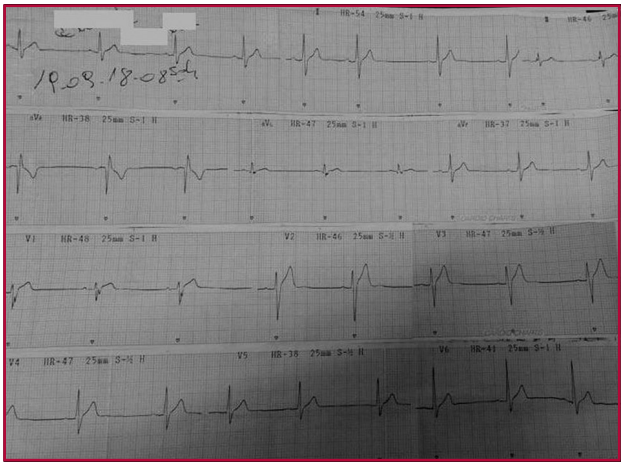

Fig. 1. Patient ECG showing heart rate of 50 bpm, absolute QT of 320 ms and a corrected QT by Bazett's formula of 292 ms.

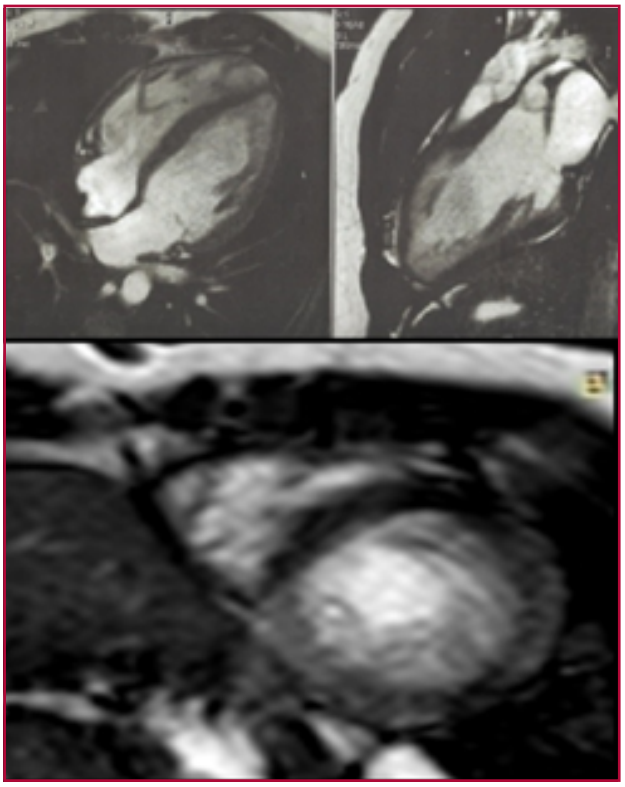

Fig. 2. Cardiac MRI. *Top left:* Four-chamber view; *on the right:* Two-chamber view of the left ventricle; both images show myocardial hypertrabeculation predominantly in the apical region. *Bottom:* Short axis view with hypertrabeculation, typical of noncompacted myocardium. Noncompacted/compacted myocardium ratio > 2.4

nificantly increased risk of SD. (6)

In view of the well-known difficulties in the lack of SD risk-stratification in this clinical scenario, and lack of information on the safety of quinidine use for structural heart disease, implantation of ICD was decided and family screening was performed, in which no phenotypes were detected.

We describe for the first time this new entity that associates a structural cardiomyopathy (SCM) with a channelopathy (SQTS). It will be difficult to discriminate whether it is merely an incidental association or it is due to a genetic mutation and constitute its phenotypic manifestation, together with persistent sinus bradycardia. Follow-up and description of further cases, as well as family genetic testing of this novel syndrome will be central to answer this question.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material).

Ethical considerations

Not applicable

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Abdominal Aortic Coarctation in Noonan Syndrome

Abdominal aortic coarctation or hypoplasia is a rare condition. Its incidence, evaluated in a serial autopsy study, was 1/62,500. (1) Magnoli et al. (2) reported 20 cases of aortic coarctation among 1,500 patients treated for aortoiliac obstruction. Three of these patients also had aneurysmal aortic dilatation. Coarctation of the abdominal aorta is more common in women than in men. In a series of 18 cases presented by Delaurentis et al., (3) there was only one man. This condition was first described

by Quain in 1847 as an aggressive variable of atherosclerotic vascular disease. (4) Since only a few cases are described in the literature, its pathophysiology is yet unclear. Some researchers hypothesize that abnormal fusion of both embryonic dorsal aortas in the first month of intrauterine life is responsible for this congenital anomaly. Moreover, some authors believe that infectious or inflammatory mechanisms, including radiotherapy, atherosclerosis and rubella, among others, could trigger this condition. (5) Pac et al. (6) have described 2 cases of aortic coarctation associated with Noonan syndrome. Narrowing of the aorta usually manifests as severe hypertension or intermittent claudication.

We report the case of a 21-year-old male patient consulting for intermittent claudication. The patient had a history of thoracic aortic coarctation treated with a stent and balloon angioplasty of the right iliac artery secondary to stenosis in 2013. Physical examination revealed facies with a peculiar phenotype, dimorphism of the auricles, eyelids and palate associated with supination of both elbows (Figure 1). Genetic testing was consistent with Noonan syndrome. Peripheral pulses and lab test results were normal. The CT angiography showed subocclusive infrarenal aortic stenosis with recanalization in the internal iliac arteries through the iliolumbar arteries and the inferior mesenteric artery (Figure 2). Hypotrophy in the origin of both primitive iliac arteries was also observed, but with adequate distal flow. Digital subtraction angiography revealed a competent aortic valve and a nondilated aortic root. The stent was patent, without restenosis, located distally to the origin of the subclavian artery. Infrarenal abdominal aortic coarctation associated with a significant long lesion in the right primitive iliac artery and complete occlusion of the left primitive iliac artery were confirmed. Assessment was completed with echocardiography and stress test, which were normal. An elective, aortoiliac bypass was performed using a Dacron graft. The patient was discharged without any complications 4 days after the procedure. Correct perfusion of the lower limbs was evidenced in the immediate postoperative course, with complete relief of symptoms. Specifically, infrarenal aortic hypoplasia lacks a clear definition. However, it typically presents as an aortic artery segment with a diameter <12 mm. Four types of aortic coarctation have been described: type I, suprarenal coarctation and renal artery stenosis; type II, infrarenal coarctation and renal artery stenosis; type III, suprarenal coarctation and normal renal arteries; and type IV, infrarenal coarctation and normal renal arteries. Therefore, our patient is included in groups III/IV. Its treatment remains open to debate, and the usefulness of new endovascular techniques for this condition is being discussed. Nonetheless, given that stenosis is generally long, conventional surgery prevails over endovascular methods. The few case series correspond mostly to conventional surgery. Since no studies compare one technique with the other, it is impossible to establish differences in technical success and outcomes between the two methods over time. In our patient, endovascular treatment for thoracic aortic stenosis was successful, with a patent period > 15 years. A conventional surgical approach was decided due to the length of the stenosis and the evidence in the literature. One year after surgery, the patient has no limitations in his daily life activities, but his assessment over time is still pending.

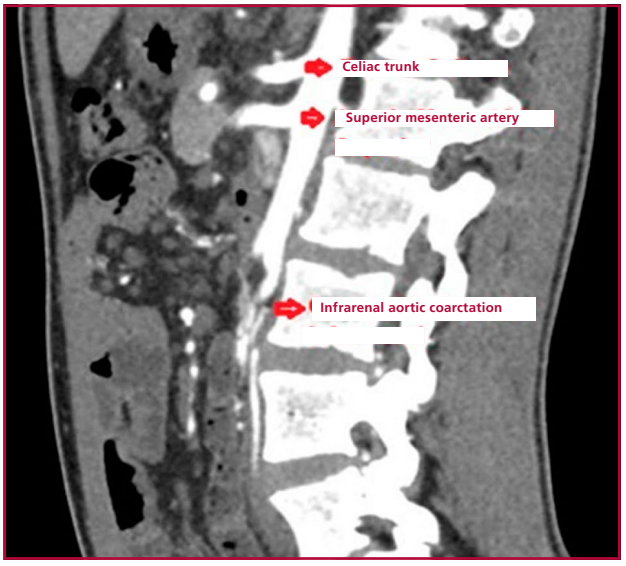

Fig. 2. Sagittal section of preoperative CT angiography showing infrarenal aortic coarctation

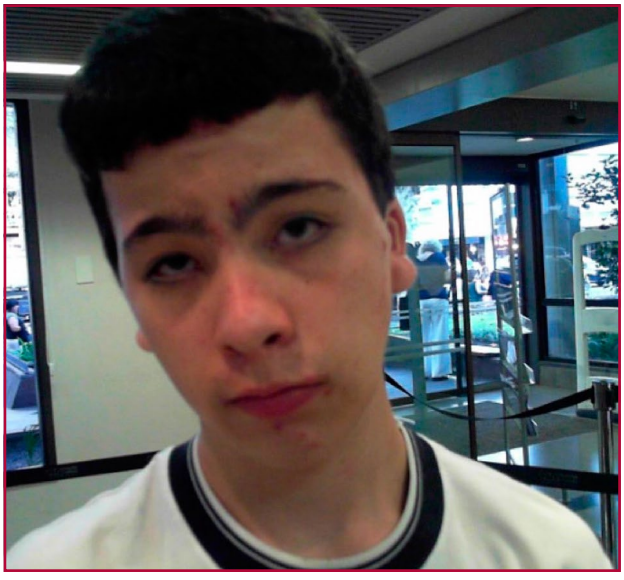

Fig. 1. Noonan syndrome phenotype

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material).

Ethical considerations

The patient has given consent for publication of his case.

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Renal Artery Aneurysm with Bilateral Renal Artery Stenosis

We report the case of a 23-year-old female patient with a history of hypothyroidism, chronic anemia due to thalassemia, obesity, sedentary lifestyle, and hereditary family history (father with type 2 diabetes). The patient refers a 3-year history of hypertension (HTN), with off-target systolic and diastolic blood pressure of 180/120 mmHg despite combination therapy with 4 drugs: hydrochlorothiazide, valsartan, amlodipine and carvedilol. The patient also refers FCI dyspnea (NYHA classification). On physical examination, the patient was lucid, oriented, and tolerating decubitus; blood pressure in the right arm was 190/120 mmHg and in the left arm, 192/120 mmHg. Cardiovascular system: Normal, regular S1 and S2; no S3; apex beat in the 5th left intercostal space. IBD. Preserved peripheral pulses.

Complementary studies

Electrocardiogram: Sinus rhythm, left ventricular overload. Chest X-ray: CTI <0.5. Doppler echocardiography: LVDD: 49 mm, LVSD: 33 mm, IVS: 13 mm, PW: 13 mm, LVMI: 130 g/m2, aortic root: 29 mm, LA: 35 mm, RVDD: 15 mm, EF: 60%, ShF: 40%, TAPSE: 23 mm, normal wall motion, aortic peak gradient 8.8 mmHg. Lab tests: red blood cells 4.7; hemoglobin 9.3 mg%; hematocrit 32%; glycemia 81 mg/dL; cholesterol 120 mg/dL; urea: 34 mg/dL; creatinine 0.88 mg/dL; creatinine clearance 124.5 mL/min; hematuria (–); HIV (–); VDRL (–); microalbuminuria 3.06 mg/L, proteinuria 12.24 mg/24 h.

Abdominal X-ray and abdominal ultrasound performed due to suspected secondary, probably renovascular, hypertension, were normal. In turn, renal ultrasound revealed an anechoic mass of 1.95 cm and 1.44 cm in the right renal pelvis. Doppler ultrasound of the renal arteries showed peak systolic velocity (PSV) in the main right renal artery (RRA) of 210 cm/s (NV <180 cm/s), resistance index (RI) >0.7 (NV <0.70), and acceleration time of 0.13 cm/s (NV <0.1). Peak systolic velocity in the left main renal artery (LRA) was 173 cm/s, and RI 0.65. Renal digital subtraction angiography evidenced a severe lesion in the middle third of the LRA, another severe lesion in the middle third and posterior RRA and a narrow neck saccular aneurysm was targeted in the main RRA bifurcation. After evaluating the therapeutic options, endovascular exclusion of the aneurysm and embolization with 5 coils (Barricade Coil System) was chosen: one of 9 x 30, two of 8 x 27 and two of 7 x 19 mm, resulting in aneurysm compaction. A main RRA angioplasty with a 4.30 mm x 16 mm stent was then performed. The following month, an angioplasty with a 4.0 mm x 16 mm Corflex stent in the main LRA was performed.

The patient made good progress after the procedure: Color Doppler ultrasound of the renal arteries: RRA: PSV: 170 cm/s, RI: 0.63; LRA: PSV: 165 cm/s, RI: 0.60; and ABPM: Systolic night–day HTN, grade I non-dipper. Selective renal angiography at one year was normal. The patient remains normotensive and under treatment with carvedilol, amlodipine, aspirin, and statins, and has discontinued clopidogrel.

In the study for suspected secondary hypertension refractory to treatment, we chose Doppler ultrasound of the renal arteries with 85% sensitivity and 92% specificity. Our patient was diagnosed RRA stenosis, and the evaluation was supplemented with renal artery angiography with digital subtraction, a gold standard method with 94% sensitivity and 93% specificity. Renal Doppler ultrasound may be negative in 10-20% of the cases, which would explain the lack of diagnosis of LRA stenosis.

Renal artery aneurysm (RAA) is a rare vascular entity (0.09-0.3% of all aneurysms) –usually saccular and mostly of extraparenchymal– affecting the bifurcation of the renal artery. (1) This condition often presents with hypertension due to renal artery stenosis or renal ischemia secondary to thromboembolization distal to the aneurysm. Mean age of presentation is 40-60 years; however, in young women with severe hypertension -in the absence of obesity, contraceptives and parenchymal renal disease-, the etiology is often attributed with more prevalence to fibromuscular dysplasia (FMD), a non-inflammatory vascular disease, or atherosclerosis, and less frequently, to congenital or fungal disorders, polyarteritis nodosa, trauma, syphilis, or tuberculosis. (2)