

Case Report

Thrombosis of The Anterior Third of the Superior Sagittal Sinus Causing Unilateral Infarct in A Patient with Thrombocytosis and Thalassemia

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Abstract

The anterior third of the Superior Sagittal Sinus (AT-SSS) is often hypoplastic. Traditional neurosurgical teaching is that the AT-SSS can be safely ligated and excised. We report a case of thrombosis of the AT-SSS with unilateral venous infarct from thrombocytosis. A 47-year-old woman with history of anemia, splenectomy and deep vein thrombosis presented with confusion, decreased ability to speak, and transient right hand shaking. Neurological exam revealed disorientation to situation, impaired comprehension and speech production, and short-term memory loss. Laboratory studies were significant for microcytic anemia and thrombocytosis. Molecular analysis of hemoglobin showed delta-beta thalassemia. CT head and MRI brain showed venous infarct in the left frontal lobe. MRV and Digital subtraction angiogram revealed thrombosis of the AT-SSS and poor venous collaterals in the left frontal region. Contrary to traditional teaching, our case report suggests that occlusion of the AT-SSS increases the risk of venous infarct in the setting of poor venous collaterals. Thrombocytosis after splenectomy is a risk factor for cerebral venous thrombosis.

Keywords: Cerebral Venous Infarct; Cerebral Venous Thrombosis; Delta Beta Thalassemia; Superior Sagittal Sinus; Thrombocytosis

Introduction

Cerebral venous thrombosis (CVT) may cause serious strokes with diverse clinical and radiological presentations [1]. Due to atypical clinical features and significant anatomic variability in venous sinuses, the diagnosis of CVT is often elusive [2, 3]. The most common etiology is hypercoagulability and the standard treatment is anticoagulation [1]. Thrombocytosis in patients with thalassemia is a risk factor for venous thromboembolism (VTE), particularly in patients with splenectomy [4-7]. In this study, we describe a case of unilateral venous infarct from CVT that occurred in the anterior third of the superior sagittal sinus (AT-SSS) in a patient with thalassemia and splenectomy.

Case Report

A 47-year-old right handed Asian woman presented to an outside hospital with confusion and decreased ability to speak for a week. She was a poor historian. The history was provided by her daughter. Briefly, the patient was in her usual state of health until late October when she was not her normal self. She was speaking less and stated that she was not feeling well. She was less interactive and would often stare off into space throughout the day. She also had abnormal behaviours such as leaving home without her shoes or purse. She was noted to have transient right hand shaking while driving. She had no appetite, energy or emotions. She denied headache, nausea, vomiting, vision changes, weakness, numbness, leg swelling, or recent trauma. CT head and MRI brain at outside hospital showed a left frontal hemorrhagic lesion with vasogenic and cytotoxic edema, as well as an 8-millimeter left-to-right midline shift (Figure 1 (A-H)).

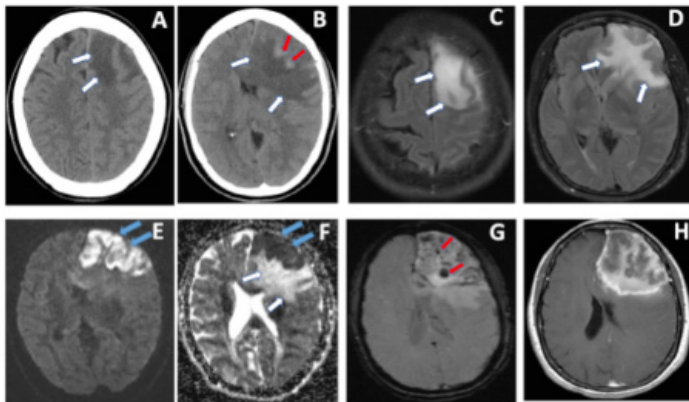


Figure 1 (A-H): Imaging of left frontal venous infarct. Initial CT head showed a left frontal hemorrhagic lesion (red arrows in B) with significant cerebral edema (white arrows in A & B). MRI brain revealed a left frontal lesion with mixed vasogenic (white arrows in C-F) and cytotoxic edema (blue arrows in E & F), as well as microhemorrhages (red arrows in G) without evidence of superimposed enhancement (H).

She was told she had a brain tumor and treated with intravenous Decadron and Keppra prior to transfer to our medical center for further evaluation. Her past medical history was notable for anemia, splenectomy and deep vein thrombosis. She was unsure of the reason for her splenectomy, but it was likely performed due to anemia that required frequent blood transfusions. Family history was significant for anemia, liver disease, and stroke. She worked as a nail technician and had recently returned from travel to another state visiting with family. Neurological examination revealed normal level of consciousness. She was oriented to name, place and date, but disoriented to situation. When asked why she was in the hospital, she answered “because I need to be here”. She had limited speech output. Her naming, repetition and comprehension were impaired. Her registration was 3/3 but recall was 0/3 at 5 minutes. There were no focal deficits in cranial nerve, motor and sensory examination. Her deep tendon reflexes were symmetric. Gait and coordination were normal.

Diagnostic Workup

Laboratory studies were significant for microcytic anemia (hemoglobin 8.8 grams/deciliter, mean corpuscular volume 66 femtoliters/cell), leukocytosis (white blood cell count 18000 / microliter blood) and thrombocytosis (platelet count 661,000 / microliter). Pregnancy test, coagulation panel, thrombophilia panel, and antiphospholipid panel were negative. Blood cultures were negative for bacteremia. Cerebral spinal fluid studies showed no evidence of infection or malignancy. Electroencephalogram and echocardiogram were unremarkable. CT chest/abdomen/pelvis revealed hepatomegaly, splenectomy, and a left renal cyst without evidence of malignancy. MRV showed a filling defect at the anterior

third of the superior sagittal sinus (AT-SSS) (arrows in (Figure 2 A, B)). Digital Subtraction Angiogram (DSA) with right ICA injection showed occlusion of the AT-SSS with significant collateral venous draining into the patent SSS and the sphenoparietal sinus/sylvian vein (blue and red arrows, respectively (Figure 2 C)). In contrast, DSA with left ICA injection showed poor collateral formation into the patent SSS and the sphenoparietal sinus/sylvian vein (blue and red arrows, respectively (Figure 2 D)).

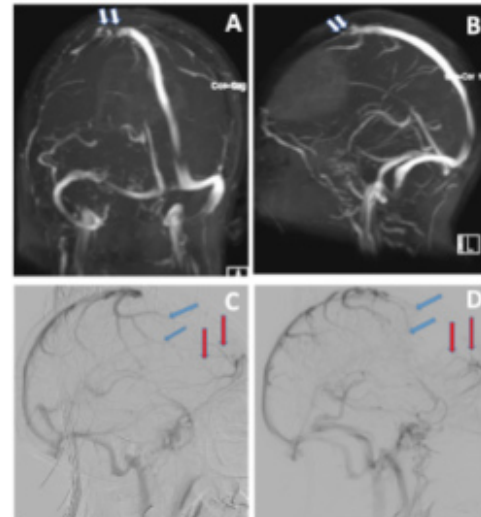


Figure 2 A-D: Neuroimaging of cerebral sinus thrombosis. MRV showed filling defect in the AT-SSS (arrows in A & B) that is consistent with sinus thrombosis. DSA with right ICA injection showed occlusion of the AT-SSS with significant collateral draining into the patent SSS (blue arrows in C) and the sphenoparietal sinus/sylvian vein (red arrow in C). In contrast, DSA with left ICA injection showed poor collateral formation into the SSS (blue arrow in D) and the sphenoparietal sinus/sylvian vein (red arrow in D).

Diagnostic Considerations

The preliminary workup was concerning for a venous infarct. However, the MRV and DSA findings may also be seen in cases of a brain mass compressing the sinus and in congenital variants of SSS. Due to atypical imaging findings and lack of evidence for a hypercoagulable state, she was not treated with anticoagulation. She improved with steroids and Keppra with some impulsivity and disinhibition. Molecular analysis of hemoglobin showed 8.1% Hgb F and 91% Hgb A, suggestive of delta-beta thalassemia.

Follow-up and Outcomes

Due to persistent thrombocytosis, she was treated with Aspirin 81 mg daily for prevention of recurrent CVT. Repeat MRI brain at 2-month follow up showed significant decrease of cerebral edema, mass effect and midline shift, and recanalization of the AT-

SSS (arrows in (Figure 3 A-D)). At one-year follow up, she was symptom-free without recurrent events.

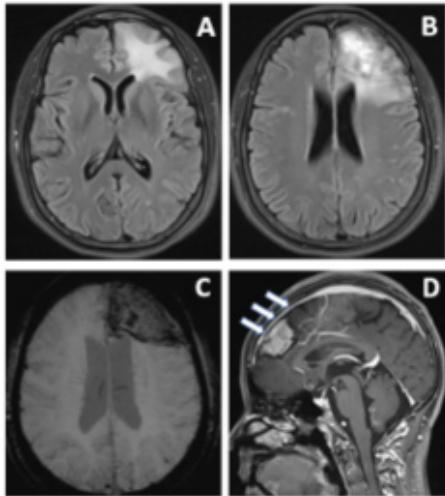


Figure 3 A-D: Follow-up Images. MRI brain at 2-month follow up showed significant decrease in cerebral edema and mass effect (A, B, C) and recanalization of the AT-SSS (arrows in D).

Discussion

This case highlights the diagnostic challenges of CVT. Vascular imaging showed a hemorrhagic lesion with cytotoxic and vasogenic edema. MRV and DSA showed filling defect of the AT-SSS, suggestive of a brain mass compressing the AT-SSS, an occlusive thrombus, or congenital sinus atresia [2]. Traditional neurosurgical teaching is that the AT-SSS can be safely excised without risk of venous infarct. This was initially described by Dr. Cushing in his surgical excision of parasagittal meningioma. The anterior third of the sinus corresponds to roughly the Rolandic point beyond which he did not suggest resection [3]. In this patient, bilateral anatomic variation in venous collateral circulation might have contributed to unilateral venous infarct from AT-SSS thrombosis. The patient had excellent venous collaterals on the right side and therefore she only developed venous infarct in the left frontal lobe. Follow up MRI showed interval improvement in cerebral edema and recanalization of the AT-SSS.

Clinical presentation was also atypical. The patient presented with progressive symptoms without headache. However, she did have significant thrombocytosis and subsequent workup revealed delta-beta thalassemia. VTE occurs in patients with thalassemia due, in part, to increased platelet activation, increased circulating abnormal red blood cells (RBCs) and endothelial cell activation [4-6]. This is particularly apparent in patients of a moderate clinical phenotype who do not require regular blood transfusions. Blood transfusions reduce the proportion of abnormal circulating RBCs and may replete anti-coagulant factors [4-7]. Following

splenectomy, risks of VTE are further increased (22.5% vs. 3.5% in non-splenectomized patients) due to worsened thrombocytosis and platelet aggregation [4].

Cerebral thromboembolism occurs in about 1.13% of patients with thalassemia, and is associated with increasing age, transfusion naivety, splenectomy, and thrombocytosis [7]. Silent ischemic injury has been noted in splenectomized patients who have platelet counts more than 500,000/mL and receive irregular blood transfusions [7]. Acute CVT with hemorrhagic infarct can be treated with anticoagulation. However, due to atypical imaging findings and lack of evidence for a hypercoagulable state, our patient was not anticoagulated during hospitalization. Aspirin was started based on the report of antiplatelet therapy for prevention of recurrent VTE in splenectomized patients with platelet counts higher than 500,000/L [6].

Conclusion

CVT may occur in patient with thalassemia, especially in the setting of splenectomy and thrombocytosis. Unilateral venous infarcts may occur in the setting of thrombosis of the AT-SSS with poor venous collaterals and the safety of ligation of the AT-SSS may be questionable in these patients.

Disclosures

The authors do not have any relevant disclosures.

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