Case Report

Paroxysmal Sympathetic Hyperactivity Due to Thrombosis of the Vein of Galen: A Significant Clinical Imitator

*Awad Magbri, Seth Harshit, Eussera El-Magbri, Mariam El-Magbri, Shaukat Rashid, Balhinder Brar

*Toledo Vascular Access Center, Toledo OH and Life-care Hospital of Pittsburgh, PA, USA

Abstract

A case of thrombosis of the vein of Galen (DCVT) is presented with hemiparesis, seizure activity, altered mental status, and sympathetic hyper-activity in young man with congenital atrial septal defect. The authors discussed the difficulties in the diagnosis in DCVT cases, the pathogenesis and the treatment of this life threatening condition. Management of sympathetic hyperactivity with bromocriptine and baclofen and anticoagulation of the DCVT are the main stay of treatment that if implemented early can make a huge difference in patient’s lives.

Keywords

Central Vein Thrombosis; Deep Central Vein Thrombosis; Anticoagulation; Sinus Venosus Atrial Septal Defect; Ventriculitis; Celebrities; Sympathetic Hyperactivity; Thrombophilia; Meningoencephalitis; Japanese Encephalitis; Vein of Galen

Introduction

Case History

The case is that of 32-year-old African American male with medical history significant of catastrophic intracranial hemorrhage with right-sided hemiparesis secondary to arterial-venous malformations. He has residual right-sided hemiparesis with spasticity, status post baclofen pump implantation. His medical history is also significant of status post ventriculo-peritoneal shunt (VP) shunt, history of pulmonary embolism, status post IVC filter placement, and history of high fevers, seizure activities, and ASD. He ambulated with quad cane and wheel-chair prior to his admission to the hospital. His family history is positive for diabetes mellitus and hypertension.

He was admitted to the hospital with altered mental status, seizure activity, tachycardia, hypotension, and poor responsiveness. In the emergency department there was a concern of non-convulsive status epilepticus along with poor responsiveness, and therefore, he was intubated for airway protection. His initial CAT scan of the head showed findings suggestive of possible VP shunt malfunction which was noted to be on the left side of the foramen of Monro associated with dilation of the left lateral ventricle and significant dilation of the vein of Galen with peripheral calcifications indicative of partial thrombosis.

The patient was evaluated by the neurosurgeon who initiated anticoagulation with heparin at high-risk protocol. He underwent MRI that showed small areas of infarct involving the right cerebellum with a questionable area of sub-acute infarct and evolving hematoma in the right thalamus, along with asymmetric enlargement of the temporal horn of the left lateral ventricle. His shunt was reprogrammed and eventually had a right VP shunt.
shunt placement which was subsequently removed. His continuous EEG was indicative of frontotemporal seizure tendency. His repeated CAT scan of the brain revealed a new left frontal infarct. The patient had echocardiography both (TEE and TEE) which showed left to right shunt and ejection fraction of 55-65%, with sinus-venous atrial septal defect (ASD).

He continued to have periods of agitations, diaphoresis, and tachycardia with increased spasticity. The patient had a lumbar puncture out of concern of ventriculitis and celebrities which showed negative CSF culture and gram stain, high protein and lymphocytic pleocytosis. His echocardiography was also negative for vegetation and endocarditis. His hospital course was stormy necessitating placement of tracheostomy with ventilator support, PEG tube for feeding, and treatment of sympathetic hyper-activities with bromocriptine and propranolol. He was investigated thoroughly for thrombophilia with no cause found. He was also treated with vancomycin, keppra, aztreonam, Neurontin, bromocriptine, propranolol, baclofen by intrathecal pump, and Coumadin. He made a remarkable recovery and eventually he was transferred to rehabilitation.

**Case Discussion**

Deep cerebral venous thrombosis (DCVT) is an uncommon disease causing stroke in adults of 3-4 per million population annually [1]. Although, there is no population based studies on the incidence of cerebral vein thrombosis (CVT) comprises 10% of all strokes in India [2]. The most frequent thrombotic sites are the lateral cavernous, and superior sagittal sinuses. Thrombosis of the deep venous system, such as internal cerebral veins, vein of Galen and straight sinus occur far less with a worse prognosis [1, 3, 4]. Deep cerebral vein thrombosis is a term used for thrombosis of internal cerebral veins, vein of Galen, basal vein of Rosenthal and its tributaries and is seen in 3-8% cases of CVT [5-7].

The non-specific clinical features of DCVT makes early diagnosis difficult, however, the applications of brain magnetic resonance imaging (MRI) is helpful. Once DCVT is confirmed, anticoagulant therapy should be started to prevent further thromboembolism [1, 3, 4]. A multicenter study of 624 adult patients with CVT estimated a mortality of 3.4% in 30 days [4].

The various precipitating factors that lead to DCVT include inherited thrombophilia states, hyperhomocystinemia, dehydration, drugs, pregnancy, malignancy and autoimmune diseases. However, in 15% of cases of DCVT no direct cause or predisposing factor could be identified [8].

The diagnosis of CVT is challenging and difficult due to diverse clinical presentation and is often missed or delayed. Headache is the most common symptom and is seen in almost 70-90% of cases [9]. Hemiparesis, seizures, loss of consciousness, papilledema and visual disturbances are other commonly observed findings in CVT. The diagnosis of DCVT is even more puzzling because of the non-specific presentation e.g. Short history of changes in mental status, aphasia, seizures, paroxysmal sympathetic hyper-activity (PSHA), coma or death [10]. It is even more difficult to differentiate DCVT from causes of altered sensorium like metabolic encephalopathy or meningocencephalitis like in our case.

Paroxysmal sympathetic hyperactivity has been mainly reported after traumatic brain injury, however, it can occur in other brain disorders like DCVT. The pathophysiology of this phenomenon is debated but probably results from dysfunction of the sympathetic centers in the diencephalon [11]. The patient under discussion has been presented with PSHA. Treatment with bromocriptine, baclofen, morphine and other agents with variable response [12].

The parenchymal changes in CVT result from increased venous pressure which leads to vasogenic edema, cytotoxic edema and hemorrhagic infarction. CT findings of DCVT often shows hypo-attenuation of bilateral thalami and basal ganglia and hyper-attenuations of the deep venous system like vein of Galen, vein of Rosenthal, internal cerebral veins and straight sinus. The MRI and MRV are more sensitive modalities for the diagnosis of DCVT [13]. Because the deep venous system drain thalamus, basal ganglia, corpus callosum, inferior frontal lobes and deep white matter of the parietal and temporal lobes, thrombosis in these deep veins causes vasogenic edema and ischemic changes which appear on MRI as hyperintensity on T2-weighted Figures (T2-WI) and hypointense on T1-weighted Figures (T1-WI). Based on MRI signal intensity on T1 and T2-weighted Figures, DCVT can be divided into; (Figures-1, 2, 3, 4)

1. Initial stage (1-5 days) – absence of flow in the vein which appear as isointense on T1-WI and hypo intense on T2-WI.
2. Intermediate stage (5-14 days) - absence of flow with hyper-intensity on both T1-WI and T-2 W1
3. Late stage (>2 weeks) - recanalization of the vessel occurs with resumption of flow in the abnormal canalized vein [14].

The finding on the MRI resembles Japanese encephalitis (JE), Wilson disease, Wernicke’s encephalopathy, hypoxic ischemic encephalopathy and auto-immune encephalopathy like multiple sclerosis [15]. However, the edema in DCVT is vasogenic and the hemorrhagic transformation appears hyper-intense on T1-WI. The MRV can confirm the presence of thrombus and the absence of flow in the deep cerebral veins [13].

The CSF analysis in CVT shows increased protein level with lymphocytic pleocytosis which mimics the CSF picture of viral meningo-encephalitis [15]. The negative test for viral antigen in the CSF along with the MRI Figures help to confirm the diagnosis of DCVT.

Any delay in the diagnosis of DCVT can be lethal and increases chance of the patient dying. Anticoagulation should be started immediately and if the patient develops signs of raised intra-cranial pressure, measure to combat edema of the brain along with surgical decompression is recommended.

The investigations for thrombophilia should be delayed for 4-6 weeks as acute thrombosis affects the level of protein C and S, and anti-thrombin III levels [16]. The recommended treatment for DCVT is initiation of heparin or LMWH along with oral Coumadin to keep international normalization ratio (INR) 2-3. This treatment should be continues for 3-6 months or as per the underlying precipitating cause of the DCVT. In cases of severe thrombophilia as in protein C and S deficiency, anti-thrombin-III mutation or anti-phospholipid antibodies syndrome the anticoagulation can be extended to 1 year or life-long.
Conclusion

The case under discussion is presented with DCVT (thrombosis of vein of Galen) with altered mental status, seizures, PSHA, and features resembling viral encephalitis on CSF examination. Because of the proximity of the vein of Galen to the foramen of Monro and the secondary hemorrhagic changes from the underlying DCVT and AVM, his course is complicated by intra-ventricular hemorrhage and malfunction of the VP shunt, respiratory failure requiring tracheostomy, and protracted hospital stay. Eventually, the patient responded well to the treatment with baclofen and bromocriptine for his PSHA and he made a good functional recovery.

References


Citation: Awad Magbri (2018) Paroxysmal Sympathetic Hyperactivity Due to Thrombosis of the Vein of Galen: A Significant Clinical Imitator. SF J Neurosci 2:1.