Scientific letters

Association of Spontaneous Coronary Artery Dissection With Fibromuscular Dysplasia

Asociación de disección coronaria espontánea con displasia fibromuscular

To the Editor,

Spontaneous coronary artery dissection (SCAD) is a very uncommon clinical entity, whose etiology and pathophysiology are still not fully understood. The main diagnostic key of this entity is the finding-on coronary angiography-of a linear contrast defect, with longitudinal distribution (Figures A and B). Intracoronary imaging techniques (intravascular ultrasound and optical coherence tomography) allow more precise morphological diagnosis (Figures C and D).

SCAD has been associated with multiple diseases, and recently, with fibromuscular dysplasia (FMD), which is another rare arterial disease, neither inflammatory nor atherosclerotic, of equally uncertain etiology. In patients with SCAD, the systematic search for noncoronary arterial vascular disease has shown a prevalence of FMD of around 70%.1-3

In this study, we describe our experience with the diagnosis of FMD in patients with diagnosed SCAD.

From April 2011 until November 2014, patients diagnosed with SCAD in our center were included in a protocol of investigation and clinical follow-up (mean follow-up, 17 months). A “conservative strategy” was followed, which only indicated revascularization in patients with persistent symptoms or recurrent ischemia. The imaging technique used was specifically directed at the detection of arterial vascular disease in noncoronary territories.

Nine consecutive patients were included: 1 man and 8 women; all patients were middle-aged (Table). The diagnosis of SCAD was made during coronary angiography, which was done because of acute coronary syndrome. In 3 patients, diagnosis was made using intravascular ultrasound or optical coherence tomography.

Eight patients underwent extracardiac arterial disease study (1 patient declined further investigation). The arterial territories studied were the renal arteries, the iliofemoral arteries, the...

**Figure.** A: Coronary angiography (patient 2) with spontaneous coronary artery dissection in the posterior descending artery (arrows). B: The same patient 6 months later, with complete resolution on angiography. C: Intravascular ultrasound in spontaneous coronary artery dissection (patient 1); true lumen and false lumen separated by an intimal flap, and the entry point (arrow); D: Intravascular ultrasound showing coronary hematoma (patient 1); a parietal hematoma (H) and guidewire artefact (*). E: External iliac artery (patient 9); angiographic irregularities compatible with fibromuscular dysplasia (arrows). F: Left renal artery (patient 7) with parietal irregularities typical of fibromuscular dysplasia (arrows). G: Computed tomography angiography of supra-aortic trunks (patient 5); Both internal carotid arteries have the characteristic string of beads appearance (arrows). H: Computed tomography angiography (patient 4) showing splenic artery aneurysm of 7 mm diameter (circle). FL, false lumen; H, parietal hematoma; TL, true lumen.

1885-5857/© 2015 Sociedad Española de Cardiología. Published by Elsevier España, S.L.U. All rights reserved.
supra-aortic trunks, and the intracranial vessels (Table). In half
the patients, invasive angiography was used (either during the
index procedure or scheduled), and in the other half of the
patients, computed tomography angiography was used.
Of the 8 patients studied, only 1 had completely normal
extracardiac arteries. In 5 patients, there were findings compati-
ble with FMD, of variable severity (Figures E-G). One patient had a
splenic artery aneurysm and an anterior communicating artery
aneurysm in the circle of Willis (Figure H). The remaining patient,
who was older, had multiple calcified plaques in the abdominal
aorta and iliac arteries, of atherosclerotic etiology.

Until very recently, our knowledge of SCAD was based on
isolated cases and short retrospective series. Nowadays, we have
access to information from longer prospective series2,−4; SCAD
generally affects women of middle age, with not without classic
cardiovascular risk factors and with no clear association with
immunological, inflammatory, or connective tissue diseases,
although isolated cases have been described in all those
situations. Traditionally, SCAD has been associated with the final
weeks of pregnancy and the postnatal period, but we now know
that this association has a low prevalence.7−4 Also, recent studies
indicate the benefit of an initial conservative strategy, reserving
revascularization for patients with persistent or recurrent
symptoms.4

Recently, several groups have highlighted the association of
SCAD with FMD.2,3 The most common form of FMD is medial
fibroplasia. The characteristic angiographic pattern of
medial fibroplasia is that of alternating dilated and stenosed
areas, forming what is called a “string of beads” appearance.
Fibromuscular dysplasia can also be associated with aneurysms
and dissections, which may form a pathophysiological link with
SCAD. Anatomopathologically, this image corresponds with
alternating zones of thickening and thinning of the arterial medial
layer. These findings have recently been visualized in vivo using
optical coherence tomography.4,6

The high prevalence of the association of SCAD with FMD,2,4
very uncommon diseases, raises suspicion of a strong
pathophysiologic relationship between them. The data that we
present support this association (75% of the patients). In our
experience, computed tomography angiography directed at
detecting FMD has an adequate diagnostic value. It must be
acknowledged that, in our series, the low number of patients
hinders evaluation of the power of this association and its possible
clinical implications. Also, the screening of extracardiac arterial
disease was done with different techniques, which was a
limitation of the study.

The clinical significance of extracardiac findings is very difficult
to establish. Probably, mild paitetal changes should only be
considered “stigmata” of FMD, with no functional repercussions,
and should not alter these patients’ management. However, more
severe changes (significant aneurysms or stenoses), may require
specific therapeutic interventions. The pathophysiological
implications of this new association are not clear. New studies are
needed with systematic screening for noncoronary vascular
disease in patients with SCAD to establish the possible implications
of this interesting association.

Teresa Bastante,a Fernando Rivero,a Javier Cuesta,a Julián Cuesta,b
Amparo Benedicto,a and Fernando Alfonsoa,b

aServicio de Cardiología, Hospital Universitario de La Princesa, Madrid, Spain
bServicio de Radiodiagnóstico, Hospital Universitario de La Princesa, Madrid, Spain

*Corresponding author:
E-mail address: falf@hotmail.com (F. Alfonso).
Available online 14 May 2015

REFERENCES

 coronary artery dissection: association with predisposing arteriopathies and
 Spontaneous coronary artery dissection. Long term follow-up of a large series of
 patients prospectively managed with a “conservative” therapeutic strategy. J Am
 Coll Cardiol Intv. 2012;5:1062–70.
5. Bastante T, Alfonso F. Insights of optical coherence tomography in renal artery
 fibromuscular dysplasia in a patient with spontaneous coronary artery dissec-
dysplasia: in vivo optical coherence tomography insights. Eur Heart J.
 2014;35:931.

http://dx.doi.org/10.1016/j.rec.2015.02.018