Case 1/2012 – 55-year-old male with multiple coronary artery aneurysms due to Kawasaki disease

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Clinical data
The patient, six months before, had complained of diffuse pain in the right hemithorax at rest, which radiated to the left elbow at exertion. At the chest radiography and tomography (Fig. 1), cardiac opacities in imaging examination led to the hypothesis of the presence of coronary stents, which was discarded, as they had not been previously implanted. On further investigation by echocardiography, giant coronary aneurysms were detected in the coronary arteries.

The CT angiography adequately identified the aneurysms as giant calcified ones, located in the proximal third of the anterior interventricular and circumflex arteries, in addition to two others in the mid-third of the right coronary artery. Stress test and myocardial scintigraphy revealed no abnormalities. Medical history disclosed prolonged fever for a few days at seven years of age, with no etiologic confirmation or specific treatment at the time.

Physical Examination
Good general status, normal skin color, normal pulses.
Weight: 115 Kg; height: 188 cm; BP: 120/80 mmHg; HR: 64 bpm.

The aorta was not palpable at the suprasternal notch. There were no precordial deformities and the ictus cordis was not palpable. Normal heart sounds were identified and there were no heart murmurs. The liver was not palpable and the lungs were clear.

The ECG showed sinus rhythm, no signs of overload or ventricular repolarization. AQRS: +90°, AP: +60°, AT: +80°.

Chest X-ray and tomography
The imaging examinations showed normal cardiac silhouette and pulmonary vasculature, as well as opacities in the cardiac imaging caused by calcified aneurysms in the coronary arteries (Figure 1).

Diagnostic impression
The CT images are compatible with the diagnosis of coronary aneurysms due to Kawasaki disease, caused by fever he had had in childhood, even though unnoticed at the time.

Differential diagnosis
Other heart diseases that are associated with multiple aneurysms of the coronary arteries are related to other vascular disorders, although they have different causes. In this context, we can cite polyarteritis nodosa, among other connective tissue diseases, Takayasu disease and other infectious nonspecific arteritis.

Diagnostic confirmation
The images were decisive elements for the diagnosis of Kawasaki disease, given the presence of multiple aneurysms. Cardiac catheterization (Figure 2) confirmed the pre-established diagnosis, as well as the location of the giant aneurysms in the coronary arteries and obstruction of about 70% of the circumflex artery after the aneurysm, in its proximal third.

Fig. 1 – Chest tomography showing normal cardiac silhouette and pulmonary vasculature. Opacities in cardiac imaging (arrow) were subsequently confirmed as being caused by calcified giant aneurysms of the coronary arteries.

Keywords
Mucocutaneous lymph node syndrome; coronary aneurysm/therapy.

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Conduct

Expectant conduct was adopted, considering the absence of characteristic symptoms and myocardial perfusion alterations.

Comments

Aneurysms caused by Kawasaki disease occur between 20% and 30% of cases, especially in patients not treated in the disease onset. With the use of high doses of gamma-globulin (2 g/kg/day) and acetyl salicylic acid (100 mg/kg/day), the occurrence of aneurysms decrease to 13% and 18% respectively to the aforementioned drugs. They are typically multiple, but small-sized aneurysms, which allows the involution process by aneurysm endothelialization, up to one year after the acute episode. Such evolution often occurs, and the aneurysms persist in only 10% of cases after the first year of the disease. As for the giant aneurysm with a diameter greater than 8 mm, which occurs in 7% to 10% among all aneurysms, it often leads to unfavorable evolution with the development of thrombi, myocardial infarction and death. These cases therefore require early myocardial revascularization.

Rare cases develop as the one presented here to adulthood, and such evolution without complications is due to the absence of thrombus formation and obstruction, which perhaps, in this patient, were prevented by aneurysm endothelium calcification.

This evolution shows that the expectant conduct might be preferred, as decades went by without adverse events. Preventive drug treatment with antiplatelet adhesive and antilipemic drugs should be recommended in addition to clinical observation and sporadic imaging assessment for an eventual change in conduct, if necessary. The literature shows that adult patients at risk correspond to those with coronary aneurysms and obstructions who developed myocardial infarction, arrhythmias and sudden death. The clinical-surgical experience determines the most appropriate conduct for each case.

Fig. 2 – Coronary angiography disclosed giant aneurysms in early-thirds of the anterior interventricular, circumflex and right coronary arteries (A, B and C).