

Intracranial capillary hemangioma mimicking a dissociative disorder

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Abstract

Capillary hemangiomas, hamartomatous proliferation of vascular endothelial cells, are rare in the central nervous system (CNS). Intracranial capillary hemangiomas presenting with reversible behavioral abnormalities and focal neurological deficits have rarely been reported. We report a case of CNS capillary hemangioma presenting with transient focal neurological deficits and behavioral abnormalities mimicking Ganser's syndrome. Patient underwent total excision of the vascular malformation, resulting in complete resolution of his symptoms.

Case Report

A 59-year-old man was brought in to the hospital with worsening confusion and progressive memory loss over past 4 months. He was noted to be unsteady with history of multiple falls over the last few months. The patient gives history of sporadic memory loss and easy irritability. The patient had increasing difficulty communicating with his wife and expressing himself. He had occasional headaches, but denied any vomiting, seizures or visual abnormalities. His family also mentioned that he had been having irrelevant talks at times. His past medical history is significant for chronic heavy alcoholism and occasional bouts of gout. He denies smoking. No significant family history of any neurological disorders or malignancy. Vitals at the time of admission were BP 150/84 mm/Hg, pulse rate of 90/min regular, respiratory rate-14/min and oxygen saturation of 100% on room air.

General examination revealed a well-built man with psychomotor agitation. Neurological examination revealed decreased levels of orientation as he could not recall the date and time. Mini mental status examination score was 25/30 with occasional absurd answers in between. Cranial nerves are intact. The patient had motor, sensory and visual neglect on the left side. No truncal ataxia noted. Gait examination revealed minimal swaying to the left.

Rests of the system examinations were normal.

His laboratory works revealed hemoglobin of 12.9 gm/dL (13.2-16.2 gm/dL), platelet count of 86,000 (100,000-450,000), plasma glucose of 104 mg/dL (60-110 mg/dL), BUN 11 mg/dL (7-20 mg/dL) and creatinine 1.18 mg/dL (0.5-1.4 mg/dL). Sodium 135 mEq/L (135-147 mEq/L), potassium 3.8 mEq/L (3.5-5.2 mEq/L), chloride 102 mEq/L (95-107 mEq/L), calcium 8.7 mg/dL (8.8-10.3 mg/dL), alkaline phosphatase 66 U/L (38-126 U/L), alanine aminotransferase 47 U/L (7-56 U/L), aspartate aminotransferase 82 U/L (5-35 U/L), protein 7.9 mg/dL (6.3-8.2 mg/dL), albumin 3.3 mg/dL (3.2-5.0 mg/dL) and TSH 0.3 U/mL (0.4-4.5 U/mL). His serum alcohol levels and serum toxicology screen was negative. Vitamin B12 and folate levels were within normal limits.

Initial non-contrast computed tomography (CT) of the head showed 5.0×4.7 cm in diameter well-circumscribed rounded hypo attenuated mass lesion involving the right temporoparietal region. There is surrounding cerebral edema and a right to left midline shift of 9.0 mm (Figure 1). Further imaging studies with gadolinium-enhanced magnetic resonance imaging (MRI) showed a large cystic intra-axial lesion centered at the right temporal-parietal junction measuring 4.8×4.7×5.1 cm. The lesion is surrounded by white matter edema involving the entire temporal and parietal lobes and portions of the occipital and frontal lobes. The cystic lesion has a well-defined wall with slight enhancement but does not contain any visible internal debris and has homogeneous fluid signal. There is 8 mm of right to left midline shift with significant compression of the right lateral ventricle but no overt hydrocephalus. The cystic lesion has a small enhancing mural nodule measuring 6×6 mm in its margin (Figure 2). There was no evidence of hemorrhage on either the CT or the MRI. Further imaging studies of the chest, abdomen and pelvis to rule out a primary malignancy or associated lesions with contrast enhanced CT were non-revealing. The differentials based on CT and MRI appearance would include a cystic metastasis, glioblastoma multiforme, ganglioglioma, hemangioblastoma and neuroglial cyst. The presence of an enhancing mural nodule favors a cystic metastasis and hemangioblastoma.

Patient underwent right temporo-parietal craniotomy. Cerebral cortex was markedly edematous and swollen. Underlying mass was cystic in nature with dilated vascular channels along with gelatinous fluid in it. It was surrounded by a thin layer of membrane.

Biopsy findings of the lesion were consistent with vascular malformation with organizing hematoma. Immunohistochemical studies with CD 34, factor V111 and smooth muscle myosin heavy chain (SMMHC) confirmed the

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Key words: intracranial capillary hemangioma, Ganser's syndrome, dissociative behavior, focal neurological deficits.

Received for publication: 8 November 2011.

Revision received: 31 January 2012.

Accepted for publication: 9 February 2012.

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Clinics and Practice 2012; 2:e35
doi:10.4081/cp.2012.e35

diagnosis as cerebral capillary hemangioma. The sample was also sent to Mayo clinic for confirmation of the diagnosis. Patient had a very rapid and complete recovery of his symptoms postoperatively. Follow up imaging studies of the brain with contrast MRI after 3 months showed no evidence of recurrence or residual lesion.

Discussion

Capillary hemangiomas are benign tumors or tumor like lesions predominantly seen in the skin and soft tissues.¹ Most of these vascular malformations occur predominantly in children. There are limited reports on the incidence of capillary hemangiomas in the central nervous system.² Capillary hemangiomas consist of vascular channels lined with a layer of endothelial cells and vascular spaces consisting of capillaries without intervening brain parenchyma.³

Capillary hemangiomas are usually solitary, but multiple lesions have been found in approximately 20% of affected newborns.⁴ Most cases of capillary hemangiomas are benign, but functional complications have been reported in 20% cases while 3-5% cases developed life-threatening complications.⁵ Histological types of capillary hemangiomas vary from hemangiomas of infancy, epithelioid hemangiomas, lobular capillary hemangiomas, also known as pyogenic granulomas and variants of capillary hemangiomas.⁶ An extensive literature review of cases of capillary hemangiomas of the neuraxis by Abe, *et al.* observed that they have the same histological findings as lesion elsewhere in the body.¹ Central nervous system capillary hemangiomas need to differentiate from other forms of vascular malformations/tumors

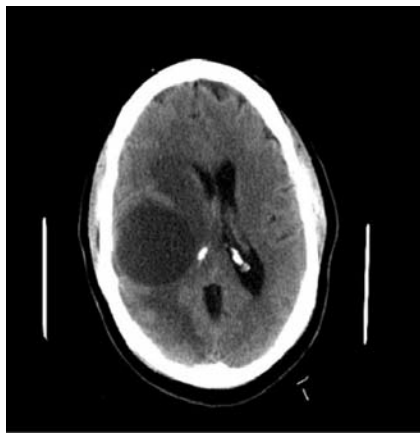


Figure 1. Brain computed tomography: there is a 5×5 cm hypo attenuated mass in right parieto-temporal region with surrounding cerebral edema and associated right to left midline shift. There is no acute hemorrhage or calcifications in the lesion.

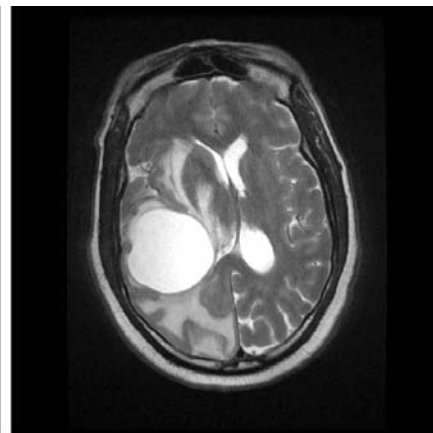
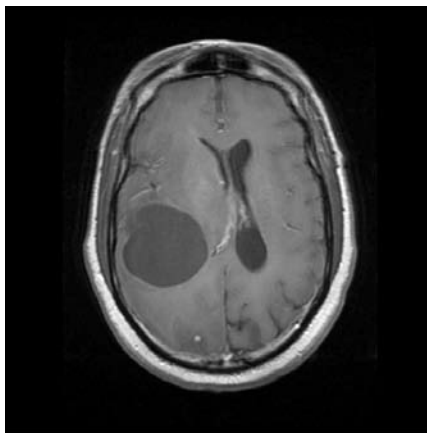


Figure 2. Brain magnetic resonance imaging with contrast: there is a 5×5 cm intra-axial cystic lesion with enhancing mural nodule in its wall in the right parieto-temporal region. There is associated right to left midline shift and surrounding cerebral edema. There is also a 3 mm intra-axial enhancing solid lesion in the right posterior parietal region. No other areas of abnormal post contrast enhancement are demonstrated in the brain. No areas of blooming artifact were demonstrated on the gradient sequence to suggest blood degradation products.

including hemangioblastoma, hemangioendothelioma and hemangiopericytoma considering the aggressive nature of the latter.

Intracranial capillary hemangiomas are rarely known to present with transient focal neurological deficits. Intracranial capillary hemangioma presenting with behavioral abnormalities in the form of dissociative disorder has not been reported in literature. Our patient clinical features were consistent with Ganser's syndrome, which is a disorder in which the person mimics behavior that is typical of a mental illness, such as schizophrenia. Ganser's syndrome has also been associated with various other functional psychiatric disorders and organic states.⁷ This condition is also seen secondary to stroke and head injury, predominantly affecting the frontal lobes. There are reports of Ganser's syndrome secondary to hemorrhage in the temporo-parietal lobe.⁷ There are also evidences in literature to support both organic and hysterical reasons for Ganser's syndrome. In our patient the intracranial space occupying lesion in the frontotemporo-parietal region was completely resected, which resulted in complete resolution of the symptoms. It has been widely accepted that surgery seems to be a reasonable treatment option for superficial CNS capillary hemangiomas presenting with neurological deficit. Risk of hemorrhage is high in deep-seated lesions. Alternative therapeutical approaches would include corticosteroids, arterial embolization and irradiation.⁸ Total resection of the lesion is possible in majority of cases. However there are case reports on multiple

intracranial capillary hemangiomas, where surgical intervention was deferred.² Post treatment it is recommended to do follow up imaging studies based on the observation that cutaneous and soft tissue hemangiomas recur, though the frequency and methods of choice are still not defined.⁶ Our patient underwent a follow up imaging study of brain with contrast MRI, which did not show any new lesions or of the residual tumor.

Conclusions

Central nervous system capillary hemangiomas are extremely rare. Clinical presentation of central nervous system capillary hemangioma may vary from transient focal neurological deficit to behavioral abnormalities or even a combination of these two. To best of our knowledge, there has been only a very few central nervous system capillary hemangiomas presenting as a dissociative disorder in English literature. Our case is one of the very few reported case of a capillary hemangioma of the central nervous system presenting with features consistent with Ganser's syndrome with complete resolution of the symptoms following surgery. Even though this disease entity is rare, physician needs to be cognizant of its presence and the need for prompt medical and surgical intervention. Early recognition of this rare disease entity would prevent unnecessary therapeutic interventions in these patients.

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