REPORTE DE CASO

# SOLITARY FIBROUS TUMOR/HEMANGIOPERICYTOMA (SFT/HPC); A CASE REPORT IN A 15 YEARS OLD PATIENT AND REVIEW OF THE LITERATURE

Tumor fibroso solitario / Hemangipericitoma (TFS/HPC); reporte de caso en un paciente de 15 anios y revisión de la literatura

Tumor fibroso solitário / Hemangiopericitoma (TFS / HPC); relato de caso em paciente de 15 anos e revisão de literatura

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## ABSTRACT

Fibrous solitary Tumors/Hemangiopericytomas are tumors of the central nervous system that over the years have been studied more intensely, they have similar findings and behaviors with meningioma's, which presents us with a

great challenge when defining a diagnosis and a medical or surgical management. They have a low incidence and their classification is strict according to their degree of cellularity and vascular compromise. Presentation is unusual in young people, the greatest incidence is between the fourth and sixth decade of life with a predominance for males, in this report we show a patient with 15 years old in which there is evidence of a lesion compatible with an anaplastic hemangiopericytoma or grade III Fibrous solitary Tumor/Hemangiopericytoma according to the WHO classification to whom neurosurgical and adjuvant management was provided with a 100% resection of the lesion.

**Keywords**: fibrous solitary tumor, hemangiopericytoma, tumors, central nervous system, pericytes.

## RESUMEN

Los tumores fibrosos solitarios / hemangiopericitomas son tumores del sistema nervioso central que a lo largo de los años han sido estudiados, tienen hallazgos y comportamientos similares a los meningiomas, lo que nos presenta un gran desafío al definir un diagnóstico y un tratamiento médico o quirúrgico. Tienen una baja incidencia y su clasificación es estricta según su grado de celularidad y compromiso vascular. La presentación es inusual en los jóvenes, la mayor incidencia es entre la cuarta y sexta década de la vida con predominio en los hombres, en este informe mostramos a un paciente con 15 años de edad que presenta una lesión compatible con un hemangiopericitoma anaplásico o tumor III fibroso solitario / hemangiopericitoma de acuerdo con la clasificación de la OMS a quien se le proporcionó un tratamiento neuroquirúrgico y adyuvante con una resección del 100% de la lesión.

**Palabras Clave**: tumor fibroso solitario, hemangiopericitoma, tumores, sistema nervioso central, pericitos.

## RESUMO

Tumores fibrosos solitários / hemangiopericitomas são tumores do sistema nervoso central que foram estudados ao longo dos anos, apresentam achados e comportamentos semelhantes aos meningiomas, o que nos apresenta um

grande desafio na definição de um diagnóstico e tratamento médico ou cirúrgico. Apresentam baixa incidência e sua classificação é rigorosa de acordo com o grau de celularidade e comprometimento vascular. A apresentação é incomum em jovens, a maior incidência é entre a quarta e a sexta década de vida com predominância nos homens. Neste relato, mostramos um paciente de 15 anos que apresenta uma lesão compatível com um hemangiopericitoma anaplásico ou tumor III / hemangiopericitoma fibroso solitário, de acordo com a classificação da OMS, que receberam tratamento neurocirúrgico e adjuvante com ressecção de 100% da lesão.

**Palavras chave:** tumor fibroso solitário, hemangiopericitoma, tumores, sistema nervoso central, pericitos.

## INTRODUCTION

Intracranial hemangiopericytoma (HPC), also known as vascular pericytes tumor, is a rare malignancy tumor, usually originates from malignant cells of meningeal mesenchyme. Initially described by Stout and Murray in 1942. It only accounts for 0.4% of all primary central nervous system tumors (Zhou et al. 2012) and less than 1% of all primary intracranial tumors (Myeong et al. 2019). These lesions were initially classified as a vascular type of meningioma; however, subsequent investigations resulted in the World Health Organization recognizing hemangiopericytomas as a distinct clinic-pathological entity in 1993 (Chantrain et al. 2006). Lesions arise from pericytes, also called mural cells. They are found across diverse human organs, pericytes were identified around arterioles, capillaries and venules, but are more frequent located in retina and brain (Murray IR et al. 2013).

Over the past decade, soft tissue pathologists have moved away from the designation HPC, diagnosing such tumors within the spectrum of solitary fibrous tumors; according to 2016 Central Nervous System – World Health Organization the correct term is Solitary Fibrous Tumor/Hemangiopericytoma (SFT/HPC) and assigns three grades: Grade I that corresponds to highly collagenous, relatively low cellularity and spindle cell lesion, previously diagnosed as SFT; Grade II corresponds to more cellularity, less collagenous, tumor with plump cells and “stag horn” vasculature that was previously diagnosed in the CNS as HPC; and Grade III that corresponds to what was termed Anaplastic Hemangiopericytoma

in the past, diagnosed by 5 or more mitoses per 10 high-power fields. (Louis et al. 2016)

Incidence of SFT/HPC occurs in only 5% to 10% of cases in children. The lower limbs are the main site of tumor location followed by head, neck region, pelvis, and visceral organs. 10% to 20% of patients have metastatic disease at the time of diagnosis (Fernandez et al. 2011). In children older than 1 year, SFT/HPC does not differ significantly from adult SFT/HPC in which its influence is between the fourth and the sixth decade of life with predominance in men (Myeong et al. 2019) and surgical therapy is the mainstay of treatment. In children younger than 1 year, SFT/HPC seems to have a better response to chemotherapy than in adults and spontaneous regression has also been documented (Rodriguez et al. 2000). Intracranial SFT/HPC represents a special location with a high proclivity toward recurrence and metastasis. Gross total resection (GTR) is associated with better outcomes among patients with intracranial location (Rutkowski et al. 2010), (Shettya et al. 2010).

## CASE PRESENTATION

We present the case of a previously healthy patient, male of 15 years old who present 10 days of right hemicranial headache associated with nausea, emesis and high intensity photophobia. The headache triggers frequent nocturnal awakenings, concomitant changes in behavior, apathy, psychomotor agitation and occasional disorientation episodes.

At admission to the emergency department of the Colombian Carlos Ardila Lulle Hospital is performed complete neurological examination that evidence patient without neurological focus, without motor or sensory deficit, eye fundoscopy with severe papilledema. Brain CT and MRI shows the presence of a mass that is compromising the greater part of the right frontal lobe, which measures approximately 6.0 cm in its greater diameter, surrounded by perilesional edema that is exerting important signs of compressive effects with deviation of the structures of the midline from right to left approximately 1.0 cm. (Figure 1)

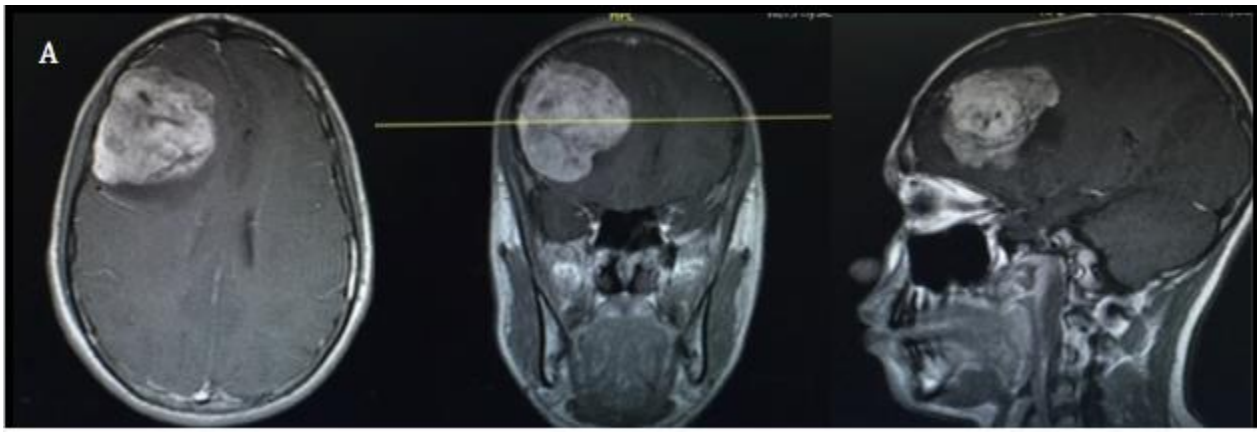


**Figure 1**: Brain CT (A) shows the presence of a mass that is compromising the greater part of the right frontal lobe, which measures approximately 6.0 cm in its greater diameter, surrounded by perilesional edema that is exerting important signs of compressive effects with deviation of the structures of the midline from right to left approximately 1.0 cm. (Source: authors).

Baseline blood and imaging studies were performed and didn't show any alteration in their results. Therefore, the patient is taken within 48 hours to surgery for tumor resection. Right coronal incision was made. During surgery, friable and vascularized necrotic tissue is found, with angiogenesis and severe infiltration of the cerebral parenchyma. Samples of pathologies are sent for immunohistochemical studies observing histological sections that show fragments of a malignant mesenchymal tumor, with densely cellularity, composed of elongated nucleus fusiform cells, vesicles of chromatin, evident nucleolus, frequent figures of mitosis (up to 10 in 12 high power fields) and cariorrexis, arranged in irregular beams and with storiform pattern, surrounding frequent branched capillars and occasional medium-sized glasses. It is associated with small foci of necrosis and hemorrhage. (Figure 2)

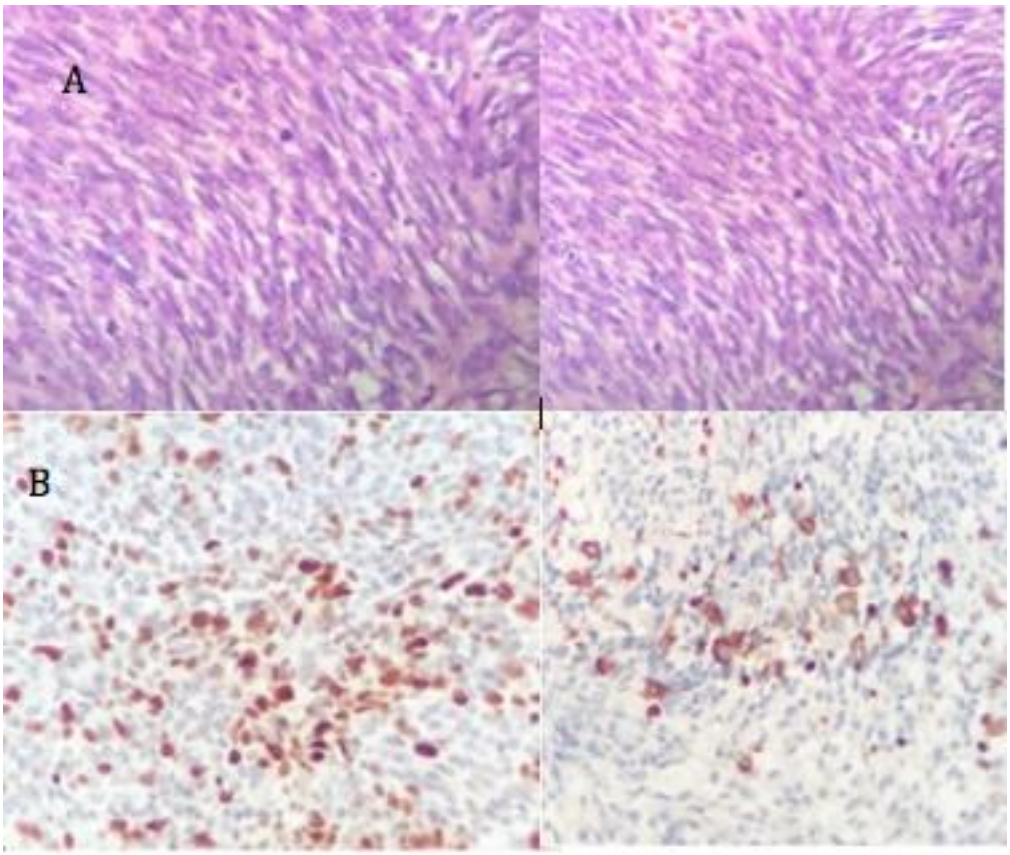


**Figure 2**. Histological sections shows fragments of a malignant mesenchymal tumor, with densely cellularity, composed of elongated nucleus fusiform cells, vesicles of chromatin, evident nucleolus, frequent figures of mitosis (up to 10 in 12 high power fields) and cariorrexis, arranged in irregular beams and with storiform pattern, surrounding frequent branched capillars and occasional medium-sized glasses. It is associated with small foci of necrosis and hemorrhage. (Source: Authors).

Immunohistochemistry reveals tumor cells that show diffuse positivity for Vimentina; focal positivity for CD34, Desmina and AML (isolated cells); are negative for progesterone receptors, EMA, S100 and PGFA, the CD34 marker highlights the vascular component of the tumor. The reticulum coloration shows a well-formed reticulinic weave around the tumor cells; the rate of vascular proliferation measured with Ki67 is up to 50% (Figure 2B) obtaining as result: brain samples compatible with anaplassic hemangiopericytoma (GRADE III) with adjacent infiltration.

The surgery was performed without complications. Posterior studies evidence a great resection had been achieved. (Figure 3)

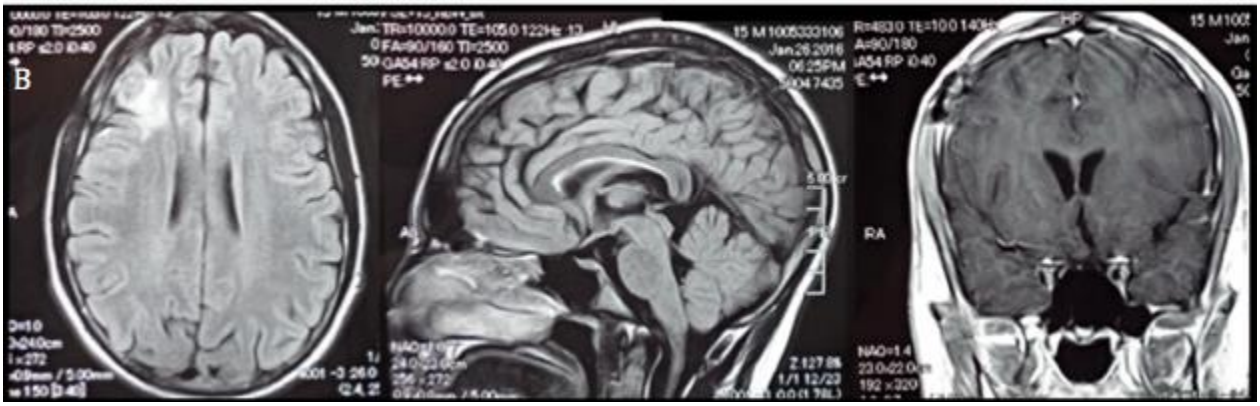

**Figure 3**. Post-operative MRI shows evidence of complete resection had been achieved. (Source: authors).

During recovery, the patient presents generalized clonic tonic seizures with adequate control with antiepileptic treatment. The radiotherapy service considers it necessary to perform radiation on the surgical bed and central nervous system.

## DISCUSSION

Hemangiopericytoma was first reported and named by Stout and Murray in 1942. It is a rare soft tissue tumor and may occur in any part of body but less commonly in the central nervous system (Thanni LO. 2005). In the past, it was believed to originate from the meninges, and hence was considered a subtype of meningioma, given by its characteristics and locations, according to 2016 CNS WHO the correct term is solitary fibrous tumor / hemangiopericytoma SFT/HPC due to its origin from interstitial capillaries Zimmerman meningeal cells, with high cell density, collagenous, fibrin brands and multi-differentiation potential cells with hypervascularity and fragile vessels in tumor. HPC is almost always attached to the dura and has a strong tendency to recur locally and to metastasize outside the central nervous system to bones, lungs and liver (Melone et al. 2014). These extremely vascular tumors are challenging to treat, especially in pediatric cases because of the smaller overall blood volume in children, the surgeon must be careful when interrupting the blood supply to the tumor, and preoperative endovascular embolization is recommended. Infantile HPC is characterized by a more benign course, with responsiveness to

chemotherapy and even a tendency to spontaneous regression. In contrast, the behavior of HPC in children older than 1 year does not seem to differ from HPC in adults. The differential diagnosis of HPC includes infantile hemangioma, infantile myofibromatosis, leukemia cutis, neuroblastoma, Langerhans cell histiocytosis, Kaposiform hemangioendothelioma, tufted angioma, pyogenic granuloma, fibrosarcoma, glomus tumor, rhabdomyosarcoma, reticulosarcoma, and angiosarcoma by the similarity in clinical behavior and radiographic findings and presentation (Laviv et al. 2012). Histological confirmation is the only definitive means of distinguishing these lesions that typically share inversions at 12q13, fusing the NAB2 and STAT6 genes which leads to STAT6 nuclear expression that can be detected by immunohistochemistry (Myeong et al. 2019).

The clinic of these patients is in relation to the location and size of the lesion, such as in the case is here presented, usually the clinic symptoms are headache, neurological deficit and seizures. According to Zhou Jun-lin *et all* a retrospectively review of Thirty-nine cases of surgically and pathologically confirmed HPC and anaplastic HPC, the lesions were commonly located in the skull base(1), lateral ventricle, pineal region and intraparenchymal area (Sulit et al. 2011) Other study by Sulit DJ *et all* with a total of 43 patients with intracranial HPC were treated from 1980 to 2010, these tumors where located frequently superficial and closely to the meninges, in the falx/parasagittal area (55%). Eight (18.6%) tumors were associated with venous sinus compression or invasion. The most common symptoms were headache (40%) and upper and/or lower limb weakness (12%) however, this study did not include children (Chen et al. 2012).
Respect to the images, HPC and meningioma's are both extra-axial tumors presenting with similar imaging findings, including dural tail and contrast enhancement. However, there are some clues not fully discriminative, that may help in differentiating HPCs from meningioma's: HPCs have heterogeneous, enhancement pattern and narrower dural tail (Kalra et al. 2012), also don't present classifications and hyperostosis of the skull. Barba et al. evaluated MRS findings in three adult patients with HPC at 1.5T. They report absence of NAA, and moderate peaks of Choline and Glutamate together with prominent myoinositol in those patients and conclude that the higher concentration of myoinositol and a glycine at the 3.56 ppm peak can be used as an important discriminator of HPC. Trimethylamine (choline) levels in HPC are higher than the ones founded in meningioma's (Barba et al. 2001). Nevertheless, according to the review of Koc AM *et all* in 2016 imaging workup of HPCs in the pediatric age

can fail to show any MRS findings that might help their differentiation from meningioma's, in contrast to the adult type (Koc et al. 2016).

## CONCLUSION

In conclusion, we preset a clinical case in which we can show one of the highest grades in terms of the classification of SFT/HPC (Grade III), in this case given by histological and pathological findings, also describing it's condition in the CNS and comparing it with one of the most frequent tumors of it, which can cause us confusion when defining a direct therapy to one of these two types of tumors, we could also demonstrate the benefit that patients have when being taken to volumetric resection of the injury. Currently, it was suggested to continue with adjuvant management with radiotherapy to eliminate the residual lesion.

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