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Imaging of acquired coronary diseases: From children to adults



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KEYWORDS

Coronary artery;
Aneurysm;
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Vasculitis;
Dissection;
Kawasaki

Abstract Acquired coronary diseases include aneurysms, fistulae, dissections, and stenosis. Aneurysms may occur secondarily to Kawasaki disease, a childhood vasculitis, the prognosis of which depends on the coronary involvement, or they may be degenerative, infectious, inflammatory, or traumatic in origin. Fistulae develop between the coronary arterial system and a pulmonary or bronchial artery, or cardiac cavity. Dissections may occur spontaneously or may be post-traumatic. These coronary abnormalities may be found incidentally or may present as complications, infarction or rupture. The goals of this article are to understand acquired childhood and adult coronary diseases and their usual means of presentation, the ways of investigating them, and the principles of their treatment.

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Acquired coronary diseases include aneurysms, ectasiae, fistulae, dissections, and stenoses. The imaging challenges are different for each of these abnormalities as whilst the positive diagnosis of aneurysm or ectasia does not in principle raise any particular imaging difficulties, identification of their cause and assessment of their risk of complications are essential, as these influence the overall diagnostic approach and treatment. Stenoses are usually atheromatous in origin but may also occur secondarily to vasculitis. The main purposes of imaging are then to assess the extent of the stenosis and its hemodynamic consequences.

Abbreviations: DES, drug-eluting stent; IVUS, intravascular ultrasound; JCS, Japanese Circulation Society; LAD, left anterior descending; TTE, transthoracic echocardiography.

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Ectasiae and aneurysms: definition

Coronary artery ectasiae and aneurysms are defined as an abnormal dilatation of a coronary artery, which are respectively diffuse, affecting over 50% of the total length of the vessel, or local, affecting less than 50% of the total length of the vessel (Figs. 1 and 2) [1]. Saccular aneurysms (whose transverse diameter is greater than the length), which are at increased risk of thrombosis and rupture, can be distinguished from fusiform aneurysms (whose length is greater than their diameter), and true aneurysms (affecting the three arterial linings) from false aneurysms (in which the arterial wall is damaged, often as a result of injury, a iatrogenic effect or fungal disease) [1]. Finally, giant aneurysms are defined as those with a diameter of over 20 mm in adults and 8 mm in children. They have many varied causes, which include degenerative, infectious, inflammatory, and traumatic [2]. Ectasiae are commoner and are usually atheromatous in origin (Fig. 3), compensating for stenosis in another coronary artery or secondary to high flow rate as in a coronary artery fistula.

Kawasaki disease

Definition and positive diagnosis

Kawasaki disease is a childhood panvasculitis affecting the medium diameter arteries with particular tropism for the coronary arteries. Its cause is not known and it is



Figure 1. Coronary ectasia (arrows). Coronary CT angiography with curvilinear reconstruction shows a dilatation that involves over 50% of the total length of the vessel.

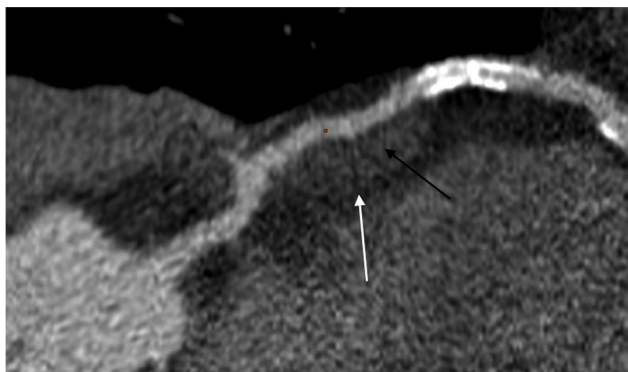


Figure 2. Coronary aneurysm (white arrow) with mural thrombus (black arrow). Coronary CT angiography with curvilinear reconstruction shows a local dilatation that affects less than 50% of the length of the vessel.



Figure 3. Atheromatous coronary ectasia (white arrow). Coronary CT angiography in the transverse plane shows irregular ectasia of the left anterior descending artery in association with a stenosed calcified atheromatous plaque (black arrow) in the main stem and proximal left anterior descending artery.

probably due to exposure to a causal or environmental agent combined with genetic predisposition. Coronary artery disease involvement may present with aneurysm, premature atheroma, stenosis, thrombosis, or occlusion with infarction, defines the prognosis of the disease and influences survival [3,4]. Fifteen to 25% of untreated children develop coronary aneurysms and ectasiae which may result in ischemic complications or sudden death [4]. Kawasaki disease has overtaken acute rheumatic disease as the most common cause of acquired heart disease in children.

In its conventional forms, Kawasaki disease is diagnosed from clinical criteria and a negative etiological assessment (Boxed text 1) [4]. In incomplete forms of the disease, cardiological investigation with transthoracic echocardiography (TTE) may be required to investigate for signs of the disease (Boxed text 2) and to begin treatment [3,4].

Natural history and features of coronary aneurysms

Whilst these often present in the subacute phase (between the 10th and 25th day), coronary artery aneurysms may also

Boxed text 1: Positive diagnosis of classical Kawasaki disease

- Fever for over 5 days + 4 of the 5 clinical criteria:
- Involvement of the extremities (acute: palmar and/or plantar erythema, palmoplantar edema).
 - Subacute: secondary "glove finger" palmoplantar scaling.
 - Polymorphic exanthema.
 - Bilateral non-exudative conjunctivitis.
 - Lip and oral cavity involvement (labial and buccal mucosa, edema, strawberry tongue, buccal and pharyngeal mucosal edema).
 - Cervical lymphadenopathy over 1.5 cm in diameter, firm and generally unilateral.

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