Continuous machinery murmur in an octogenarian

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Abstract

Background: sinus of valsalva aneurysm is a rare congenital anomaly and usually presents in adolescence to early adulthood. Manifestations are varied, ranging from asymptomatic murmur to sudden death.

Case report: an elderly male presented with chest pain and machinery murmur, which was confirmed as ruptured sinus of valsalva aneurysm on echocardiogram. Emergency surgical repair led to a successful outcome.

Discussion: review of the literature revealed that this patient is the oldest reported case. Urgent echocardiography and surgical advice are essential. This condition is potentially treatable even in the older age group and the prognosis is good after surgical repair.

Keywords: sinus of valsalva aneurysm, rupture, older

Case Report

Sinus of valsalva aneurysm (SVA) is an uncommon condition, especially in the elderly, but it is a condition with varied manifestations. An 80-year-old male non-smoker with no previous ischaemic heart disease was admitted with sudden chest pain radiating to the back, associated with breathlessness. In the past, he had chronic airways disease managed by inhalers. Examination revealed a collapsing pulse; 84 beats/min in sinus rhythm, blood pressure (BP) 200/70 mmHg with no difference in BP in the two arms and continuous machinery murmur heard all over the precordium. ECG and chest X-ray were unremarkable, and serum troponin T was not elevated. An urgent chest CT did not reveal any aortic dissection. An urgent transthoracic echocardiogram showed a rupture of the right SVA into the right outflow tract (Supplementary figure 1a and b available at http://www.ageing.oupjournals.org). The patient was transferred to the regional cardiothoracic centre where right heart catheterisation revealed saturation of 60% in the right ventricle and 82% in the pulmonary artery. Angiography revealed mild plaque disease in the coronary tree. An emergency repair of the ruptured SVA was performed (Supplementary figure 2a and b). Postoperatively, the patient required inotropic support, ventilatory support and venovenous haemofiltration for two days. The remaining course was satisfactory and he was discharged home.

Discussion

SVA was described as ‘a rare congenital anomaly’ by John Thurnam in 1840. A congenital SVA is usually clinically silent, but manifestations may vary from a mild asymptomatic dilatation detected on routine two-dimensional echocardiogram...
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Table 1. Published studies in the literature on SVA

<table>
<thead>
<tr>
<th>SVA</th>
<th>Number of patients studied</th>
<th>Age (years)</th>
<th>Year study published</th>
<th>Reference number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ruptured</td>
<td>13</td>
<td>mean 24.5</td>
<td>1991</td>
<td>3</td>
</tr>
<tr>
<td>12 unruptured</td>
<td>104</td>
<td>5–65</td>
<td>1997</td>
<td>4</td>
</tr>
<tr>
<td>34 ruptured</td>
<td>53</td>
<td>4–60</td>
<td>2001</td>
<td>5</td>
</tr>
<tr>
<td>All ruptured</td>
<td>68</td>
<td>mean 29.5 ± 10.7</td>
<td>2001</td>
<td>6</td>
</tr>
<tr>
<td>All ruptured</td>
<td>18</td>
<td>20–65</td>
<td>2001</td>
<td>2</td>
</tr>
<tr>
<td>Unruptured</td>
<td>1</td>
<td>43</td>
<td>2001</td>
<td>7</td>
</tr>
<tr>
<td>All ruptured</td>
<td>67</td>
<td>2–57</td>
<td>2002</td>
<td>8</td>
</tr>
<tr>
<td>4 ruptured</td>
<td>5</td>
<td>18–48</td>
<td>2002</td>
<td>9</td>
</tr>
<tr>
<td>1 unruptured</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unruptured</td>
<td>1</td>
<td>75</td>
<td>2002</td>
<td>10</td>
</tr>
<tr>
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<td>1</td>
<td>75</td>
<td>2002</td>
<td>11</td>
</tr>
<tr>
<td>Unruptured</td>
<td>1</td>
<td>64</td>
<td>2002</td>
<td>12</td>
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<tr>
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<td>1</td>
<td>39</td>
<td>2002</td>
<td>13</td>
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<tr>
<td>Ruptured</td>
<td>35</td>
<td>7–64</td>
<td>2002</td>
<td>14</td>
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<tr>
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<td>52</td>
<td>2002</td>
<td>15</td>
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<tr>
<td>Unruptured</td>
<td>1</td>
<td>40</td>
<td>2002</td>
<td>16</td>
</tr>
<tr>
<td>Unruptured</td>
<td>1</td>
<td>62</td>
<td>2002</td>
<td>17</td>
</tr>
<tr>
<td>Ruptured</td>
<td>1</td>
<td>54</td>
<td>2003</td>
<td>18</td>
</tr>
</tbody>
</table>

Echocardiography can also quantify the severity of the defect. Cine MRI, if available, is also a diagnostic tool. Cardiac catheterisation reveals a shunt in ruptured SVA.

The main differential diagnoses of continuous murmur are coronary artery aneurysm with fistula to right heart, ventricular septal defect and patent ductus arteriosus.

Medical management involves optimising medications for heart failure. Urgent surgical repair is however recommended in all patients with ruptured SVA, especially with intra-cardiac shunting [22]. Clinical deterioration can be rapid, hence urgent cardiothoracic consultation is needed. Associated surgical procedures could include aortic root reconstruction/replacement, aortic valve reconstruction/replacement, Bentall procedure (valved conduit), repair of associated ventricular septal defect and atrial septal defect. Surgical repair has low mortality (2%) and morbidity. Complications of unruptured SVA include myocardial infarction from coronary artery compression, complete heart block, cardiac tamponade, infective endocarditis and sudden death. Without surgical correction, prognosis is poor as most unruptured SVAs have been found to progress and rupture.

Key points
- SVA is a ‘rare congenital anomaly’ and its first presentation with rupture in an octogenarian is extremely rare.
- Rupture of SVA causes machinery murmur.
- Urgent two-dimensional echocardiography is necessary, which gives reliable diagnosis by a non-invasive method.
- Definitive treatment is available and urgent surgical repair is necessary.
- Prognosis after surgery is good even in older age.

to symptomatic presentations related to compression of adjacent structures or to rupture of SVA into the heart. Approximately 65–85% of SVAs originate from the right sinus of valsalva, while SVAs originating from non-coronary (10–30%) and left sinuses (<5%) are rare [1].

The aetiology of SVA is usually a congenital defect in aortic media tissue and abnormal development of bulbous cordis. Other diseases which rarely produce SVA include atherosclerotic aneurysm, syphilis, endocarditis, trauma, cystic medial necrosis and Marfan syndrome [2]. The male:female ratio is 4:1 for both ruptured and unruptured SVA. Aneurysms may remain dormant for many years. Rupture of SVAs usually occurs after puberty and up to the age of 30 years and presents clinically in this age group [2]. A literature search revealed that, to our knowledge, our report highlights the case of the oldest patient with ruptured SVA to undergo surgical repair (Table 1).

Approximately 25% cases of SVA are clinically asymptomatic (unruptured) and are detected by routine echocardiography. Unruptured aneurysms may rarely cause symptoms by right ventricular outflow tract obstruction [19]. Rupture of the aneurysmal sac may occur spontaneously. Rupture of dilated sinus causes intra-cardiac shunting due to communication with the right atrium (Gerbode defect 10%) or is more commonly directed into the right ventricle (60–90%). Cardiac tamponade may occur if rupture involves pericardial space. If present, a loud superficial ‘machine type’ continuous murmur, accentuated in diastole, is characteristic. There may also be associated structural valvular heart defects. Progressive heart failure is the main cause of death.

Transthoracic echocardiography may detect 75% of all patients with SVA and a ‘windsock’ appearance may be identified [20]. However, trans-oesophageal echocardiography provides comprehensive information about diagnosis of SVA including structural cardiac anomalies and shunts [21].

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