

# Intraspinal dissemination of an intracranial hemangiopericytoma in a child: A case report

<sup>1</sup>Burcin Agrıdag Ucpınar MD, <sup>1</sup>Cennet Sahin MD, <sup>2</sup>Canan Tanık MD

<sup>1</sup>Department of Radiology, <sup>2</sup>Department of Pathology, Sisli Hamidiye Etfal Training and Research Hospital, Istanbul, Turkey

## Abstract

Hemangiopericytoma (HPC) is a rare tumor with malignant potential, high local recurrence rate and risk of metastasis. The average age of presentation is 35 to 44 years and only 10% of the HPCs occur in children. Spinal metastasis of intracranial HPC is extremely rare that only two illustrated cases have been reported in the literature. A child who was diagnosed with WHO Grade III intracranial HPC which was complicated with intra-spinal metastasis on follow-up is presented. Intra-spinal dissemination of the HPC in child age group has not previously been reported. We emphasize with this report that intracranial HPC can cause intra-spinal drop metastasis in the child age group.

**Keywords:** hemangiopericytoma; intracranial hemangiopericytoma; CNS tumors; spinal mass.

## INTRODUCTION

The term hemangiopericytoma (HPC) was firstly named in 1942 by Stout and Murray to describe a rare tumor originated from Zimmerman pericytes of capillaries.<sup>1</sup> The pathologic classification of HPC is an area of ongoing discussion that; 2016 World Health Organization (WHO) classification included HPC in the “extrapleural HPC and solitary fibrous tumor” subgroup of the group of “fibroblastic/myofibroblastic tumors”. Furthermore; intracranial HPC is divided into two categories named as WHO grade II hemangiopericytoma and WHO Grade III anaplastic hemangiopericytoma.<sup>2</sup>

HPC may occur at any sites of the body where there are capillaries; 16-33% occur in the central nervous system (CNS), mainly near the meninges of the dura. In all CNS tumors; HPCs account for only 1.6%.<sup>3</sup> The average age of presentation is 35 to 44 years and only 10% of the HPCs occur in children. HPCs are rare tumors with malignant potential and high local recurrence rate and risk of metastasis; mainly to bone, lung, and liver.<sup>4</sup>

In this case report; we describe a child with WHO Grade III intracranial hemangiopericytoma presenting with intradural spinal metastasis on follow-up. To the best of our knowledge, intra-spinal dissemination of the HPC during childhood has not previously been reported in literature.

## CASE REPORT

A six-year-old boy was referred to our emergency department because of an acute onset of headache and nausea for one day. The parents reported that the patient had difficulty in walking since the symptoms began. Previous clinical history was unremarkable and the family reported no traumatic event. On ocular examination; mydriatic pupils and right-sided nystagmus were observed. On neurologic examination; the patient also had ataxia. Sensation could not be evaluated due to patient’s non-cooperation. On otoscopic examination; both tympanic membranes were intact and unremarkable. The patient had no fever, and blood tests were in normal values. An immediate non-contrast CT scan showed a 35x35 millimeter in diameter heterogeneous hyperdense mass lesion with perilesional extra axial hemorrhage located in the posterior cranial fossa next to the right cerebellar hemisphere. Fourth ventricle was also effaced without hydrocephalus. (Figure 1. A) On magnetic resonance imaging (MRI) of the brain and whole spine; intracranial lesion was in extra axial location surrounded with cerebrospinal fluid (CSF) and in close relationship with right sigmoid sinus. The lesion was T2 (Figure 1. B) and T1 (Figure 1. C) isointense in unenhanced scans. Following injection of Gd DTPA, heterogeneous enhancement was seen (Figure 1. D). Signal

Address correspondence to: Burcin Agrıdag Ucpınar, Department of Radiology, Sisli Hamidiye Etfal Training and Research Hospital, Etfal Hastanesi, Halaskargazi Cad., 34371 Şişli/Istanbul, Turkey. Email: drburcinagridag@gmail.com

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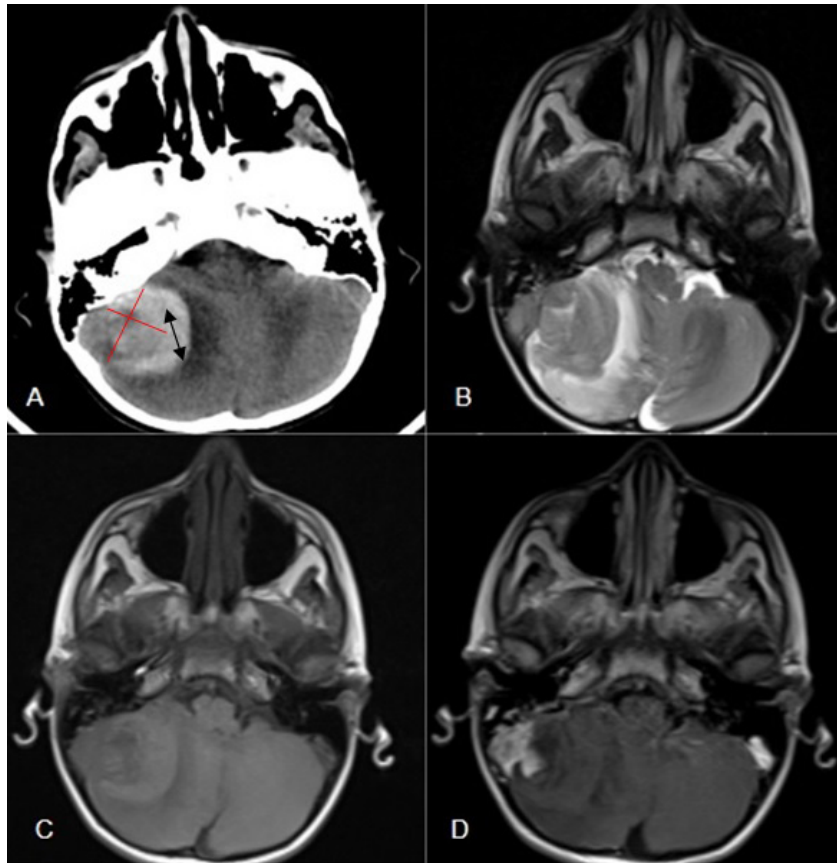


Figure 1. (A) Axial cranial CT showing hemorrhagic mass lesion in posterior cranial fossa close to right sigmoid sinus (red lines) with perilesional extra axial hemorrhage (black arrow) and significant effacement of the fourth ventricle due to compression. (B) Axial T2-weighted image and (C) T1-weighted image at the level of the cerebellum showing isointense extra axial mass lesion in close relation with right sigmoid sinus. (D) Axial T1-weighted image after Gadolinium contrast injection demonstrates heterogeneous enhancement. Contrast enhancement of the right sigmoid sinus is also noted.

void areas in the lesion and perilesional extra axial hemorrhage were also observed. On whole spinal MRI; no accompanying mass lesion or any remarkable pathology was observed (Figure 2. A, B). Accordingly; total excision of the mass and drainage of the perilesional hematoma were performed through a suboccipital craniectomy on the following day. The operation was conducted carefully by not cutting the tumor into pieces to prevent dissemination. Pathology specimens revealed malignant tumor with high cellular components composed of big rounded nucleus mesenchymal cells and containing vascular structures and spreading throughout the dura (Figure 3). The patient was diagnosed as malignant hemangiopericytoma WHO Grade III. Chemotherapy and local radiotherapy (50 Gy) were initiated to the patient. After the radiotherapy was completed, he was referred to a local physician

for follow-up serial brain MRI.

After 25 months of follow-up; patient was referred to the emergency department with symptoms of gait disturbance and urinary incontinence. On spinal MRI; intradural extramedullary 76x15 millimeter in diameter mass lesion was observed at the level of L4-S2 vertebral bodies (Figure 2. C, D). Biopsy was performed and pathology specimen revealed the spinal seeding of HPC. Hemilaminectomy with metastasectomy was performed due to acute onset spinal cord compression symptoms. Postoperative period was uneventful. However, although his acute spinal compression symptoms were relieved after the surgery, he did not recover fully from his gait disturbance. The patient was referred to radiation oncology for assessment of adjuvant fractionated proton beam therapy.

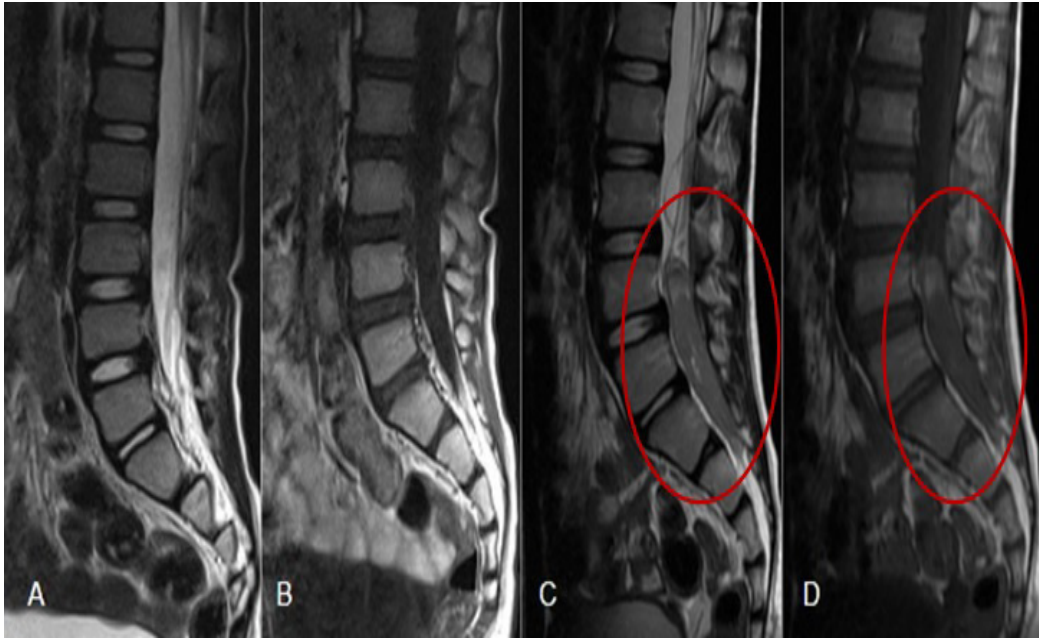


Figure 2. (A) Sagittal T2-weighted and (B) T1-weighted image at initial spinal magnetic resonance imaging show no abnormality. (C) Sagittal T2-weighted and (D) T1-weighted image 25 months after the initial diagnosis shows intradural spinal mass lesion which represents spinal drop metastasis of intracranial hemangiopericytoma (red circles).

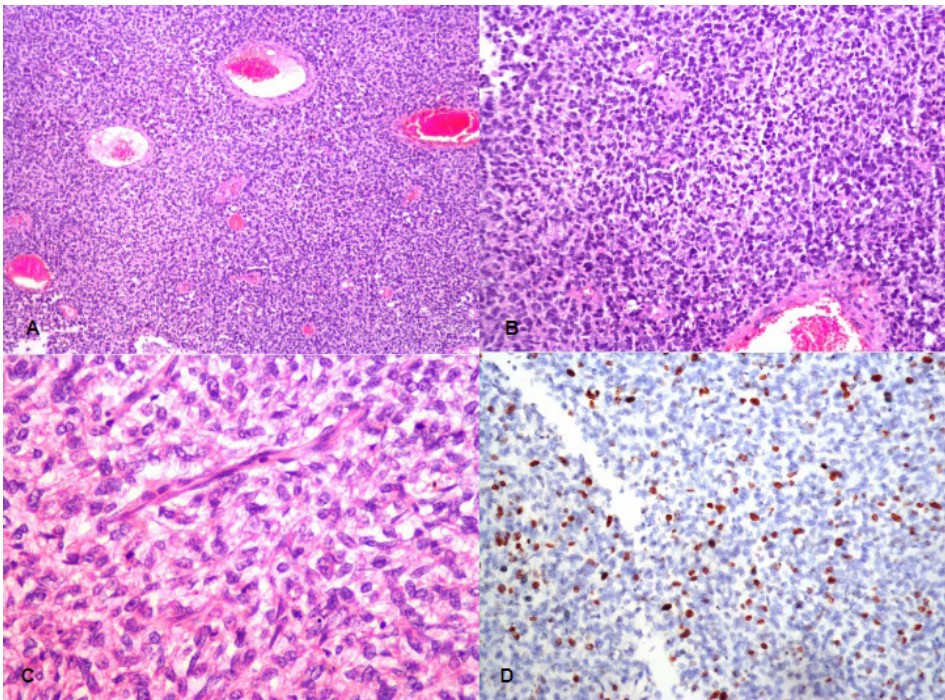


Figure 3. (A) Tumoral infiltration that consists of thin walled vascular structures and shows diffuse growing pattern: hematoxylin and eosin stain (H&E);  $\times 40$  magnification. (B) Tumor cells with spindle-shaped: hematoxylin and eosin stain (H&E);  $\times 100$  magnification. (C) Malignant tumoral infiltration with obvious mitotic activity: hematoxylin and eosin stain (H&E);  $\times 100$  magnification. (D) Ki-67 proliferation index was 15% percent: KI 67; X100 magnification.



## DISCUSSION

HPCs have a tendency to metastasize to extracranial localizations at a rate of 11.6 to 69% within 10 years after diagnosis; mainly to bone, lung and liver.<sup>4</sup> So far, only twelve HPCs with spinal metastasis either in intradural, extradural or both of the locations have been reported. Of these, only two were illustrated.<sup>3,6</sup> Furthermore; all of the reported cases were in adult age group. Infantile HPCs (<1 years) are reported to be less aggressive and have a better prognosis.<sup>4</sup> But there is no known difference in prognosis of the cases that occur under 18 years of age. This report highlights the rare but probable aggressive course of tumor dissemination through the CSF in child age group.

MRI is highly useful in detection and follow-up of HPC.<sup>3</sup> The tumor is generally isointense to hypointense on T1-weighted images; and isointense to hyperintense on T2 weighted images. Since the tumor consists of vessels; high vascularity of the tumor is also important during surgery that intraoperative bleeding is a possible complication.<sup>7</sup> As shown in our case; total neuraxis imaging in the initial presentation as well as follow up imaging for all children diagnosed with intracranial hemangiopericytomas is essential to avoid missing any progression of the disease.

The primary treatment for intracranial HPCs is surgery. Gross total resection improves the overall survival and progression free survival; when compared to subtotal resection. The role of adjuvant radiotherapy after resection is controversial.<sup>5</sup> It is probably due to heterogeneity of the patients with WHO grade II and grade III tumor pathologies. The largest study with 52 WHO grade III HPC patients with a mean of 36.8 months follow-up showed that adjuvant radiotherapy increased both overall survival and progression free survival. The recommended standard dose of radiotherapy is 50 Gy.<sup>5</sup> Unfortunately, there is no evidence that complete resection and adjuvant radiotherapy decrease the distant metastasis rates.<sup>8</sup> As a prognostic factor; Kumar *et al.* reported that; WHO grade III tumors had aggressive courses and there is 37 months of median recurrence free interval compared to a median of 68 months with Grade II tumors.<sup>4</sup> Even so; metastasis can occur after 10 to 15 years after diagnosis.<sup>8</sup>

In the setting of spinal HPCs, main treatment option is resection and adjuvant radiotherapy similar to intracranial HPCs.<sup>9</sup> Preoperative endovascular embolization of the tumor is recommended to reduce blood loss during

surgery.<sup>3,7</sup> Our case did not receive preoperative embolization since there were symptoms of acute spinal cord compression. Systemic chemotherapy in the treatment of HPCs has been found ineffective.<sup>9</sup> Still, doxorubicin seems to be preferred chemotherapeutic agent by some.

In conclusion, intracranial HPC can cause intraspinal drop metastasis also in the child age group. Initial and follow-up imaging of whole neuraxis with MRI should be obtained not to miss any progression of the disease in pediatric age group.

## DISCLOSURE

Financial support: None

Conflict of Interest: Nothing to declare.

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