Post-streptococcal central nervous system vasculitis

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CASE DESCRIPTION

A 5-year-old previously healthy female presented to the hospital after an episode of headache and acute right-sided weakness. One week prior, the patient had a fever and upper respiratory symptoms. Initial imaging revealed a left basal ganglia infarct with an occlusion of the left A1 and multiple stenoses of left M1 and M2 segments suggestive of vasculitis. CSF studies, hypercoagulable, and autoimmune work-up were unremarkable except for an elevated ANA. The patient was transferred to pediatric rehabilitation service for further care and unfortunately experienced another episode of headache, generalized weakness, and decreased responsiveness. Cerebral angiogram showed diffuse left ICA, ACA, and MCA arteriopathy. Additional testing revealed elevated DNase-B antibody. She was started on high dose steroids and rituximab for possible post-streptococcal cerebral vasculitis. During the outpatient rehab follow-up visit, the patient exhibited new onset spasticity most prominent in her right upper extremity for which she was started on baclofen with improvement of her symptoms.

DISCUSSION

Secondary cerebral vasculitis in children is well-described in literature. Common causes of secondary central nervous system (CNS) vasculitis in children include infections, rheumatological diseases, and other inflammatory diseases. It is thought that a CNS infection (i.e. bacterial, viral, or fungal meningitis), much like rheumatic diseases, causes vessel wall inflammation which leads to vascular stenosis, endothelial activation, and clot formation.[1]

Particularly, cerebral vasculitis associated with post-streptococcal infections, while a rare complication, has been mostly associated with post-streptococcal glomerulonephritis, which the patient did not present with.[2-4] To our knowledge, this is the first case of isolated CNS vasculitis after a streptococcal infection. In this case, intracerebral vasculitis led to an arterial ischemic stroke (AIS), a rare occurrence that is one of the leading causes of pediatric deaths. AIS most commonly presents with focal or diffuse neurological deficits or seizures. Risk factors for developing AIS include arteriopathy, cardiac disorders, prothrombotic disorders, or acute illness including infection. While the rate of common pediatric infections leading to AIS is poorly established, it is pertinent to consider this unfavorable sequela.

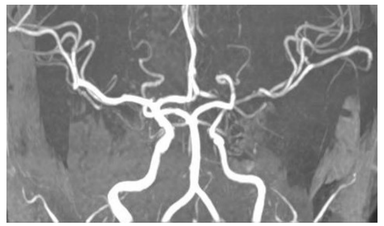

Figure 1: Images from MRA Brain showing evidence of diminished left-sided cerebral blood flow

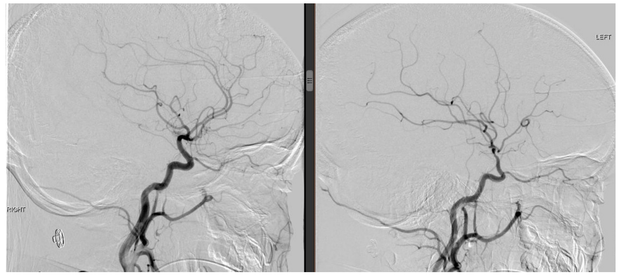

Figure 2: Images from cerebral angiogram showing diffuse arteriopathy of left-sided anterior circulation

CLINICAL SIGNIFICANCE

Cerebral vasculitis associated with streptococcal infection, although rare, should be considered in children presenting with stroke-like symptoms after URI symptoms. Timely initiation of acute rehabilitation and close outpatient rehab follow-up to monitor for complications can improve overall functional outcomes.

REFERENCES

1. Gowdie P, Twilt M, Benseler SM. Primary and secondary central nervous system vasculitis. *J Child Neurol*. 2012;27(11):1448-1459.
2. Wong W, Morris MC. Cerebral vasculitis in a child following post-streptococcal glomerulonephritis. J Paediatr Child Health. 2001;37(6):597-9.
3. Rovang RD, Zawada ET, Santella RN, Jaqua RA, Boice JL, Welter RL. Cerebral vasculitis associated with acute post-streptococcal glomerulonephritis. Am J Nephrol. 1997;17(1):89-92.
4. Deveber GA, Kirton A, Booth FA, et al. Epidemiology and Outcomes of Arterial Ischemic Stroke in Children: The Canadian Pediatric Ischemic Stroke Registry. Pediatr Neurol. 2017;69:58-70.