Unruptured sinus of Valsalva aneurysm presenting with concurrent Morgagni hernia

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We describe a patient with dyspnea and intermittent cyanosis who was found to have concurrent right diaphragmatic and right atrial masses, initially thought to have advanced vascular sarcoma. She was ultimately diagnosed with an unruptured sinus of Valsalva aneurysm, a Morgagni hernia, and a patent foramen ovale. Her dyspnea and cyanosis resolved after sequential surgical correction of these defects.

Unruptured sinus of Valsalva aneurysm (SOVA) and Morgagni hernia (MH) are both uncommon findings. SOVA usually presents with a continuous murmur after rupture (1–3). In addition to causing abnormal intracardiac physiology (4), SOVA can be diagnosed before rupture as an asymptomatic finding on cardiac imaging and can be confused with an intracardiac mass lesion (5). MH may be discovered incidentally on imaging studies or may cause numerous symptoms including chest or abdominal pressure, dyspnea, or intestinal obstruction (6, 7). The occurrence of unruptured SOVA, MH, and patent foramen ovale (PFO) in the same patient has not been previously reported, based upon review in PubMed, nor has the simultaneous occurrence of these disorders in association with cyanosis.

CASE REPORT

A 52-year-old woman was noted to have exertional dyspnea and intermittent cyanosis of several months' duration. She had no previous cardiac history. Computed tomography angiography (CTA) of the chest demonstrated a mass involving the right hemidiaphragm and right atrium. Based on pulse oximetry, the patient had an oxygen saturation of 93% on room air while standing upright, which decreased to 79% when she bent forward. Reinterpretation of the chest CTA (*Figure 1*) demonstrated a large right-sided MH displacing the heart leftward. The MH defect was approximately 6 to 7 cm and contained visible omentum and transverse colon. There was significant mass effect in the mediastinum that was not itself thought to fully explain the observed intermittent cyanosis. A transthoracic echocardiogram demonstrated asymmetric dilatation of the noncoronary cusp bulging into the right atrium and pushing on the atrial septum, with right-to-left shunting across a PFO (*Figures 2 and 3*). A right and left cardiac catheterization

revealed normal coronary arteries and no evidence of pulmonary hypertension. Pulmonary function testing showed moderate obstruction without bronchodilator response and normal lung volumes, suggesting that the observed hypoxemia was due to right-to-left shunting.

Sequential surgical management followed. The MH repair was pursued, with a subsequent repair of her SOVA anticipated. Vascular air filters were used to prevent paradoxical air embolism. For the hernia, a laparoscopic transabdominal repair was chosen, which required the patient to tolerate steep reverse Trendelenburg positioning and intraabdominal insufflation with CO₂ to a pressure of 12 to 15 mm Hg. Careful attention to the risk of air embolus was discussed preoperatively, and potential shunt exacerbation due to insufflation increasing right-sided pressures was anticipated. Surgery was completed as planned, with operative findings including herniation of the majority of the omentum and midportion of the transverse colon. The defect was sizable, and there was attenuation of the diaphragm such that it was repaired using an underlay polyester prosthetic in a tension-free manner, using absorbable fixation and sutures to fixate the mesh to the defect, providing broad coverage, and taking care not to injure mediastinal structures above the diaphragm. The procedure was completed with only three working cannula sites, and the patient tolerated the procedure well.

After an interval of 2 months, the SOVA and PFO were repaired surgically. The PFO was closed primarily. Intraoperative findings also showed severely thinned out aortic sinus segments necessitating root replacement. Due to normal aortic valve anatomy, a valve-sparing root replacement was performed. An appropriately sized graft was chosen, plicated at the annulus and sinotubular junction and subsequently sewn to the base of the heart. The aortic cusps were then reimplanted inside the graft to preserve aortic valve function. The coronary arteries were reimplanted to complete the repair. Two seizures (the

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Figure 1. Chest computed tomography angiography. (a, b) Axial images reveal a large, predominantly fat attenuation lesion at the right cardiophrenic angle that exerts mass effect on adjacent structures (asterisk) and displaces the heart leftward, with elongation of the inferior vena cava (I). This lesion represents omental fat herniating through a Morgagni hernia, with the diaphragmatic defect better visualized on the sagittal reformat (c). There are several linear soft tissue attenuation structures that represent herniated omental vessels within the hernia (arrows, c), a characteristic finding of this lesion. Additionally, despite cardiac motion on this non-echocardiographic gated study, aneurysmal dilatation of the aortic root to approximately 5.5 cm is present (arrowheads, a).

patient had a history of epilepsy) and a brief episode of atrial fibrillation occurred postoperatively without sequelae. Following repair of the SOVA and closure of the PFO, the patient had marked improvement in her symptoms and complete resolution of her cyanosis.

DISCUSSION

We report the diagnosis and staged surgical management of MH, unruptured SOVA, and PFO in a patient who presented with dyspnea and intermittent cyanosis elicited by changes in position. The patient's initial presentation was radiographically concerning for vascular sarcoma given the presence of an intracardiac mass lesion and a mass involving the right hemidia-

phragm. Further interpretation of the CTA of the chest and subsequent echocardiography established the proper diagnosis. Thorough transthoracic two-dimensional echocardiography and color Doppler can be diagnostic in evaluating for the presence and cause of intracardiac shunts in patients with cyanosis. Acquired cyanosis in adults due to shunting is unusual and most commonly occurs in the situation of a PFO with an anatomic or hemodynamic reason for shunt reversal. The additional shifting of infradiaphragmatic and intrathoracic structures by the MH contributed to her symptoms.

The authors are unaware of any known unique genetic explanation for the concurrence of MH and SOVA. While SOVA can be congenital or acquired (8, 9), this patient demonstrated no known aortopathy-associated genetic mutations in a specific limited aortopathy panel (ACTA2, CBS, COL3A1, COL5A1, COL5A2, FBN1, FBN2, MYH11, MYLK, PLOD1, SKI, SLC2A10, SMAD3, SMAD4, TGFB2, TGFBR1, TGFBR2;

Figure 2. Transthoracic echocardiogram, four-chamber view. The arrow shows the dilated noncoronary sinus and cusp creating the appearance of a masslike structure in the right atrium (RA). LA indicates left atrium, LV, left ventricle; RV, right ventricle.

Figure 3. Color Doppler demonstrates right-to-left intraatrial shunting across a patent foramen ovale due to distortion of the atrial septum by the noncoronary cusp. LA indicates left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

aortopathy panel performed by ARUP Laboratories, Salt Lake City, Utah) (10). MH is congenital and is reported to be associated with multiple congenital disorders (11, 12). This report is the first known case, based on PubMed review, of SOVA and MH occurring simultaneously in the same patient.

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1. Harkness JR, Fitton TP, Barreiro CJ, Alejo D, Gott VL, Baumgartner WA, Yuh DD. A 32-year experience with surgical repair of sinus of Valsalva aneurysms. *J Card Surg* 2005;20(2):198–204.
2. Moustafa S, Mookadam F, Cooper L, Adam G, Zehr K, Stulak J, Holmes D. Sinus of Valsalva aneurysms—47 years of a single center experience and systematic overview of published reports. *Am J Cardiol* 2007;99(8):1159–1164.
3. Takach TJ, Reul GJ, Duncan JM, Cooley DA, Livesay JJ, Ott DA, Frazier OH. Sinus of Valsalva aneurysm or fistula: management and outcome. *Ann Thorac Surg* 1999;68(5):1573–1577.
4. Ellis A, Kalogeropoulos A, Chung A, Martin RP, Caras D, Clements SD Jr. Cyanosis due to unruptured aneurysm of the sinus of Valsalva: an unusual complication. *Tex Heart Inst J* 2011;38(4):448–449.
5. Sasaki S, Asano M, Fukuda K, Nishimura K, Mizuno A, Nomura N, Mishima A. Unruptured sinus of Valsalva aneurysm suspected to be a cardiac tumor. *Ann Thorac Surg* 2009;87(5):1619.
6. Nasr A, Fecteau A. Foramen of Morgagni hernia: presentation and treatment. *Thorac Surg Clin* 2009;19(4):463–468.
7. Horton JD, Hofmann LJ, Hetz SP. Presentation and management of Morgagni hernias in adults: a review of 298 cases. *Surg Endosc* 2008;22(6):1413–1420.
8. Goldberg N, Krasnow N. Sinus of Valsalva aneurysms. *Clin Cardiol* 1990;13(12):831–836.
9. Pomianowski P, Elefteriades JA. The genetics and genomics of thoracic aortic disease. *Ann Cardiothorac Surg* 2013;2(3):271–279.
10. ARUP Laboratories. Aortopathy Panel, 17 Genes. Available at <http://ltd.aruplab.com/Tests/Pdf/100>; accessed June 20, 2015.
11. Holder AM, Klaassens M, Tibboel D, de Klein A, Lee B, Scott DA. Genetic factors in congenital diaphragmatic hernia. *Am J Hum Genet* 2007;80(5):825–845.
12. Longoni M, High FA, Russell MK, Kashani A, Tracy AA, Coletti CM, Hila R, Shamia A, Wells J, Ackerman KG, Wilson JM, Bult CJ, Lee C, Lage K, Pober BR, Donahoe PK. Molecular pathogenesis of congenital diaphragmatic hernia revealed by exome sequencing, developmental data, and bioinformatics. *Proc Natl Acad Sci U S A* 2014;111(34):12450–12455.