Acute posterior circulation infarct due to bicuspid aortic valve vegetation: An uncommon stroke mechanism

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Abstract

Acute ischemic stroke may be the first clinical manifestation of the underlying cardioembolic source. We are reporting a 28-year-old man presenting with acute posterior circulation infarct due to underlying bicuspid aortic valve disease with vegetation detected by transesophageal echocardiography in the absence of clinical features of heart disease and infective endocarditis. The case report highlights the importance of routine evaluation of cardioembolic sources in all cases of ischemic stroke.

Key Words

Bicuspid aortic valve vegetation, posterior circulation infarct, stroke in young

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Introduction

A significant number of strokes occur due to cardioembolic sources.[1] However, some of these patients may not have clinical features referable to the cardiovascular system. A high index of suspicion and routine cardiac evaluation in all patients with acute ischemic stroke is necessary to establish the underlying cardiac source of embolism. We are reporting a case of acute posterior circulation infarct thought to be of undetermined etiology, till Bicuspid aortic valve (BAV) with valvular vegetation was documented on transesophageal echocardiography (TEE).

Case Report

A 28-year-old man presented with acute weakness of the right upper limb and lower limb associated with one episode of generalized tonic–clonic seizure. The patient had a history of fever 2 weeks before admission to this hospital, for which he was treated in the outlying hospital. On general examination, the patient was afebrile. There was no clubbing, cyanosis or splinter hemorrhages on the nails. Pulse was 76/min, regular, and the blood pressure was 140/80 mmHg. There was no hepatomegaly or splenomegaly. Cardiac examination revealed a basal short systolic murmur. On neurological examination, the patient was conscious and well oriented in time place and person. Cranial nerves were normal. He had triparesis comprising power of 3/5 in the left upper and lower limbs, and power of 2/5 in the right lower limb with reduced deep tendon reflexes and extensor plantar on the left side. There were no sensory or cerebellar signs. Magnetic resonance imaging showed bilateral parasagittal parietal hemorrhagic infarcts [Figure 1a], infarcts in splenium of corpus callosum and left thalamus [Figure 1b] and right cerebellar hemisphere [Figure 1c]. Magnetic resonance venography was normal. Transthoracic echocardiogram (TTE) showed bicuspid aortic valve with mild regurgitation, moderate aortic sclerosis and moderate concentric left ventricular hypertrophy. TEE showed vegetation attached to the anterior commissure in the 5 'O' clock position [Figure 2]. His vasculitic workup was negative. Serum homocysteine, antinuclear antibody, anticardiolipin antibodies, lipid profile, protein C and S and antithrombin III levels were normal. Magnetic resonance angiography was normal. Blood cultures were sterile after incubation for 48 h. The patient was treated with intravenous Ceftriaxone 2 grams IV od for 4 weeks, of which he had received 2 weeks therapy before coming to our hospital. In addition, he received intravenous Gentamycin 3 mg/kg od for 2 weeks. This treatment regimen is in accordance with the most recent ACC/AHA guidelines, which covers most of the organisms (*Streptococcus viridens* and *S. bovis*, Enterococci and *Staphylococcus aureas*) that cause infective endocarditis (IE).[1]

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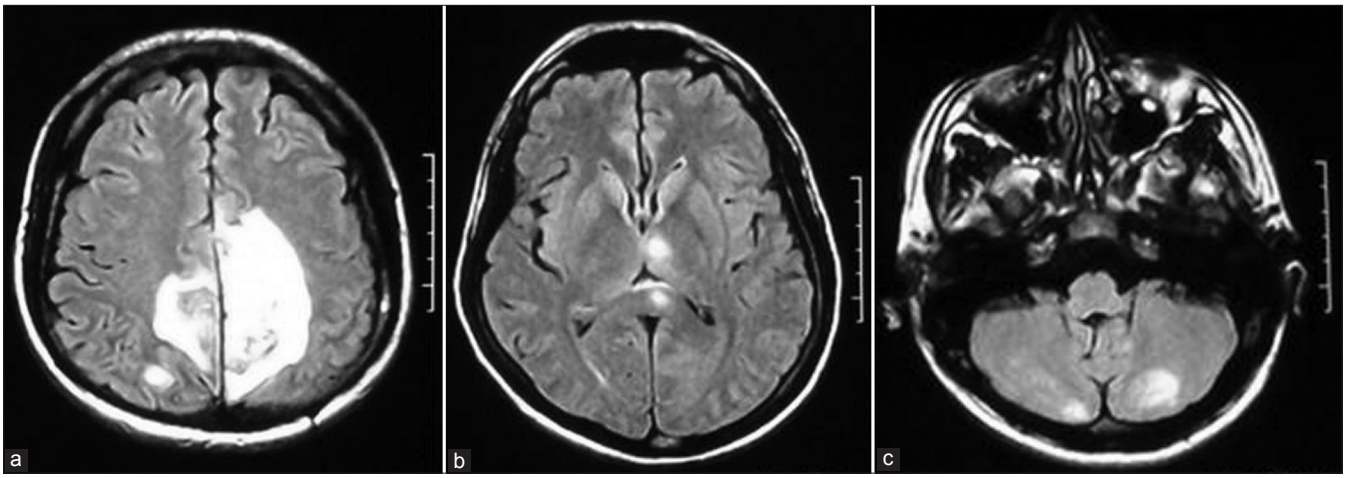

Figure 1: (a) Magnetic resonance imaging (MRI) (Flair) showing bilateral parasagittal parietal hemorrhagic infarcts. (b) MRI of the brain (Flair) showing infarcts in the splenium of corpus callosum and left thalamus. (c) MRI (Flair) showing infarcts in both the cerebellar hemispheres

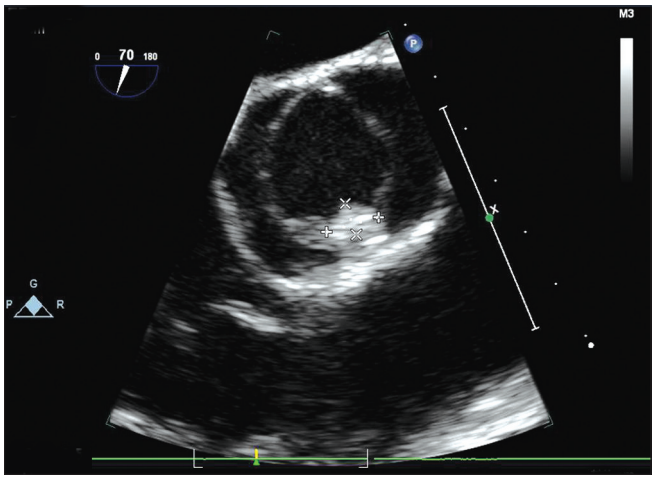

Figure 2: Transesophageal echocardiography showing a vegetation attached to the anterior commissure in the 5 "O" clock position

The patient had significant recovery of his weakness and his power improved to 4/5 in the affected limbs, bringing his modified Rankin's score to 1 and Barthel score to 90 at the time of discharge.

Discussion

BAV is the most common congenital heart disease occurring in 1-2% of the population, but most patients remain asymptomatic and therefore go undetected, without clinical consequences, for their lifetime.[2] BAV is the most common congenital cardiac abnormality and may result from defects in the genes that encode matrix elements. It is the result of abnormal aortic cusp formation during valvulogenesis. Adjacent cusps fuse to form a single aberrant cusp, larger than its counterpart yet smaller than two normal cusps. BAV malformations are inherited, particularly in males. BAV is a disease of the entire aortic root and has a propensity for both valvular and aortic complications, often requiring surgery; endocarditis is a devastating complication that can be prevented by antibiotic prophylaxis.[3] Embolism has been considered to be a much less common occurrence in patients with aortic valve disease compared with those with mitral valve disease, but is likely to occur in patients with IE superimposed upon bicuspid valve.[4] However, embolism can occur without IE, as reported recently in a 42-year-old case of BAV with recurrent posterior circulatory ischemic strokes resulting in ataxia and cognitive impairment without the evidence of endocarditis. TEE demonstrated a BAV with mild stenosis, moderate calcification and a dilated ascending aorta without vegetations. The degree of calcification and the valve phenotype might be important factors implicating the BAV as a rare cause of ischemic stroke even in the absence of endocarditis.[5] In view of the history of fever, our patient had probable IE, the clinical manifestations of which were suppressed due to antibiotic treatment. IE is known to have neurological manifestations in 30-40% of patients, and in about 15-20% of patients systemic manifestations like clubbing, splinter hemorrhages and hematuria may be absent.[6,7] In such cases, blood culture is helpful. However, if blood culture is noncontributory, then the diagnosis relies on the demonstration of vegetations on echocardiography. TTE sometimes may not be able to pick up the underlying vegetation. TEE allows visualization of smaller vegetations, and has been shown to be significantly more sensitive (85-95%) than TTE and highly specific in both confirming the clinical diagnosis of IE as well as in identifying valvular vegetations in patients at risk for this infection. TEE also achieves better resolution of the image.[8,9] Thickened valves, ruptured chordae or valves, valve calcification and nodules may be mistaken for vegetations, indicating the specificity limitations of isolated echocardiography.[10]

Conclusion

A cardiac source of embolism should always be considered in all patients of acute ischemic stroke, particularly when there is no other obvious etiology to explain the stroke mechanism. High clinical suspicion coupled with TEE is helpful in detecting the vegetations even when the other systemic manifestations are missing.

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