Stent Thrombosis and COVID-19

The outbreak of the COVID-19 pandemic has changed people's lives in countless ways worldwide. In the medical environment, the involvement of the respiratory tract is vital, but no less important than the involvement of the coagulation cascade, causing thrombotic events. (1) Endothelial dysfunction, atheromatous plaque destabilization, cytokine release with hyperinflammation, and hypoxia aggravation have been postulated as the origins of coagulation disorders and subsequent disseminated intravascular coagulation, although much of this complex mechanism is unknown. Cardiovascular complications of COVID-19 include myocardial injury (up to 30% of the cases) and venous thromboembolic events (up to 20%). However, arterial thrombotic events have been described more recently. In Spain, extensive thrombosis in multiple coronary territories, and an increase in stent thrombosis during the COVID-19 pandemic have been reported. (2) Our purpose was to report a dramatic increase in stent thrombosis cases during the pandemic.

The first case was a male patient with a history of diabetes, renal failure, and non-ST-segment elevation myocardial infarction in July 2020, requiring implantation of 2 drug-eluting stents in the anterior descending artery (ADA). The patient progressed to bilateral pneumonia and was transferred to the Intensive Care Unit. Nasopharyngeal swab was performed, with a positive result for COVID-19. Later, the patient presented class IV angina and ST-segment elevation in the anterior wall. Stent thrombosis was confirmed, and balloon angioplasty, thrombus aspiration, intracoronary tirofiban, and 2 drug-eluting stent implants were performed, with positive technical results and clinical outcomes.

The second case was a 67-year-old male patient with a history of diabetes and active smoking, who presented with stress angina, and ischemia detected by gamma camera. In August 2020, the patient underwent catheterization, and a subocclusive lesion was found in the ADA and in the right coronary artery. Angioplasty with a sirolimus-eluting stent in the ADA, and angioplasty with two sirolimus-eluting stents in the right coronary artery on the third day were performed with positive outcomes. Eleven days after discharge, the patient was readmitted for sudden-onset angina at rest. ST-segment elevation was detected in the anterior wall; the patient was swabbed as per institutional protocol, and was then admitted to the interventional cardiology unit. Proximal stent thrombosis of the ADA was diagnosed. Balloon angioplasty was performed and partial vessel opening was achieved. However, the patient developed complex ventricular arrhythmia and ventricular fibrillation, and died in the general ward. Twelve hours after his death, the swab result was positive for COVID-19.

The third case reported was a 62-year-old male patient with a history of diabetes, who was admitted for non-ST elevation acute coronary syndrome. Coronary angiography revealed a big and dominant circumflex artery with severe diffuse disease. Angioplasty with 4 drug-eluting stents was successfully performed. The patient was swabbed as per institutional protocol and the result was positive for COVID-19. Postoperative course was good, and the patient was discharged. Six days later, he presented an episode of severe angina at rest. He went to another hospital, where ST-segment elevation in the inferior wall was detected, and died before undergoing catheterization.

In the reports and case series before the COVID-19 pandemic, the rate of coronary stent thrombosis worldwide was around 1% and 2.5%. (3)

The three cases reported were treated in centers with a high number of cardiac patients, a hemodynamic team with a high rate of monthly angioplasties and experience in its practice, and a rate of stent thrombosis of 0.67% in June-July, 2019. Globally –and also in our setting–, acute myocardial infarctions and elective angioplasties decreased between 40% and 80%, without specifying the mechanisms that measured this reduction, probably attributed in part to an excessive number of heart attacks and deaths at home not reaching health centers due to the pandemic. (4) In this context, the 3 cases of stent thrombosis in the period of June-July 2020 in COVID-19 positive patients represent an increase from 0.67% (1/148) to the current 5.66% (3/56) of all angioplasties (odds ratio 8.32, 95% CI 0.84-81; p [Fisher] = 0.0638). Despite the limitation of the low number of subjects analyzed, the results are alarming. Moreover, of the 6 patients positive for COVID-19 undergoing angioplasty due to coronary events, 3 had stent thrombosis and 3 presented events on stable plaques or atherosclerotic le-

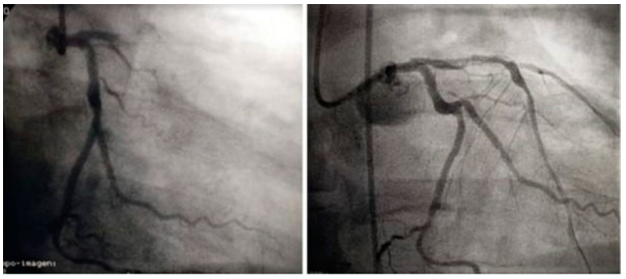

Fig. 1. Stent thrombosis in the anterior descending coronary artery on admission

Table 1. Characteristics of the three cases

	CASE 1	CASE 2	CASE 3
Age	55	67	62
Sex	Male	Male	Male
CRF:	HTN - DM	DM - Ex SMK - HTN	HTN - Ex SMK - DM - Overweight
Previous PTCA	July 2020 - ADA	August 2020 - ADA & RCA	July 2020 - CXA
Implanted stent	2 biodegradable polymer-based DES (Biolimus)	Sirolimus DES 3.5 × 33 2.5 × 23 – 3.5 × 18 mm	Everolimus DES x 4: 2.5 × 32 – 2.5 × 20 – 3.0 × 24 – 3.5 × 18 mm
Antiplatelet drug	ASA - Clopidogrel	ASA - Clopidogrel	ASA - Clopidogrel
Clinical COVID-19 codition	Bilateral pneumonia	Asymptomatic	Asymptomatic
COVID-19 diagnosis	PCR swab test +	PCR swab test + (sample taken on last admission)	PCR swab test + (sample taken as per protocol during hospitalization for PCI)
New coronary condition	Previous AMI	Previous AMI	Inferolateral AMI
CA	ADA Stent thrombosis	ADA Stent thrombosis	-
Time from previous PTCA	15 days	11 days	6 days
Treatment	Balloon thrombus aspiration Tirofiban 2 Sirolimus DES implanation	Balloon thrombus aspiration	-
Antiplatelet agent	ASA – Ticagrelor	-	-
Evolution	Good	Intraprocedural death	Death before admission to the catheterization laboratory

CRF: Cardiovascular risk factors. HTN: Hypertension. DM: Diabetes mellitus. SMK: Smoker. ASA: Aspirin ADA: Anterior descending artery. RCA: Right coronary artery. CXA: Circumflex artery. PTCA: Percutaneous transluminal coronary angioplasty. DES: Drug-eluting stent. AMI: Acute myocardial infarction. CA: Coronary angiography.

sions (personal data not previously published). Since no intracoronary ultrasound was performed to assess the stent thrombosis, the correct stent position and expansion cannot be confirmed, nor can previous hidden coronary dissections. Neither can it be confirmed that there has been intolerance to clopidogrel, polymorphisms associated with clopidogrel non-responders, or even that the three patients have omitted medication doses after hospital discharge. If our suspicion is confirmed, we should consider whether prasugrel or ticagrelor are more effective than clopidogrel for COVID-19-positive patients, whether we should increase the doses, whether all antiproliferative drugs in the stents interact in the same way with hypercoagulability associated with COVID-19, whether these phenomena observed in coronary stents are reproduced in stents implanted in other territories, (5) and many other questions for which not only do we have no answers, but we do not even imagine the questions.

Conflicts of interest

None declared.

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

Ethical considerations

Not applicable.

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Heart Failure Due to External Iliac Arteriovenous Fistula After Laser Ablation of the Saphenous Vein

Chronic venous failure caused by varicose veins is a common medical condition in adults. A meta-analysis demonstrated that endovenous laser ablation was more effective in reducing the rate of complications compared with conventional surgery. (1) However, complications are different, including nerve injury, thrombophlebitis, superficial burns, and arteriovenous fistulas (AVF).

A 78-year-old male patient with a history of hypertension and endovenous laser ablation (1,470 nm diode) of the left internal saphenous vein due to severe venous failure 8 years ago, was admitted to the emergency room for functional class (FC) IV dyspnea, with anasarca and edema of both lower limbs, and a diagnosis of heart failure. During questioning, the patient referred two hospitalizations due to the same clinical presentation in the last 6 months in another medical center, with a positive response to the medical treatment.

Physical examination revealed systo-diastolic murmur with thrill in the left inguinal region. The Echo-Doppler reported an external iliac arteriovenous fistula with 6 mm neck diameter, high-velocity and low-resistance flow pattern, without associated venous thrombosis in the deep and superficial venous system. The echocardiogram showed a 57 mm left ventricular diastolic diameter, 64% EF and moderate diastolic dysfunction, enlarged right ventricle, TAPSE 17, dilated pulmonary artery trunk, dilated inferior vena cava of 33 mm diameter and 9% collapse, bilateral pleural effusion, and ascites. A CT angiography confirmed the fistula between the artery and the left external iliac vein adjacent and immediately proximal to the inguinal ligament, with a diameter of about 6.2 mm, determining proximal dilation of the ipsilateral common iliac vein (Figure 1). Since the patient was in anasarca, and considering the high risk and the vascular anatomy, endovascular treatment was decided, and a 16 mm - 10 mm × 7 cm endoluminal Excluder® iliac extension was implanted in the left external iliac artery under neuroleptanalgesia and by

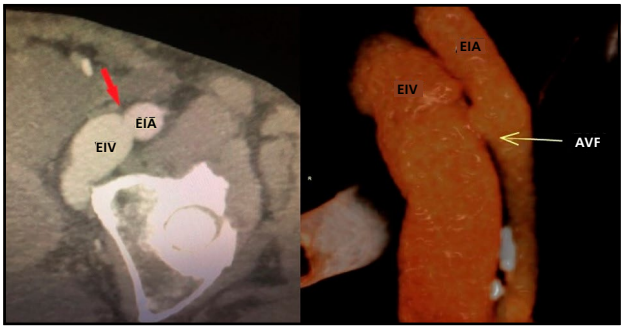

Fig. 1. CT angiography: arteriovenous fistula (AVF) with dilation of external iliac (EI) vessels.

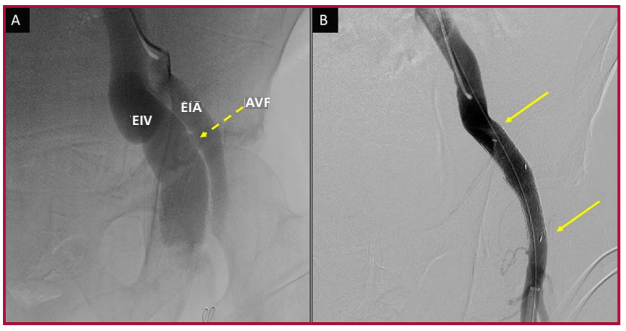

Fig. 2. A. Arteriography with arteriovenous fistula (AVF) in external iliac (EI) vessels. **B.** Complete AVF exclusion following covered stent implantation

superficial femoral access, achieving the complete exclusion of the A-V fistula between the artery and the left external iliac vein (Figure 2). The patient made good progress and was discharged under clopidogrel 75 mg, aspirin 100 mg, enalapril 5 mg, and bisoprolol 5 mg/d. During the first month, anticoagulation with 5 mg/d apixaban was indicated to prevent deep vein thrombosis due to the sudden decrease in the high venous flow of the pelvic and intra-abdominal collateral veins. At the 12-month follow-up, the patient was in FC I, and with absence of murmur and thrill in the left inguinal region.

Endovenous laser ablation is a percutaneous treatment under local tumescent anesthesia, in which distal saphenous vein cannulation is performed under ultrasound guidance, and a laser fiber is inserted. The laser energy heats the blood and vessel endothelium, causing thrombotic and fibrotic occlusion of the treated vein. Tumescent anesthesia not only makes the procedure tolerable for the patient, but also protects the structures adjacent to the saphenous vein, since it physically separates the tissues and also acts as a heat sink, avoiding thermal damage. Furthermore, this type of anesthesia compresses the vein, thus reducing its diameter and increasing the effectiveness of the treatment.

Arteriovenous fistula formation after endovenous thermal ablation is an extremely rare complication, and has been described in the early or late postoperative period, both in the treatment of internal or external saphenous vein. In patients undergoing endovenous thermal laser ablation, Theivacumar et al. (2) reported < 0.2% development of arteriovenous fistula, while Rudarakanchana et al. (3) reported this complication in 0.15% of cases in 2,500 procedures analyzed after venous thermal ablation in the same medical center, mainly after laser ablation (73%), and the rest with radiofrequency. Most authors describe small, asymptomatic arteriovenous fistulas, diagnosed as a control Echo-Doppler finding, which resolve spontaneously. (2, 3) Two possible mechanisms of arteriovenous fistula formation are considered during venous laser ablation: the concomitant venous and arterial

injury with the needle during administration of tumescent anesthesia, and the thermal energy transfer with the laser probe that could cause degradation and perforation of the vein wall with arterial injury.

Very few cases of arteriovenous fistula in the external iliac system are reported in the literature, probably caused by the inadvertent advancement of the probe through the saphenous vein arch in the external iliac vein either with mechanical perforation through the vein into the artery or vessel wall damage due to activation of the laser while still within the external iliac vein. Lack of tumescent anesthesia at this level likely increases the risk of damaging the arterial wall. (4-5) Echo-Doppler and CT angiography are the non-invasive imaging methods to confirm the diagnosis, and allow for the characterization of the arteriovenous fistula (location, size and flow); an additional angiography is only required if an intervention is considered for marked lower limb edema, recurrent varicose veins, venous complications (phlebitis, ulceration), intermittent lower limb claudication from steal syndrome or heart failure. (2) The cases of external iliac arteriovenous fistula –including our case– were reported in the post-treatment of the internal or great saphenous vein, and all of them presented systo-diastolic murmur with thrill in the homolateral inguinal region to treatment. Diastolic heart failure was the most common clinical presentation, together with dyspnea, ascites, edema and lower limb heaviness with unilateral predominance, and occurred between 3 weeks and 2 years after surgery. (4, 5) Cases of heart failure and superficial femoral artery arteriovenous fistula have also been described after laser ablation of the saphenous vein. (6) All patients required either conventional surgery or endovascular treatment with covered or hybrid stent, depending on the anatomical proximity of the arteriovenous fistula to the inguinal ligament and the size of the fistula. In every case, symptoms were resolved, with disappearance of the murmur and inguinal thrill. (4, 5)

In summary, arteriovenous fistula is a rare complication of endovenous laser treatment, but it can develop high flows and cause heart failure in some patients. Therefore, the presence of iatrogenic arteriovenous fistulas should be ruled out in patients with signs and symptoms of heart failure and a recent or long-term history of saphenous vein failure treatment with laser ablation.

Conflicts of interest

None declared.

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

Ethical considerations

Not applicable

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Persistent ST-Segment Elevation Due to Myocardial Infiltration

In most cases, ST-segment elevation is due to acute coronary occlusion. However, atypical presentation or atypical evolution should make us consider other etiologies.

We present the case of a 71-year-old patient with persistent chest pain and shortness of breath. The electrocardiogram (ECG) showed ST-segment elevation in leads V2 through V4. A severe non-occlusive lesion was found in the distal left anterior descending artery and a stent was implanted, resulting in unsuccessful ST-segment normalization. Days later, symptoms recurred and a computed tomography (CT) scan revealed lung cancer with multiple metastases including the myocardium. ST-segment elevation in the ECG could indicate tumour invasion of the myocardium at the level of the left ventricle.

A 71-year-old man presented at the emergency department with persistent chest pain and shortness of breath. The ECG showed ST-segment elevation in leads V2 through V4 (Figure 1a). He had a history of arterial hypertension and smoking. He was referred to another hospital to undergo urgent revascularization. Coronary angiography showed critical narrowing at the distal segment of the left anterior descending artery (LAD), which was successfully treated with a drug-eluting stent (Figures 2a and 2b). Atypical evolution was observed, with persistent ST-segment elevation but without necrosis (no Q-waves [Figure 1b], no troponin el-

evation, no abnormalities in ECG). Symptoms recurred after 7 days (refractory chest pain, dyspnea and orthopnea). CT revealed primary lung cancer in the right upper lobe, myocardial interventricular septum infiltration (Figure 3) and metastases in bones and suprarenal glands. During this hospitalization, the ECG showed ST-segment elevations with a similar pattern, which was probably due to tumor invasion of the myocardium. Histopathological examination was not performed; however, CT findings suggested lung adenocarcinoma. Patient died one month after percutaneous coronary intervention (PCI), under palliative treatment.

Although acute myocardial infarction (AMI) is the most frequent cause of ST-segment elevation, it could occur in other circumstances, as in tumor with myocardial involvement. (1)

The most common underlying malignancies with secondary cardiac involvement include carcinoma of the lung, breast, esophagus, stomach, and kidneys, as well as melanoma, lymphoma and leukemia. Primary lung carcinomas account for around one third of cardiac metastases. (2, 3)

Myocardial metastasis from neoplastic disease is often clinically unapparent, and very difficult to diagnose. Of 151 consecutive autopsies of lung cancer patients, cardiac metastasis was found in 67 patients (44.4%), while myocardial metastasis was found in 11.9% of patients. ECG of patients with myocardial metastasis revealed ST-segment and T wave abnormalities and various types of arrhythmia. These alterations were observed in 4 patients with myocardial metastasis, and in 6 without this pathology (pericardial metastasis alone). (2) Case reports of ST-segment elevation without coronary occlusion have been described in different ECG leads depending on the infiltrated area of the heart. (2, 4, 5)

Patients with cancer and normal ECGs are unlikely to have cardiac metastasis. The ECG finding of myocardial ischemia or injury has high specificity (96%, p <0.000001) for cardiac metastasis. (4) On the other hand, ST-segment and T wave abnormalities are an unspecific finding of myocardial metastasis. (2)

Myocardial involvement by direct lung cancer invasion is unusual and is often clinically silent, although it can cause malignant pericardial effusion with or without symptoms of pericarditis, arrhythmias, heart failure, and rarely AMI. (2) AMI might be caused by tumor embolization or direct compression of the coronary arteries. (5)

The present case was initially misdiagnosed as acute coronary syndrome with ST-segment elevation due to the presence of cardiac pain and ST-segment elevation on ECG. However, after fatal evolution of the patient in one month, we should conclude that the abnormal ST-segment elevation in this patient was not a manifestation of AMI, but possibly caused by an alteration in the myocardial electrical properties associated with tumor invasion. Moreover, the presence of normal cardiac enzymes was also an important evidence that the ECG abnormalities were not associated with either AMI or myocarditis. (6)

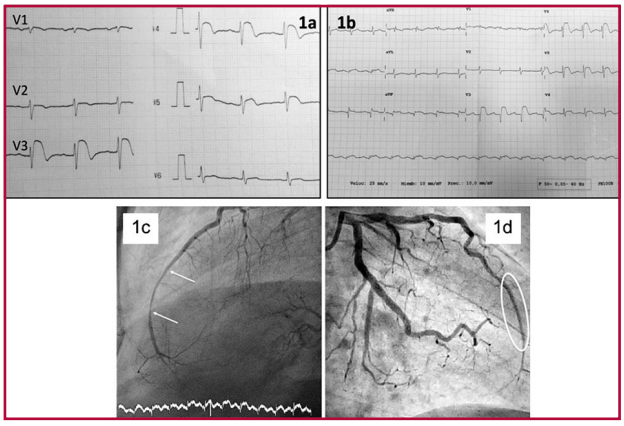

Fig. 1. 1a. Baseline ECG: ST-segment elevation in leads V2 to V4. **1b.** Discharge ECG: ST-segment elevation in leads V3 to V6. Q-wave in V3. Minimal ST-segment elevation in leads D2-D3-avF. **1c.** Left anterior descending artery angiography before treatment. **1d.** Image following stent implantation.

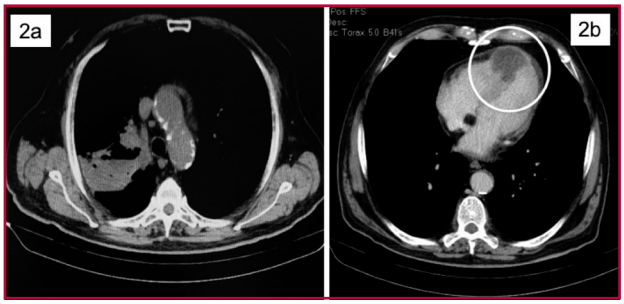

Fig. 1. Computed tomography. **2a.** Lung cancer in the right upper lobe. **2b.** Myocardial interventricular septum and apex infiltration.

Conflicts of interest

None declared.

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Ethical considerations

Not applicable

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Short QT Syndrome Associated with Noncompacted Myocardium

Cardiomyopathies are a heterogeneous group of myocardial diseases associated with mechanical or electrical dysfunction that usually (but not invariably) exhibit inappropriate ventricular hypertrophy or dilation and are due to a variety of frequently genetic causes. Cardiomyopathies are confined to the heart or are part of generalized systemic disorders, which may lead to cardiovascular death or progressive heart failure-related disability. (1)

Noncompacted myocardium is a primary genetic cardiomyopathy first described in 1984 by Engberding, characterized by myocardial hypertrabeculation with sinusoids in communication with the ventricular cavity, resulting from a sudden arrest in the normal embryogenesis prior to compaction, and involving predominantly the distal portion of the left ventricle. (2)

Short QT syndrome (SQTS), introduced as an association with sudden death (SD) in 1993 by Algra, but first described by Ghusack in 2000, who reported 6 unrelated family cases with atrial fibrillation as the form of presentation, is a primary cardiomyopathy described within the channelopathies generating electrical dysfunction. (3) Diagnosing SQTS is not easy, since in a first instance, numerous studies associated corrected QT interval (QTc) <300 ms with SD, but then, the limit point proposed was two standard deviations from the normal QTc (350 ms), so the cut-off point to consider a short QT interval would be 320 ms. In 2011, Gollob et al. proposed a diagnostic score based on QTc, the JT interval, the genotype, and the patient's medical record and his/her family history, assigning points to each category. An overall score of 4 points or greater indicates a high-probability of SQTS, 3 points an intermediate probability and 2 points or less a low probability of SQTS. (4) In 2015, the European Society of Cardiology (ESC) standardized criteria and determined that the presence of QTc <340 ms (Class I, level of evidence C), or a QTc <360 ms with at least one associated genetic mutation, family history of SQTS, family history of SD before the age of 40, or survival from an episode of ventricular fibrillation (VF) or documented ventricular tachycardia (VT) in the absence of structural heart disease (Class IIa C) constitute diagnosis of SQTS.

A 19-year-old male patient, without known personal pathological history, with a history of a 4th degree relative (cousin) who died of SD at the age of 1 year, and a 2nd degree relative (maternal uncle) who also died of SD at the age of 15 months, consulted for paroxysmal palpitations and dyspnea in FC II-III associated with an episode of chest pain. Sinus bradycardia at 40 bpm was detected, with adequate chronotropic response.

The ECG showed absolute QTc of 320 ms, and corrected by Bazett's formula of 292 ms (Figure 1). The echocardiogram revealed myocardial hypertrabeculation in the mid-apical region of the low and lateral walls, suggestive of noncompacted myocardium (Figure 2). The MRI reported preserved left ventricular size and function and normal myocardial wall thickness. Within the intraventricular cavity, abundant trabeculae in the lateral-apical region and apical segment were observed (Figure 1), with a relation between noncompacted and compacted areas of 2.4 (noncompacted area 16.5 mm, and compacted area 6.8 mm in a short axis view), meeting the diagnostic criteria for noncompacted myocardium (Figure 2).

The 24-hour Holter evidenced average HR of 45 bpm, minimum 33 bpm, maximum 94 bpm, without ventricular arrhythmia, maximum and minimum QTc of 375 ms and 276 ms respectively, and minimum absolute QT of 360 ms, with scarce daily variability despite high HR variability.

No familial phenotypes have been found in the screening for familial SQTS.

With the diagnosis of SQTS and the patient's consent, a cardioverter defibrillator (ICD) was implanted.

So far, SQTS associated with noncompacted myocardium has not been described.

Short QT syndrome was diagnosed according to the 2016 ESC guidelines for ventricular arrhythmias and prevention of SD due to QTc <340 ms (class IC). Based on the Gollob score, we have a patient with high probability of SQTS (4 points QTc <330 ms, and second-degree family history of SD) without performing genetic testing. The yield of genetic testing for SQTS is low (14%). (4)

So far, noncompacted myocardium has been described in association with other conditions, such as long QT syndrome; recently, a novel entity has been introduced, which identified the mutation in the HCN4 gene that is a phenotypic manifestation of sinus node dysfunction, noncompacted myocardium, mitral valve prolapse, and aortic dilation. Of all the family cases analyzed, no patients with short QT have been reported. (5)

Moreover, SQTS has been associated with Brugada syndrome and early repolarization syndrome, with sig-

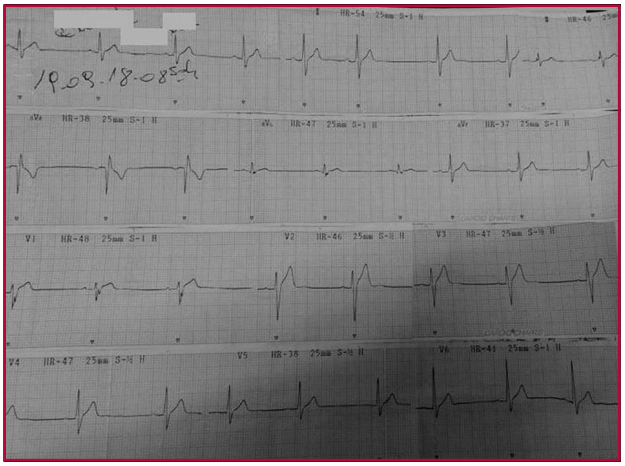

Fig. 1. Patient ECG showing heart rate of 50 bpm, absolute QT of 320 ms and a corrected QT by Bazett's formula of 292 ms.

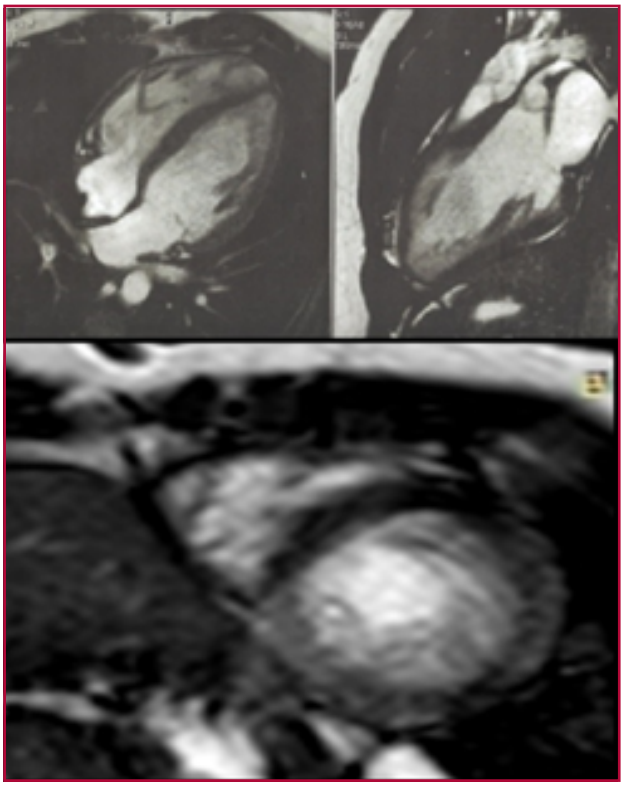

Fig. 2. Cardiac MRI. *Top left:* Four-chamber view; *on the right:* Two-chamber view of the left ventricle; both images show myocardial hypertrabeculation predominantly in the apical region. *Bottom:* Short axis view with hypertrabeculation, typical of noncompacted myocardium. Noncompacted/compacted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

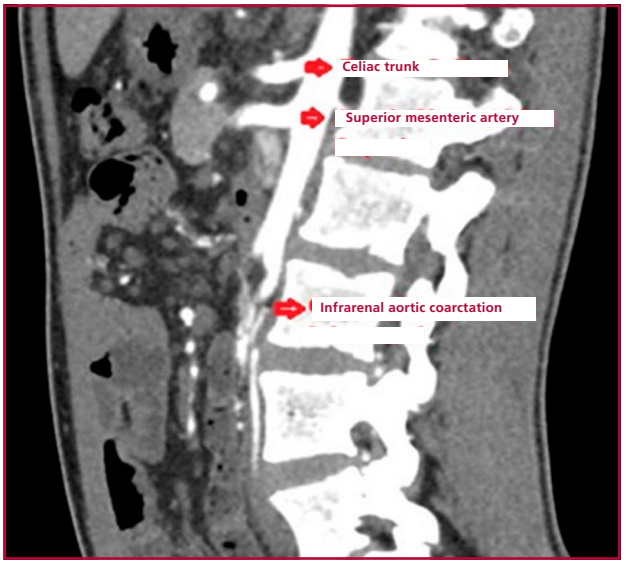

Fig. 2. Sagittal section of preoperative CT angiography showing infrarenal aortic coarctation

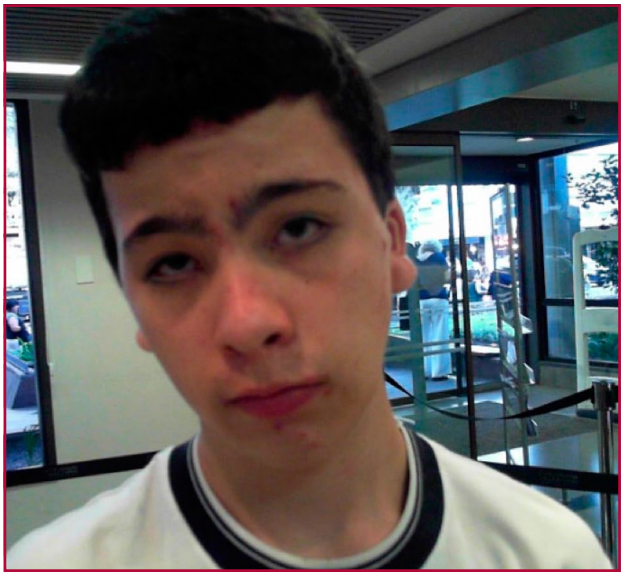

Fig. 1. Noonan syndrome phenotype

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)

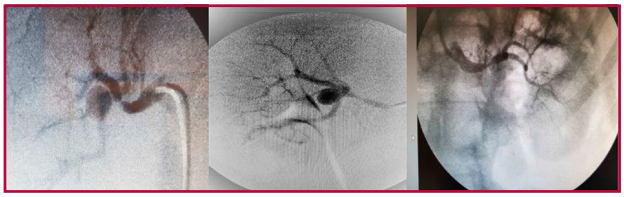

Fig. 1. (**A & B**): Selective arteriography of the right renal artery showing a saccular image with caudal pole adjacent to the main bifurcation of that artery, is a specific finding of a right renal artery aneurysm. (**C**): Left main renal artery stenosis.

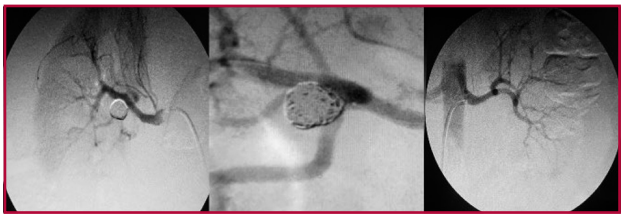

Fig. 2. (**A & B**): Selective arteriography of the right renal artery showing aneurysmal embolization with 5 Barricade coils. (**C**): Left renal artery angioplasty.

Associated complications including RA thrombosis, hematuria due to aneurysm rupture, renal infarction due to aneurysmal dissection and hypertension have been described. Aneurysmal rupture is rare (3%) but fatal. Rupture during pregnancy has a mortality rate of 50% for the mother. (3)

There are 2 registries: One is from the University of Michigan Hospital, in which the RAA were: 68% solitary, 31.5% multiple, and 19% bilateral, with average size of 1.5 cm. (4) Their presentation can produce flank pain in 15% of the patients, hematuria in 30%, and hypertension in 55%; the latter triggers the rupture in 90% of cases. The other is the ARCADIA Registry (Assessment of Renal and Cervical Artery Dysplasia), in which 48% of the cases had multisite FMD, and 20% had bilateral pathology. Among patients with a renal presentation, the prevalence of cervical lesions was higher in patients with bilateral than in those with unilateral RA lesions. (5) In our patient, results from the Doppler ultrasound of the neck vessels, abdominal aorta, and iliac, subclavian, brachial, radial, and bilateral ulnar arteries was normal.

Considerable controversy continues to surround the treatment of these aneurysms: specifically, what size of RAA needs surgery in asymptomatic patients; experts recommend repair in diameters ranging from 15 mm to 30 mm, although treating diameters > 20 mm is generally accepted.

Endovascular treatment is used, either direct aneurysm embolization, extended-release coils and stents with aneurysm embolization, or surgery only for complex lesions in case of large aneurysms with high risk of perforation or radiological signs of expansion. Nephrectomy is necessary only if the aneurysm is ruptured.

Renal angioplasty is the treatment of choice for renal artery stenosis due to FMD. (6) Other indications for revascularization include hemodynamically significant lesions, bilateral lesions, accelerated hypertension, and in patients with uncontrolled blood pressure despite drug therapy, as in our case.

Fibromuscular dysplasia should be considered in young women with the combination of aneurysm and renal artery stenosis. Renal artery angioplasty and endovascular exclusion of the aneurysm with coils should be the treatment of choice in these cases.

Conflicts of interest

None declared.

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

Ethical considerations

Not applicable

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