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Criptogenetic Cerebral Ischemia and Spongious Atrial Septum. A New Culprit?

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Abstract

A case of unusual atrial septal anatomy with right-to-left shunting in a patient with cryptogenetic cerebral ischemia is presented. Percutaneous correction and technical tips are described.

Introduction

The appropriate treatment strategy for secondary stroke prevention in patients with cryptogenic stroke and Patent Foramen Ovale (PFO) remains challenging. The clinical and anatomic variables reported to be risk factors associated with stroke recurrence include older age [1], large PFO [2-5] and large right-to-left shunting [6] and combined atrial septal aneurysm [7,8].

Patent Forame Ovale (PFO) is an anatomical variant of the atrial septum, providing communication between the atrial chambers of the heart through fossa ovalis on the right side and the ostium secundum on the left side. Septum primum acts as a one-way valve allowing blood flow from the right to left atria, bypassing the lungs. This septum normally remains patent before birth and closes with the first breath of air that a baby takes because of increased left sided pressures [9].

Autopsy studies have shown an overall prevalence of 27% in the general population, decreasing with increasing age (35% and 20% in age groups less than 30 years and more than 80 years, respectively) [10,11]. This could be secondary to either selective mortality of patients with PFO with age or late spontaneous closure.

Transthoracic Echocardiography (TTE) diagnosis requires detection of a shunt with either color Doppler or agitated saline contrast with Valsalva maneuver, and its prevalence in the population varies anywhere between 10% to 22% with this technique [12-15].

Trans-Cranial Doppler (TCD) provides the possibility to show the specific cerebral flow disturbance in case of right to left shunt during Valsalva maneuver. It confirms PFO (patho) physiology significance [16,17].

This report deals with a patient with trans-cranial Doppler positivity and non-apparent PFO with initial impossibility to cross the atrial septum during PFO percutaneous closure attempts.

Case Report

A 49 year old Caucasian male came to our Outpatient Clinic after several transient ischemic neurological attacks without any evidence of Carotid, aortic or cardiac embolic disease. Hypercoagulability screening tests were completely negative.

TTE had confirmed the presence of right to left shunt at atrial septum level, according to a positivity of TCD with curtain effect during Valsalva maneveur. Patient was thus scheduled for PFO percutaneous closure.

The procedure was performed during general anesthesia, oro-tracheal intubation, and Trans-Esophageal Echo (TEE) monitoring. Atrial septum showed some grade of mixoid degeneration with increased thickness (4 mm) (Figure 1). Surprisingly, no evidence of septal defects and/or PFO was shown, as neither a 5 F MP catheter nor the common angiographic wires (0.021 to 0.035") were able to cross the septum in spite of several and accurate attempts.

However, echo-contrastography (agitated saline solution from the venous femoral route) during Valsalva maneveur showed right to left passage of multiple swirling contrast jet through the septal wall (Figures 2 and 4). This unusual pattern of right to left shunting suggested the possibility to engage a small wire into and through the septum.

In fact, after multiple attempts, the septum was crossed by a 300 cm long 0,014" coronary medium weight guidewire followed from a 5F MP catheter.

Following the left atrium selective catheterization, a high support angiographic 0.035" wire was left into the left superior pulmonary vein and a 9 F Mullins long sheath was easily inserted into the left atrium (Figure 3).

Then a 23/25 mm Figulla atrial septal occluder (Occlutech®) was used aiming to stent the major part of the septum without encroaching

Figure 1: Atrial septal mixoid degeneration.

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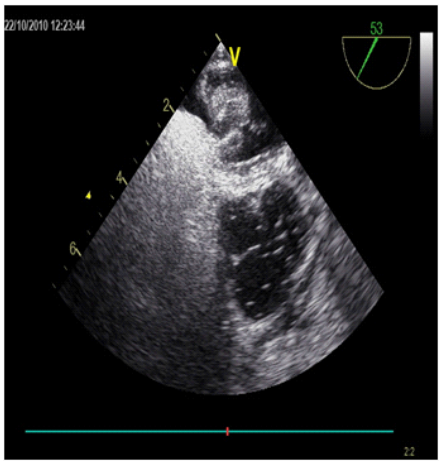


Figure 2: TEE: no PFO evidence and right to left shunting during Valsava.

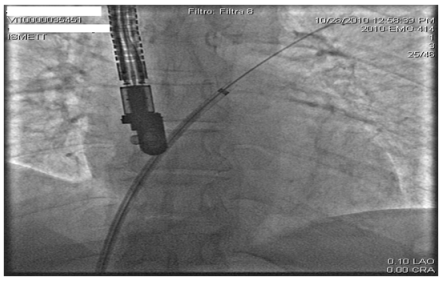


Figure 3: Hardware through the atrial septum.

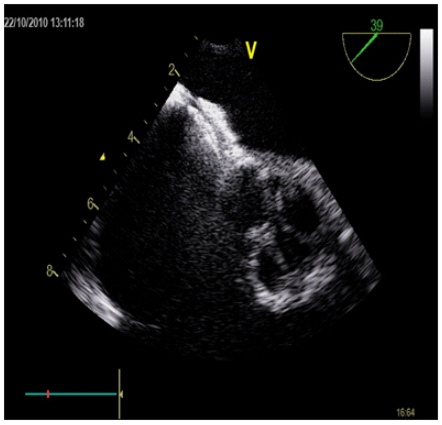
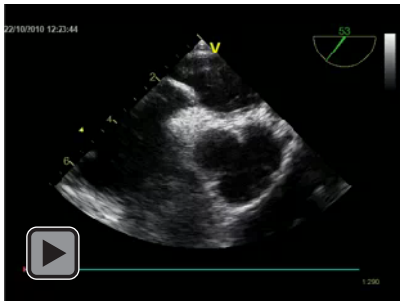


Figure 4: On site occluding atrial septal prosthesis.



Movie 1

the surrounding anatomic structures (Aorta, mitral valve, caval veins) (Figure 4).

After the septal occluder deployment, the right to left shunt during Valsalva maneveur rapidly disappeared with persistence of only minimal intra-prosthetic shunt.

Discussion

PFO initial screening is generally done with TTE using color Doppler and agitated saline contrast [13]. TCD with Valsalva and bubble test can also be used as an initial screning test (with higher sensitivity and lower specificity than TTE).

Administration of contrast through the femoral vein as opposed to the ante-cubital vein has shown to increase sensitivity of detection of PFO. This is due to the fact that blood from the superior vena cava is directed towards the tricuspid valve, while venous return from the inferior vena cava is directed towards the PFO [18,19].

During interventional approach, PFO is generally easily crossed by a common MP catheter or a 0.035" angiographic wire.

Sometimes, in cases of complex septal anatomy (multi-cribrosity, hypertrophic Eustachian valve, septal aneurysm) crossing the atrial septum may request multiple attempts under the guide of TEE.

In this reported case, no definite communication between right and left atrium through a patent foramen ovale and/or a (micro) septal defect was present. However, the atrial septum, that showed some thickening and looked non-homogeneous in the region of the fossa ovalis, let echocontrast pass from right to left through multiple sites during Valsalva maneveur, apparently behaving as a spongious reservoir.

There are no definite cases described in literature of such septal behavior, while some report of non-PFO embolic anatomy such as the so called pouch septum does exist [20,21].

In the described case, the atrial septum behave like a reservoir, ejecting blood from right to left as squeezed by the increased thoracic pressure (Valsalva). A mechanical approach with stenting of a wide part of the atrial septum was adopted in the hope to reduce the propensity to paradoxical shunting of blood in this patient.

A transeptal puncture was avoided as a thin coronary wire was able to pass from right to left, probably through intraseptal micro channels.

This anatomic variant was thus defined spongious micro-channeled atrial septum.

The patient is maintained under strict clinical (double oral antiplatelet therapy) and TTE and TCD follow up to confirm the favorable evolution of the septum stenting procedure.

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