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CASE REPORT

Robert F. Bonvini · Aljoscha Rastan · Sebastian Sixt
Marc Righini · Roland Hofstetter · Thomas Zeller

Diffuse fibromuscular dysplasia successfully treated with scoring balloon angioplasty in a 3-year-old boy

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Abstract In children, up to 10% of the cases of arterial hypertension may be caused by a renovascular disease. The etiology of this renovascular disease is most of the time due to a fibromuscular dysplasia (FMD), which causes a noninflammatory intimal–medial fibroplasia leading to luminal compromise. Percutaneous transluminal angioplasty of FMD is a worldwide-accepted treatment modality for this serious arterial disease with, so far, good safety and long-term efficacy data. Once FMD involves several arterial compartments leading to symptoms the outcomes are poor. Herein we report the case of a 3½-year-old boy with severe arterial hypertension and abdominal angina due to a diffuse multivisceral FMD involvement, successfully managed by a percutaneous angioplasty approach using a new balloon catheter for plaque modulation.

Key words Percutaneous angioplasty · Fibromuscular dysplasia · Children · Renal artery · Mesenteric artery

Introduction

In children, arterial hypertension may be caused, in up to 10% of the cases, by renovascular diseases.[1] Most children presenting with renovascular hypertension have few if any symptoms, but if untreated, devastating complications (e.g., neurologic injury and congestive heart failure) may occur. The etiology of this juvenile renovascular disease is most of the time due to a fibromuscular dysplasia (FMD), followed by Takayasu arteritis, neurofibromatosis, and other rare syndromes.[1]

Fibromuscular dysplasia is a nonatherosclerotic, noninflammatory arteriopathy characterized by intimal, medial, and/or adventitial fibroplasias leading to luminal compromise, aneurysm formation, or arterial dissection. The pathological classification of fibromuscular lesions is based on the arterial layer in which the lesion predominates. Medial fibroplasia represents the most common dysplastic lesion, followed by intimal fibroplasia, which occurs in less than 10% of cases, and adventitial (periarterial) hyperplasia, which is the rarest type.[2] The natural history of renal FMD includes, in up to 37% of the patients, the progression of the angiographic disease (i.e., occurrence of new focal lesions, worsening of arterial stenosis), or the enlargement of a mural aneurysm.[3]

Fibromuscular dysplasia mostly affects the renal and the carotid arteries but it may involve several other muscular arterial compartments, including the celiac and the mesenteric arteries and the iliac arteries. The clinical presentation may vary from an asymptomatic condition to a multiorganic disease that mimics necrotizing vasculitis. This variety of symptoms depends on the arterial segment involved, the degree of stenosis, and the type of fibromuscular dysplasia. Usually, in the case of multiple symptoms, if FMD is not promptly treated the outcomes are poor.[4]

Because of the absence of specific symptoms, the diagnosis of FMD may be challenging and most of the time it remains speculative. However, the combination of the clinical history and the characteristic angiographic imaging are sufficient to suggest this pathology.

For nearly 30 years, percutaneous transluminal renal angioplasty (PTRA) has been used to treat renovascular diseases and the first described case of PTRA in a child was reported in the early 1980s.[5] Renal angioplasty, especially in the case of FMD, is nowadays the treatment of choice for renovascular hypertension,[6] and its efficacy and safety, also in children, has already been suggested in several reports.[7,8]

The percutaneous treatment of FMD is also described in all vascular fields.[9] However, to the best of our knowledge,

R.F. Bonvini (✉) · A. Rastan · S. Sixt · T. Zeller
Angiology Division, Heart-Center Bad Krozingen, Südring 15, 79189 Bad Krozingen, Germany
Tel. +49-7633-402-2431; Fax +49-7633-402-2439
e-mail: robert.bonvini@hcuge.ch

R.F. Bonvini · M. Righini
Angiology and Hemostasis Division, University Hospital, Geneva, Switzerland

R. Hofstetter
Pediatric Cardiology Division, University Hospital, Frankfurt a.M., Germany

this minimally invasive approach in more than one arterial sector, and especially in a child, has so far never been reported.

Here we report the case of a 3½-year-old boy with severe arterial hypertension and abdominal angina due to a diffuse multivisceral FMD involvement. The boy was successfully managed by a percutaneous angioplasty approach using a new balloon catheter for plaque modulation, with a 12-month favorable outcome thus far.

Case report

In December 2006, a 3½-year-old boy was found to have arterial hypertension (right arm: 148/52 [84] mmHg, left arm: 140/60 [87] mmHg) during a routine examination by his pediatric physician. After having excluded other possible etiologies of the arterial hypertension and following the discovery of a high peripheral blood renin level, a renovascular hypertension was suspected and finally confirmed by a selective renal and abdominal angiography which showed a diffuse multivisceral FMD involving both renal arteries, the superior mesenteric artery (SMA), and the splenic artery.

Under strict creatinine surveillance (i.e., potential risk of decreasing the glomerular filtration rate by giving angiotensin-converting enzyme inhibitors in case of renovascular hypertension), the pediatric physician initiated a regimen of enalapril 2.5 mg b.i.d. and metoprolol 12,5 mg b.i.d. With this antihypertensive combination, a few days thereafter the boy experienced postprandial abdominal pains, which were interpreted as an abdominal angina due to the newly developed relative hypotension. Despite having suspended the beta-blocker, the abdominal angina persisted and the blood pressure rose again (131/83 [99] mmHg). Finally the boy was sent to our tertiary center for the percutaneous management of this multivisceral FMD.

Intervention

The intervention was performed under general anesthesia. A 5-F Internal Mammary guiding catheter was selectively placed in the left renal artery. A 0.014-inch guidewire was then easily advanced through the main artery stenosis and a 2.0-mm coronary angioplasty balloon was utilized to dilate the most relevant stenosis. Due to an important elastic recoil at all dilated segments, despite several prolonged balloon inflations and the administration of intra-arterial nitroglycerine, we decided to utilize a 2.5-mm Scoring Balloon (AngioSculpt, AngioScore, Fremont, CA, USA), which with its special laser-cut nitinol spiral scoring elements could better "break" the membranous septa of the FMD stenoses, thus achieving a better final result (Fig. 1A,B). For the right renal artery and the SMA a Judkins Right guiding catheter was adopted and similarly to the left kidney, the most severe stenoses were initially treated with a 2.5-mm coronary angioplasty balloon. Again, an insufficient angiographic result was obtained. Thereby, all the lesions were additionally dilated with the same 2.5-mm Scoring Balloon (Fig. 2) for the renal artery and a 3.0-mm one for the SMA. Due to technical difficulties in catheterizing the truncus coeliacus and the already advanced procedural time, the splenic artery stenosis was not treated in this session. Femoral access hemostasis was obtained with a D-Stat Dry hemostatic bandage (Vascular Solutions, Maple Grove, MN, USA). After discharge, the boy's parents were instructed to give him Aspirin 50 mg for 4 weeks.

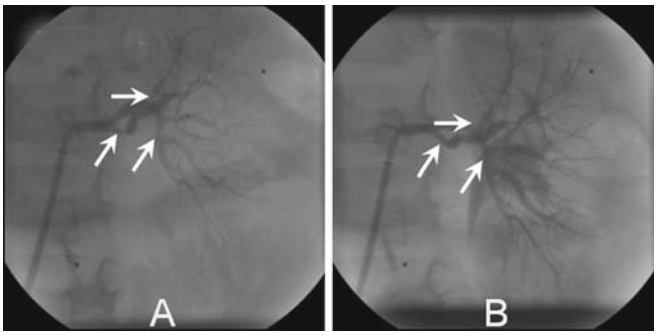

Fig. 1A,B. Left renal artery angiography before (**A**) and after (**B**) the 2.5-mm Scoring Balloon angioplasty (*arrows* show the dilated segments)

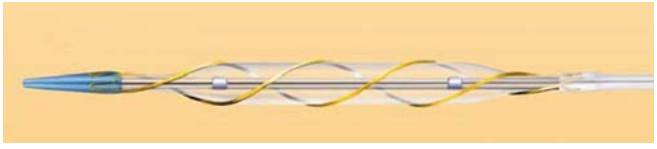

Fig. 2. Scoring Balloon with special laser-cut nitinol spiral scoring elements wrapping around a conventional nylon angioplasty balloon (AngioSculpt, AngioScore, Fremont, CA, USA)

At the 6-month follow-up visit blood pressure was 112/86 [95] mmHg, despite the fact that the boy did not take any antihypertensive medication and at 12 months, according to his age/height percentile interpretation, he had a persistent but still significantly improved systolic hypertension (121/63 [82] mmHg). The kidney function did not change significantly (pre-interventional: creatinine value: 0.2 mg/dl, at 6 months: 0.3 mg/dl), and the boy had so far no recurrence of abdominal angina.

Discussion

Fibromuscular dysplasia of the renal arteries is usually suspected in young patients presenting with severe or newly developed arterial hypertension.[1] This case highlights how FMD can be a diffuse disease involving more than one organ, and how the conservative management of this multivisceral disease can be difficult when multiple symptoms are already present.[4]

Despite the young age of the patient, the vicious circle of abdominal angina due to antihypertensive drugs, which could not be suspended, mandates the attempt of this off-

label percutaneous renal and mesenterial angioplasty. In the case of FMD renal lesions, PTRA is the first-line treatment and stenting should be reserved only as bailout procedure in case of a suboptimal result after balloon angioplasty or renal artery dissection. In our patient insufficient angiographic results with severe elastic recoil formed the rationale to continue the intervention with a lesion modulating catheter device like a Scoring or a Cutting Balloon. Due to a better crossing profile (<2.7 F of the distal shaft) compared to standard Cutting Balloons and very small balloon diameters (from 2.0 up to 5.0 mm), Scoring Balloons are probably the best alternative after PTRA failure, and this especially by treating small FMD renal arteries as observed in children.

As recently shown in different trials, a significantly low dissection rate compared to standard balloon angioplasty (~13% vs ~30%), no device slipping for more accurate placement, and no "geographic miss," are other advantages of this new Scoring Balloon technology.[10,11] These points are very important to take into consideration especially in performing percutaneous interventions in children, in whom stenting should be avoided because of the theoretical future stent-undersizing due to the continuous growth of the vessel diameter during childhood. Because FMD is secondary to an intimal–medial arterial fibroplasia, angioplasty alone, especially if performed with a Scoring or a Cutting Balloon, is most of the time sufficient to break these fibrodysplastic membranes, thus assuring a near normal vessel lumen with an acceptable long-term patency rate.[6,12]

Even if children's FMD treated by PTRA is already largely reported,[7,8] to the best of our knowledge, this is the first described case where a successful angioplasty using a plaque modulation technology was performed on a 3½-year-old boy, in both renal arteries and in the mostly diseased branches of the SMA. Furthermore, the mid-term favorable outcome (no recurrence of hypertension or abdominal angina) suggests the safety and efficacy of this procedure also in children.[10,11]

In conclusion, our case confirms that even in young children affected by symptomatic multivisceral FMD, percutaneous arterial angioplasty is technically feasible, safe, and efficacious in terms of symptoms regression, thus suggesting that angioplasty should be rapidly proposed before the affected organs are irreversibly damaged. Furthermore, in case of immediate angiographic failure with standard balloon angioplasty, Scoring Balloons offer a valid and efficacious solution to better modulate these FMD lesions where, especially in children, stent implantations are relatively counter-indicated.

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