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J. Christopher Cole
L. Gill Naul

Intracranial infantile hemangiopericytoma

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J. C. Cole (✉) · L. G. Naul
Department of Radiology,
Scott & White Clinic and Memorial Hospital, 2401 South 31st Street, Scott, Sherwood and Brindley Foundation, Texas A&M University Health Science Center, College of Medicine, Temple, TX, USA

Abstract Hemangiopericytoma (HP) is a rare vascular tumor that usually occurs in adults, but 10 % occur in children. Tumors occurring in the first year of life are even more rare and are referred to as infantile hemangiopericytoma. In this article, we report the imaging, operative, and pathological findings in a patient with an infantile HP.

Introduction

Hemangiopericytoma (HP) is a rare vascular tumor that was first described by Stout and Murray in 1942 [1]. The majority of these tumors occurs in adults, with only 10 % of cases occurring in children [1]. Infantile HPs, those occurring within the first year of life, are even more infrequent and are associated with a more benign course [1]. A review of the literature revealed numerous cases in adults, but only six cases of intracranial HP have been reported in children under the age of 1 year [1]. In this article, we report the imaging, operative, and pathological findings in a patient with an infantile HP and, through a review of the literature, discuss the characteristic pathological and imaging features of this tumor.

Case report

A 6-week-old boy, whose initial symptoms were three episodes of tonic-clonic seizures, was admitted to our hospital. Each episode lasted approximately 15 s with a subsequent postictal state. Projectile vomiting was also associated with each episode. The infant had been healthy without fever or trauma. He was full term and had an uncomplicated vaginal delivery. He exhibited, on examination, no increased irritability or change in his normal behavior. The patient was afebrile, alert, active, and in no distress. The neurologic examination was unremarkable.

A computed tomography (CT) scan (Fig. 1) was obtained and revealed a 5 × 2-cm lobulated mass lesion intracranially in the right parieto-occipital region. There were multiple hyperdense areas within the mass consistent with calcifications. There was a minimal amount of associated surrounding edema without significant shift of the midline structures. There was very dense contrast enhancement of the entire mass. On magnetic resonance imaging (MRI) (Fig. 2), T1-weighted (T1W) images were performed before and after administration of gadolinium diethylene triamine penta-acetic acid (DTPA). In addition, T2-weighted (T2W), proton density, and gradient echo images were performed. A vigorously enhancing intra-axial lesion with a small amount of edema was identified. On T2W images, the mass was hypointense, relative to adjacent gray matter. On T1W images the mass was isointense, relative to the normal gray matter and hyperintense to white matter. There were areas of increased signal intensity consistent with calcifications on the T1W images. Gradient echo images did not show evidence of hemorrhage; however, there were areas of diminished signal intensity compatible with calcifications. There were no abnormal flow voids, and the magnetic resonance angiogram of the brain was negative. The differential diagnosis included a primitive neuroectodermal tumor (PNET), an intra-axial ependymoma, a teratoma, and a vascular lesion.

At surgery, a rubbery hypervascular tumor was identified, intra-axially, in the right parietal region. The tumor was encapsulated and large vessels were present over the anterior surface. Gross removal of the tumor was performed without significant bleeding.

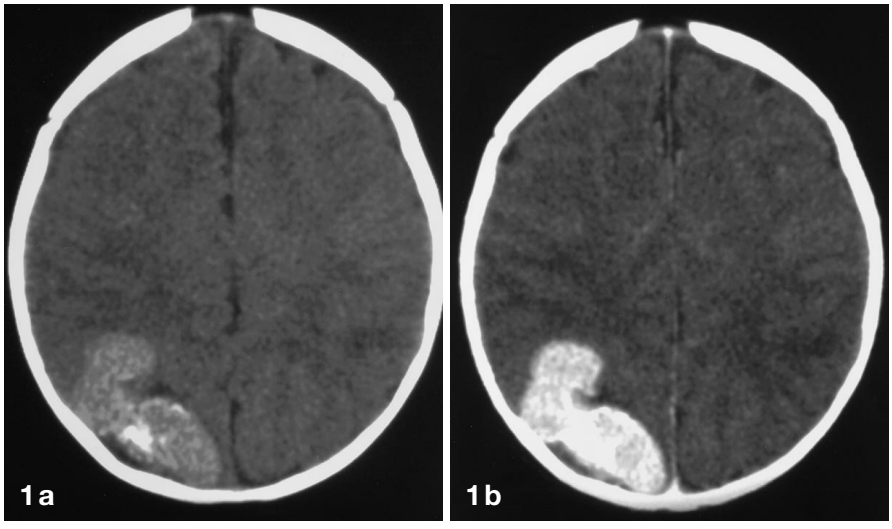

Fig. 1 a Axial CT image without contrast demonstrates a lobulated intra-axial mass with calcifications (84 HU) in the right parietal-occipital region. **b** Axial CT image after contrast demonstrates vigorous enhancement of the mass

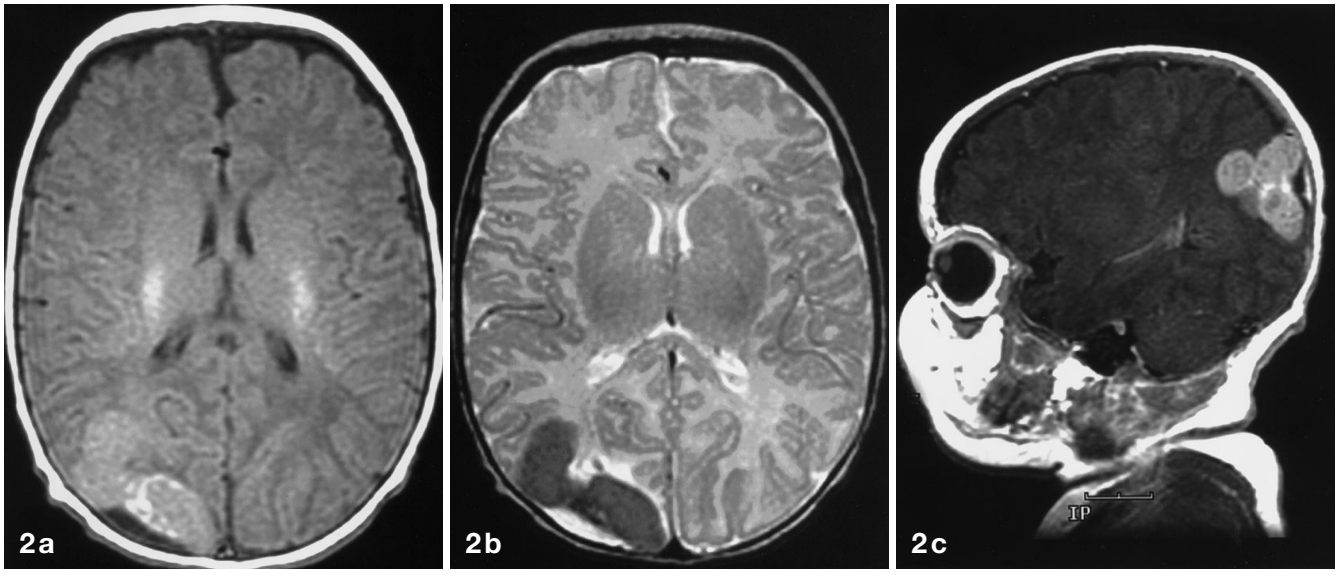

Fig. 2 a T1W (450/9) MRI shows a mass isointense to gray matter and hyperintense to white matter. **b** T2W (4000/90) MRI shows a hypointense mass, relative to gray matter. **c** Vigorous contrast enhancement with Gadolinium DTPA is demonstrated on sagittal T1W (450/9) MRI

On microscopic examination, the tumor had a variable appearance in multiple areas. Some areas were more cellular than others and were composed of epithelioidlike cells with staghornlike intervening spaces. There was a fairly sharp demarcation between the tumor and the adjacent brain parenchyma without evidence of invasion. Because of the unusual nature of the tumor, immunohistochemical stains were performed. The keratin stains were negative, ruling out an epithelial neoplasm. Epithelial membrane antigen (EMA) stains were negative, making it unlikely that this would represent a meningioma. Glial fibrillary acidic protein (GFAP) stains were negative in the neoplastic tissue, ruling out a glial origin neoplasm. Vimentin stains were positive, demonstrating a mesenchymal origin of the lesion. Reticulin stains demonstrated a staghornlike arrangement of the cellular proliferation. The final pathologic diagnosis was hemangiopericytoma.

Following resection, the patient did well and follow-up CT scan at 1 month revealed postoperative changes without evidence of residual or recurrent tumor.

Discussion

The tumor arises from the pericytes of Zimmerman, first described in 1923, which may occur at any site throughout the body where capillaries are found [2]. The incidence of HP of the central nervous system (CNS) is less than 1% of all CNS tumors, presenting most often in the third to fourth decade of life, but can occur at any age [2]. Because these tumors were thought to arise from the meninges, they were originally categorized as an angioblastic meningioma, but they are now recognized as a distinct entity [3]. Arguments for this include: (1) morphological similarity of the tumor with the hemangiopericytoma in other parts of the body; (2) existence of pericytes in the blood vessel walls within the CNS; (3) fine structural differences from the arachnoid cells of meningiomas; and (4) similarities found on elec-

tron microscopy [4]. Also, reticulin staining demonstrates its relationship to vascular channels [3]. The 1993 classification of the World Health Organization (WHO) eliminated the term "angioblastic meningioma" in favor of "hemangiopericytoma" [3, 5]. The diagnosis of primary HP of the CNS is histological.

Intracranial HPs are neoplasms of pericytes that originate in a histologically pure form without a meningioma component. These tumors are rare multilobulated masses. They are well-demarcated masses, which are reported to be heterogeneous and predominately isointense on T1W and T2W images, some of which demonstrate signal flow voids [5, 6]. On unenhanced CT, they are slightly hyperdense and may show bony erosion, but usually lack the calcifications and hyperostosis associated with meningiomas. Heterogeneous contrast enhancement is usually demonstrated on both CT and MRI [5, 6]. They tend to have a high recurrence rate and metastasize frequently to lung, liver, kidneys, pancreas, adrenals, and bone [5].

An infantile form of HP, those that occur within the first year of life, has been recognized as having a benign course. Histologic features of frequent mitosis, increased cellularity with pleomorphism, necrosis, and hemorrhage are usually indicative of malignant behavior in HP, but can also be demonstrated in benign infantile HP. There is no clear distinction, histologically, between benign HP and malignant HP, and the diagnosis is frequently based solely on the tumor's clinical behavior [1].

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