Neurofibromatosis type I associated with abdominal aorta and renal artery stenosis: a case report

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I have the following potential conflicts of interest to report:

- Consulting
- Employment in industry
- Stockholder of a healthcare company
- Owner of a healthcare company
- Other(s)

X do not have any potential conflict of interest
Introduction

- Neurofibromatosis type 1 (NF1), also called von Recklinghausen's disease, is an autosomal dominant genetic disorder.

- It may present with arterial lesions, most commonly stenosis or aneurysmal formation of the aorta at the level of the renal arteries.
Case presentation

- A 4-year-old girl, diagnosed with NF1 since she was 14 days old, was admitted to our hospital on April 2015 due to hypertension.

- The patient presented multiple café-au-lait spots throughout the trunk and limbs, two big spots in her left thigh and unequal lower limbs.

- No other abnormalities except a heart murmur (2/6)
Case presentation

- Her systolic blood pressure was 125 mmHg, diastolic 83 mmHg, and her pulses were 99 bpm.

- The blood pressure screening included a 24-hour holter which indicated increased systolic and diastolic blood pressure during day and night measures.

- Brain MRI/MRA revealed a node in the middle cerebellar peduncles, characterized as possible hamartoma.
Case presentation

- Absence of bilateral femoral pulses and bilateral claudication.
- Creatinine level (0.58 mg/dl).
- **MRA of the abdomen** revealed aortic stenosis at the level of the superior mesenteric artery.
- Left ostial renal artery stenosis.
- Collateral circulation between superior and inferior mesenteric artery (artery of Riolan).
INTERVENTIONAL PROCEDURE

- Femoral artery cut down.
- Lesion angioplasty with MONORAIL balloon.
- 5mm diameter and 2cm length for the aortic lesion.

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<th>suprarenal</th>
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<tr>
<td><strong>1\textsuperscript{st} PTA</strong></td>
<td>4 atm for 20 s</td>
<td>4,5 atm for 90s</td>
<td>5 atm for 60s</td>
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<tr>
<td><strong>2\textsuperscript{nd} PTA</strong></td>
<td>14 atm for 80 s</td>
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<td><strong>3\textsuperscript{rd} PTA</strong></td>
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<td>15 atm for 75s</td>
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- Completion intraop. angio: good aortic result with left renal artery stenosis.
Postoperative Result

• Bilateral femoral pulses
• Dual Antihypertensive Medication
• Creatinine level (1.07 mg/dl)
Six-month postoperative MRA
Discussion

- The cardiovascular spectrum of NF1 includes stenotic and aneurysmal lesions of aorta and renal arteries, congenital heart disorders and secondary hypertension.

- Renal artery and aortic stenosis, is the most common cause of renovascular hypertension in young children and treatment rarely reported in the literature.
Discussion

- Endovascular treatment with simple balloon angioplasty in children seems to be an acceptable approach.
- Nevertheless dedicated devices are not available.
- Long-term follow up is necessary.