

Case Report

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Acute ischemic stroke in a caucasian male with long chiari network, interatrial septal aneurysm and patent foramen ovale: A case report

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Case presentation

A 49-year-old Caucasian male, left-handed, with no known comorbidities, working as a middle school teacher, presented to the emergency room with a sudden-onset left hand weakness described as difficulty gripping with no other associated symptoms. A non-smoker, non-alcoholic beverage drinker, with no illicit drug use. There was no significant history of any neurological disease in the family background but his father has a history of interatrial septal aneurysm. Physical, cardiologic and neurologic examination at the emergency department were unremarkable aside from the weakness on his left hand and a mild sensory deficit on the left upper extremity. A cranial magnetic resonance imaging with magnetic resonance angiography was done which showed a hyperacute infarct in the right frontal lobe, with paucity of vessels and attenuation of flow-related signals in the M4 perforators of the right middle cerebral artery and contour irregularities along the intradural segment of the right vertebral artery relating to atherosclerotic changes (Figure 1). Patient was given a recombinant tissue plasminogen activator since he came in at the 3rd hour post-ictus. Laboratory tests

done at the emergency department showed normal complete blood count, routine blood chemistry, A further work-up was done which showed a slightly elevated LDL on Lipid profile, and other parameters within normal limit.

The contrast-enhanced transesophageal echocardiography had an ejection fraction of 59% and showed a large filamentous Chiari network. Intermittent separation of the septum primum and septum secundum creating a tunnel-like space at the interatrial septum measuring 0.31 cm that leads to a small predominantly bidirectional shunting consistent with functionally patent foramen ovale. Contrast echo was done revealing full opacification of the right atrium and right ventricle with passage of contrast seen on the left atrium, indicative of positive bubble contrast study. Interatrial septum aneurysm also noted (Figure 2).

Patient was then referred to Cardiovascular Thoracic Surgery service and was given the treatment option of PFO closure. Patient eventually underwent open Patent Foramen Ovale closure with Bipore Graft and resection of Chiari network on his 14th

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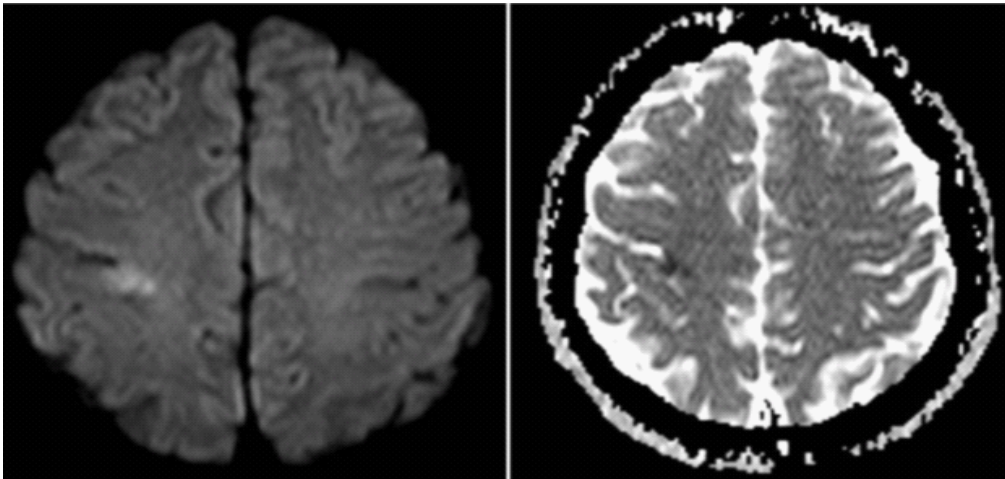


Figure 1: Axial cuts of cranial MRI without contrast showing an area of restricted diffusion on DWI with corresponding signal drop on ADC at the right frontal lobe.

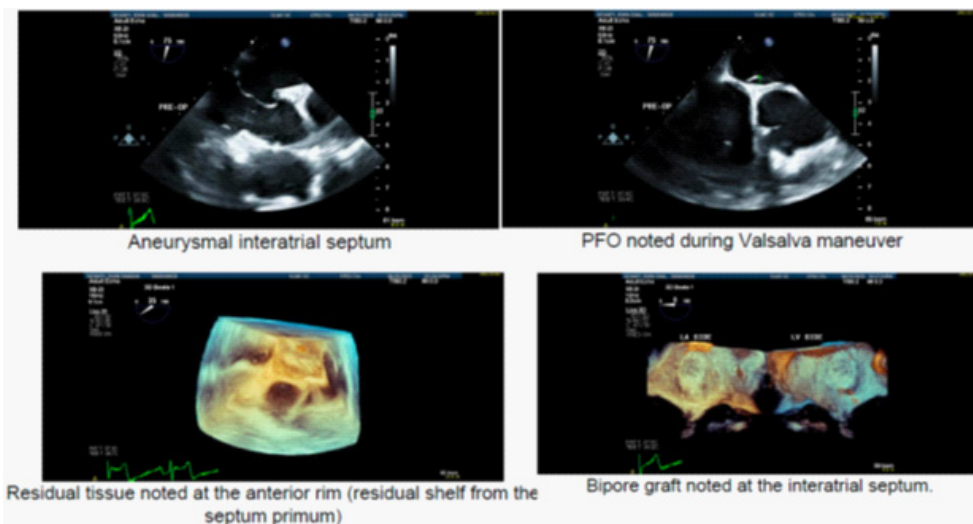


Figure 2: Transesophageal echocardiography findings preoperative (above) and postoperative (below).

day post-ictus with no untoward events noted intraoperatively and post-operatively. A postoperative transesophageal echocardiography was done revealing an ejection fraction of 69% with the presence of a Bipore graft at the interatrial septum. Aneurysmal interatrial septum is no longer seen and no residual shunt is observed between the atria. After the procedure, the patient was maintained on Aspirin 80 mg per day and had neither recurrence nor worsening of symptoms, his left hand grip was back to baseline and had no sensory deficit.

Discussion

Patent foramen ovale (PFO) has been associated with cryptogenic stroke, however, there is conflicting data and it remains uncertain whether PFO is the direct cause, a risk factor or an incidental finding. The mechanism by which PFO causes stroke is uncertain but paradoxical embolism is the most acceptable hypothesis to date. PFO size is an independent risk factor for stroke occurrence and recurrence when the size is ≥ 2 mm. The diagnosis of a PFO in terms of clinical significance entails both anatomic and physiologic aspects. The physiologic assessment of a right-to-left shunt (RLS) is usually performed using contrast transthoracic echocardiography (TTE) or Transcranial Doppler (TCD) before an anatomic confirmation is demonstrated, usu-

ally by transesophageal echocardiography (TEE). Each of these 3 ultrasound modalities has advantages and disadvantages in the workup of a stroke patient, but equivalent in a sense that they all detect right-to-left shunt by microbubbles, either visually (TTE and TEE) or by Doppler shift (TCD). Therefore, it can be used in diagnosing the presence of PFO.

Atrial septum aneurysm on the other hand is associated with increased stroke risk, but is only present in 1-4% of the general population. It is associated with higher risk of stroke particularly if co-existent with PFO, in fact a stronger risk factor than PFO. It has been recognized as an independent predictor of embolism recurrence, and the risk increases by two to three times when it coexists with PFO. In relation to this, the recurrence rate for stroke or TIA within 4 years after the initial event was 19.2% for PFO combined with atrial septal aneurysm versus 5.6% for PFO alone.

Chiari's network are fetal features that interfere with the normal embryonic R-L shunt but this anatomic variation is believed to be the mostly clinical in apparent. Although often an incidental finding, the Chiari network has been shown to be associated with PFO and atrial septal aneurysm. The proposed mechanism for this phenomenon is that the Chiari network results in an

embryonic right atrial flow pattern, which directs blood toward the interatrial septum, leading to an increased likelihood of aneurysm or PFO development. In addition to this, the Chiari network has also been posited as a source of thrombi and emboli. Hence, the combined presence of PFO with Chiari network and interatrial septum aneurysm increases an individual's risk of developing an ischemic stroke.

In a retrospective study done by Nakayama et. al wherein they evaluated patent foramen ovale characteristics by trans-esophageal echocardiography and they projected the risk score of having a stroke based on high-risk features of PFO. They found out that large-size PFO (≥ 2 mm in height), long-tunnel PFO (≥ 10 mm in length), the presence of hypermobile interatrial septum, the presence of prominent Eustachian valve or Chiari's network, the large R-L shunt during Valsalva maneuver, and low-angle PFO were independent risk factors related to stroke, hence, presence of more than two of these features are seen on TEE, it adds up to the risk of having an ischemic stroke.

The Randomized Evaluation of Recurrent Stroke Comparing PFO Closure to Established Current Standard of Care Treatment (RESPECT) trial performed at 69 sites in the United States and Canada revealed that PFO closure had a lower rate of recurrent ischemic strokes than medical therapy alone, however, it was associated with a higher rate of venous thromboembolism than medical therapy alone. In another study, Patent Foramen Ovale Closure or Anticoagulants versus Antiplatelet Therapy to Prevent Stroke Recurrence (CLOSE) trial which compared transcatheter closure of PFO plus long-term antiplatelet therapy versus antiplatelet therapy alone on stroke recurrence in a 5-year analysis revealed that the rate of recurrent stroke was significantly lower with closure of the PFO plus long term antiplatelet therapy than with anti-platelet therapy alone. These studies then suggest that closure of patent foramen ovale lessens the risk of recurrence of stroke. In relation to the patient, since he has an interatrial septum aneurysm, long Chiari network and a patent foramen ovale, which poses a higher risk for him to develop recurrent strokes, surgical intervention combined with antiplatelet therapy would be beneficial.

Conclusion

In conclusion, presence of high-risk features of patent foramen ovale poses a high risk of an ischemic stroke, which might warrant investigation in patients suspected to have PFO or those who have a familial history of PFO. The recent trials have shown that PFO closure is superior to medical treatment in the secondary prevention of ischemic stroke. The benefit in the reduction of stroke recurrence is particularly evident in patients who have documentation of a PFO with high-risk characteristics.

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