CASE REPORT

An alien in the heart

Yashwant Agrawal [a,*], Jagadeesh K. Kalavakunta [b], Vishal Gupta [b]

[a] Department of Internal Medicine/Pediatrics, Western Michigan University Homer Stryker School of Medicine, Kalamazoo, MI
[b] Department of Cardiology, Michigan State University/Borgess Medical Center, Kalamazoo, MI

[a,b]USA

We report a case of a 38-year-old-man who presented with altered mental status. The patient was diagnosed with infective endocarditis (IE) originating from the GORE HELEX septal occluder device, which was placed 15 months earlier for symptomatic atrial septal defect. Brain imaging revealed shower emboli phenomena from the known IE. The patient developed hydrocephalus for which external ventriculostomy was performed. Improved neurological status warranted open heart surgery. The patient was later confirmed to be an intravenous drugs abuser, prejudicing IE. This case highlights the importance of meticulously monitoring patients with suspected high-risk behavior with an implanted intracardiac prosthetic device.

Keywords: Atrial septal defect, GORE HELEX septal occluder device, Infective endocarditis, Interatrial septum, Ventriculostomy

Introduction

We report the first case, to the best of our knowledge in the USA, of GORE HELEX septal occluder device (GORE-HSO) infective endocarditis (IE), with shower emboli to the brain requiring external ventriculostomy drainage, where the patient's intravenous drug abuse predisposed to the pathology described.

Case report

A 38-year-old man presented in altered mental status and acute respiratory failure for which he was emergently intubated. Vitals were significant for fever of 40.6 °C. Physical examination was remarkable for multifocal pustules on palms, fingers and arms. Laboratory results were significant for leukocytosis of 18.7 × 10^9/L, creatinine 2.5 mg/dL, troponin 0.74 ng/mL, and procalcitonin 60.76 ng/dL. Urine drug screen was positive for benzodiazepines and cannabinoids. The patient's family confirmed him being an intravenous drug abuser. Computed tomography of the brain revealed multiple infarcts. Blood cultures were drawn and empiric antibiotic therapy was commenced. Fifteen months previously, the diagnosis of symptomatic single defect atrial septal defect (ASD) was made when the patient presented with worsening exertional dyspnea, transient ischemia attacks, and cryptogenic stroke. This ASD was determined as the cause of his symptoms

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* Corresponding author at: Department of Internal Medicine/Pediatrics, Western Michigan University Homer Stryker School of Medicine, 1521 Shaffer Road, Borgess Medical Center, Kalamazoo, MI 49048, USA.
E-mail address: yashwantagrawal.agrawal@gmail.com (Y. Agrawal).

P.O. Box 2925 Riyadh – 11461KSA
Tel: +966 1 2520088 ext 40151
Fax: +966 1 2520718
Email: sha@sha.org.sa
URL: www.sha.org.sa

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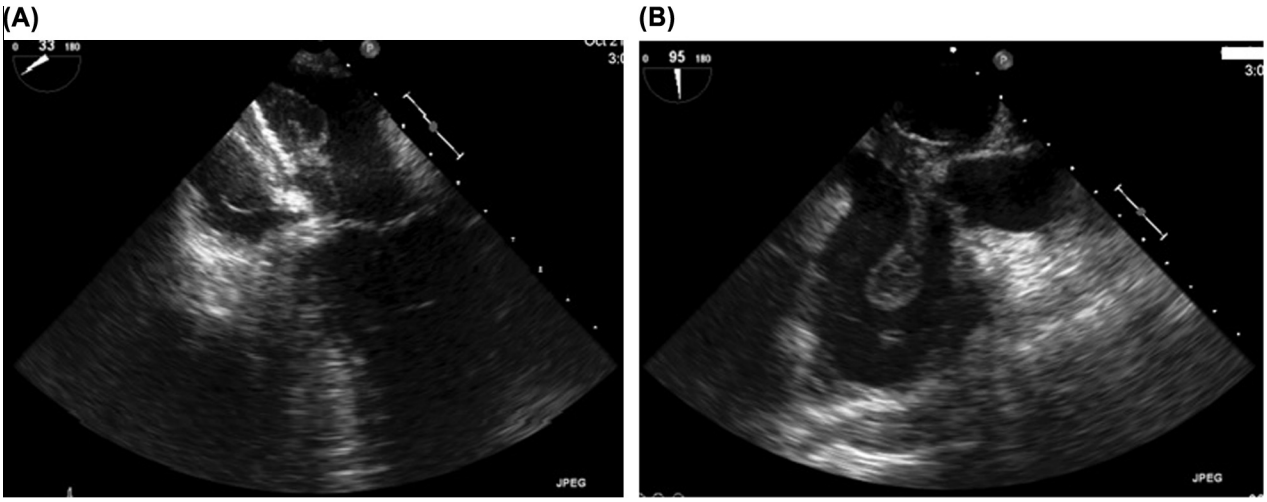

Figure 1. Transesophageal echocardiogram at the mid esophageal level (A at 33°, B at 95°) showing large, multilobulated and mobile echodensities (arrowheads) on either side of the GORE-HELEX septal occluder device (arrow) consistent with vegetations. LA = left atrium; LV = left ventricle; RA = right atrium.

prompting a 30-mm GORE-HSO to be implanted, which covered the entire defect.

Transesophageal echocardiography revealed a large, multilobulated mass and a small mobile mass on either side of the GORE-HSO, consistent with vegetation (Fig. 1). Magnetic resonance imaging of the brain demonstrated widespread small nonenhancing punctate acute ischemic infarcts and punctate parenchymal hyperintensities consistent with shower emboli from the IE. The patient's two blood cultures were positive for methicillin-resistant *Staphylococcus aureus*.

Asymmetric right pupillary dilation (7 mm vs. 5 mm) 2 days after admission warranted repeated computed tomography of the brain. It revealed new left temporal, occipital horn dilatation, and increased dilatation of the third and both lateral ventricles, indicating early hydrocephalus. Neurosurgery was consulted and external ventriculostomy drainage performed to relieve the intracranial pressure.

The patient's neurological and hemodynamic status improved over the next 3 weeks and he underwent open heart surgery. Intraoperatively, extensive scarring of the GORE-HSO with vegetations encompassing the device on both atrial sides and the entire atrial septum were appreciated. The GORE-HSO was removed through a right atrial approach and the infected interatrial septum was excised. The resultant large ASD was closed using the patient's autologous pericardium. The resected specimen cultures remained negative. He was treated for 6 weeks with vancomycin, ceftaroline, daptomycin, and rifampin. His postoperative course was unexceptional and he recovered well. The patient is asymptomatic with biannual follow-ups in the cardiology clinic.

Discussion

Secundum type ASD is a common congenital heart defect and newer imaging modalities have fashioned their percutaneous closure popular. The three commonly used devices are the Amplatzer (St. Jude Amplatzer, AGA Medical Corporation, Plymouth, MN, USA), the CardioSEAL-STARflex (NMT's CardioSEAL-STARflex, W.L. Gore & Associates, Inc., Flagstaff, AZ, USA), and the GORE-HSO (Helex Septal Occluder, W.L. Gore & Associates, Inc., Flagstaff, AZ, USA), with CardioSEAL withdrawn in October 2006 and ceased operations in 2011.

The first description of the GORE-HSO implantation was in 2001 [1] and it was approved by the United States Food and Drug Administration in 2006. A clinical trial of the GORE-HSO proved successful implantation in 96% of the patients with no reported IE [2]. The *study cohort* pooled clinical results on the feasibility, pivotal, continued access, and postapproval studies on the GORE-HSO. It demonstrated a composite clinical success of 93%, with no stated IE in this largest study of 435 patients [3]. A nonrandomized, multicenter trial of 119 patients who underwent HSO placement showed clinical success of 91.7% versus 83.7% who underwent surgery, with no described IE [4].

A case of GORE-HSO culture-negative IE has been reported in The Netherlands [5]. We report the first case, to the best of our knowledge, in

the USA with culture-positive IE of GORE-HSO with shower emboli to the brain causing hydrocephalus, requiring external ventriculostomy drainage and removal of the infected device.

Generally, 12 months of postprocedural antibiotic prophylaxis are recommended. Nonetheless, intravenous drug abuse renders any antibiotic prophylaxis inadequate and prejudices to progression of IE. The all-embracing infection could be explained from contiguous spread from the device, through the septum and to the left side of the septum as evidenced with the infected interatrial septum. Neither the patient nor his family reported intravenous drug usage before the GORE-HSO placement; moreover, the patient did not take any postprocedural antibiotics. Practitioners' vigilance in drug screening patients prior to intracardiac prosthetic device implantation can prevent such high morbidity and mortality complications.

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