A 16-year-old female with a family history of coronary artery disease was referred to hospital because of recurrent chest oppression unrelated to exertion. Although a selective coronary angiogram showed no atherosclerotic lesions, coronary spasm was provoked by acetylcholine in the left coronary artery, accompanied by chest pain and depression of blood pressure. This is the youngest healthy young female to be diagnosed with coronary vasospasm by provocation test. Because there are no risk factors, the vasospasm must be related to unknown hereditary factors. (Circ J 2003; 67: 467–469)

Key Words: Adolescent; Family history; Vasospastic angina

A 16-year-old female with VSA has been referred to hospital because of recurrent chest oppression unrelated to exertion. Although a selective coronary angiogram showed no atherosclerotic lesions, coronary spasm was provoked by acetylcholine (ACH) provocation test. She had no conventional coronary risk factors other than a family history of CAD or sudden death. We also review the literature on CAD in adolescents.
anterolateral wall in the right anterior oblique view, and an ejection fraction of 0.64. A selective coronary angiogram showed no atherosclerotic lesions. In order to confirm VSA, we provoked a spasm with intracoronary ACh (Ovisot®, Daiichi Pharmaceutical Co, Ltd, Tokyo, Japan). Just after injection of ACh (100 μg) into the left coronary artery, she showed symptoms of chest oppression and her systolic blood pressure dropped from 110 mmHg to 60 mmHg. At that time the coronary angiogram diffuse spasm of the left coronary artery (Fig 3). The ECG showed electronic pace-maker rhythm (50 beats/min) and so we were unable to assess ST-T changes. We immediately administered NTG into the left coronary artery and the coronary spasm improved. When comparing the coronary artery diameters after ACh and after NTG by quantitative coronary angiography (QCA) with CARDIO 500 (Kontron, Munich, Germany), the diameter at segment 6 changed with NTG from 1.63 mm to 4.09 mm (%stenosis by Ach was 60%), at segment 8 from 0.81 to 1.98 (59%) and at segment 15 from 0.99 to 1.51 (32%). Thus, the average %stenosis induced by ACh was 75% by the American Heart Association (AHA) classification. Consequently, we diagnosed VSA and prescribed a calcium antagonist agent. The patient has not had symptoms of chest oppression for 20 months.

Discussion

Vasospastic angina occurred in a 16-year-old female who was in otherwise good health and had no risk factors other than her family history. CAD, apart from congenital heart disease or in association with Kawasaki disease, is rare in adolescents, especially VSA, to our knowledge, there are only 4 other reported cases2–4 (Table 1), none of which have been in such a young female.

Table 1  Variant or Vasospastic Angina in Cases Under 20 Years of Age

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Author</th>
<th>Age (years)/gender</th>
<th>Clinical diagnosis</th>
<th>Past history</th>
<th>Family history of CAD</th>
<th>Spasm</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Hosei et al</td>
<td>13/M</td>
<td>AMI, VSA</td>
<td>None</td>
<td>None</td>
<td>Positive</td>
</tr>
<tr>
<td>2</td>
<td>Wilkes et al</td>
<td>11/M</td>
<td>VA</td>
<td>Sickle cell anemia</td>
<td>None</td>
<td>Unknown (not performed)</td>
</tr>
<tr>
<td>3</td>
<td>Ivy et al</td>
<td>16/M</td>
<td>AMI, VA</td>
<td>None</td>
<td>None</td>
<td>Unknown (not performed)</td>
</tr>
<tr>
<td>4</td>
<td>Flohr et al</td>
<td>18/F</td>
<td>AMI, VSA</td>
<td>None</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>Patient</td>
<td>Kobayashi et al</td>
<td>16/F</td>
<td>VSA</td>
<td>None</td>
<td>Positive</td>
<td>Positive</td>
</tr>
</tbody>
</table>

CAG, coronary angiogram; AMI, acute myocardial infarction; VA, variant angina; VSA, vasospastic angina; CAD, coronary artery disease.

Vasospastic angina occurs in adults over 40 years of age and the etiology is thought to be endothelial cell dysfunction caused by atherosclerosis and various other factors associated with atherosclerosis, such as aging, smoking, hypertension, hyperlipidemia, and diabetes mellitus. However, the present patient has no such risk factors. Vasospastic angina appears to occur more frequently in certain geographical locations, namely, Japan, Italy and Canada. Recently, cases of VSA in families with a history of CAD have been reported8–10 and it is suspected that the etiology involves a gene defect, such as an abnormality of the endothelial nitric oxide synthase (eNOS) gene, or of the HLA antigen11–13. However, the association between a mutation of the eNOS gene and coronary spasm is still controversial13–15.

The present case of VSA is the youngest to be diagnosed by an ACh provocation test. The prognosis of VSA in adolescents remains unknown, so careful and frequent follow-up examinations are needed.
References


