Characteristics and Fate of Systemic Artery Aneurysms caused by Kawasaki Disease

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FINANCIAL DISCLOSURE:
We have no relevant financial relationship.
Introduction

Systemic artery aneurysms (SAA) develop in approximately 2% of untreated patients and 10% of patients with giant coronary aneurysm caused by KD.

*Circulation. 1996;94 1379-85 Kato H et al*

SAA refers to aneurysms developing anywhere in the arterial system other than the coronary circulation.
Objective

The objective of this study is to clarify the characteristics and fate of SAA caused by KD.
Methods

20 pts with SAA between 1980 and 2013 at NCVC in Japan

1. The characteristics of the patients with SAA was analyzed, and the distribution and fate of SAA were also evaluated.

2. We analyzed the residual rate of SAA in the late period and the incidence of stenotic lesions in 11 pts with SAA who underwent an initial angiogram less than 4 months after the onset of KD.
**Interval from the the onset of KD to respective angiogram**

<table>
<thead>
<tr>
<th></th>
<th>Range</th>
<th>Median</th>
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</thead>
<tbody>
<tr>
<td>Initial angiogram</td>
<td>1-53 months</td>
<td>3 months</td>
</tr>
<tr>
<td>(n=20)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Latest angiogram</td>
<td>2-26 years</td>
<td>18 years</td>
</tr>
<tr>
<td>(n=17)</td>
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</tbody>
</table>
Patients

Sex

- Female: 6 (30%)
- Male: 14 (70%)

Onset age of KD

- \( \leq 1 \) year: 15 (75%)
- \( > 1 \) year: 5 (25%)

All patients had bilateral coronary aneurysms.

\( n = 20 \)

Median 6 months

1 ~ 20 months
Treatment of acute KD

n = 20

Initial therapy 5 ± 2 of the illness day
The mean duration of fever 24 ± 12 days

IVIG 2 (10%)
Steroid 3 (15%)
IVIG, Aspirin and Steroid 3 (15%)
Aspirin 4 (20%)
IVIG and Aspirin 7 (35%
Unknown 1
Treatment in the late period

- Coumadin with antiplatelets: 8 (40%)
- Antiplatelets: 12 (60%)

n = 20
# Cardiac events

<table>
<thead>
<tr>
<th>Event</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Acute myocardial infarction</td>
<td>7 pts (35%)</td>
</tr>
<tr>
<td>Coronary artery bypass grafting</td>
<td>8 pts (40%)</td>
</tr>
<tr>
<td>Death</td>
<td>4 pts (20%)</td>
</tr>
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</table>
Distribution of SAA

Upper extremities

Brachial 28%

Common iliac 19%

Internal iliac 19%

Lower extremities

total = 108
Regression of aneurysm in brachial artery

6 months

6 years

5.0 mm
Changes of aneurysms in brachial arteries

7 months

Left

10.1 mm

Right

13.4 mm

18 years

Residual aneurysm

Segmental stenosis with calcification
Regression of aneurysms in bilateral common and internal iliac arteries

20 months

8.7 mm  7.0 mm
9.9 mm  8.7 mm

13 years
Changes of aneurysms in bilateral iliac arteries and abdominal aorta

7 months

22 years

Occlusion With calcification
7 months old
Abdominal aorta aneurysm and bilateral common iliac aneurysms $\geq 30$ mm

23 months old
Replacement using artificial vessels
Outcome of SAA in 17 pts

- Subcravian
- Brachial
- Common Iliac
- Internal Iliac
- Femoral
- Abdominal

Regression: Yellow
Aneurysm: Orange
Stenotic lesion: Green
Surgery: Blue

n = 78
Residual rate of SAA in 11 pts (Brachial, common and iliac arteries)

Residual SAA included persistent aneurysms and stenotic lesions.
### Cut-off values of the initial diameter leading to residual SAA

<table>
<thead>
<tr>
<th></th>
<th>Diameter (mm)</th>
<th>AUC</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brachial (n=14)</td>
<td>6.8</td>
<td>0.96</td>
<td>0.0003</td>
</tr>
<tr>
<td>Common Iliac (n=16)</td>
<td>9.4</td>
<td>0.75</td>
<td>0.0339</td>
</tr>
<tr>
<td>Internal Iliac (n=12)</td>
<td>10.7</td>
<td>0.92</td>
<td>0.0035</td>
</tr>
</tbody>
</table>
Incidence of stenotic lesions in 11 pts
(Brachial, common and iliac arteries)

Stenotic lesions (n = 6)
- Occlusion 4
- Segmental stenosis 1
- Localized stenosis 1

Years after acute Kawasaki disease

20-year
25%

10-year
6%
1. All patients with SAA had had acute KD within 20 months with a median of 6 months.

2. All patients had at least one symmetric pair of aneurysms in bilateral peripheral arteries. Sixteen pts (80%) had multiple aneurysms.

3. The prevalence of SAA was high in the brachial, common and internal iliac arteries.
4. The incidence of cardiac events was high in patients with SAA.

5. The larger aneurysms persist into the late period, and the acute phase diameter of SAA leading to late stenotic lesions was more than 10.0 mm.
Conclusions

SAA occurred symmetrically and were multiple in younger infants and those with severe acute vasculitis. The fate of SAA resembles that of coronary artery aneurysms, and depends on its acute phase diameter. The larger SAA can lead to stenotic lesions in the late period.