and eosin, A), focal high mitotic rate (5 or more mitoses per 10 high-power fields; PHH3 staining, B) and nuclear reactivity for STAT6, C**

**Bibliography**

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