The young girl with renovascular hypertension of unknown origin

P. Manunta¹, C. Lanzani¹, R. Chiesa², R. Castellano², A. Ianello², G. Melissano², M. Venturini³, A. Del Maschio³ and D. Cusi¹

¹Chair of Nephrology, Dialysis and Hypertension. University of Milan, ²Division of Vascular Surgery, ³Division of Radiology, San Raffaele Hospital, Milan, Italy

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Case Report

A 16-year-old girl was admitted to the Division of Nephrology at San Raffaele Hospital in March 1995 with severe hypertension (170/120 mmHg). Fifteen days earlier she had been found by the school physician with high blood pressure values (160/110 mmHg) after an epistaxis. Twenty-four-hour ambulatory blood pressure monitoring (ABPM–SpaceLabS 90207) confirmed the presence of elevated blood pressure values (daytime SBP 170/DBP 123 mmHg; night-time SBP 143/DBP 92 mmHg). Six months before, the same school physician had found normal blood pressure values.

The family history was negative for essential hypertension, cardiovascular diseases, or other familiar-related diseases. Physical examination revealed that all the pulses were present. The blood pressures in the right and left arms were similar (170/120 and 167/122 mmHg respectively). There were no abnormal findings on physical examination, in particular there was no abdominal bruit. Both the neurological and ophthalmological examinations were negative.

The results of laboratory analyses showed: serum creatinine concentration 0.63 mg/dl (n.v. 0.7–1.25 mg/dl), with body mass index 17 kg/m². The urinalysis was negative for proteinuria, haematuria, and urinary infections. There was no evidence for systemic vasculitis by history, serologies, or other laboratory data. The thyroid function, the corticosteroid hormones, sex hormones, and urinary catecolamine were normal. Blood samples showed an elevated erythrocyte sedimentation rate (21 mm/h, n.v. 0–10 mm/h), a decrease of complement fraction C4 (15 mg/dl, n.v. 20–47 mg/dl) with normal values of C3 and the presence of anticardiolipin antibody (IgM 27 IU/ml, n.v. 0–12.5 IU/ml). Plasma renin activity (PRA) and serum aldosterone in supine and standing positions were increased (PRA supine 4.2 ng/ml/h with n.v. 0.2–2.8 ng/ml/h; PRA standing 8.0 ng/ml/h with n.v. 1.5–5.7 ng/ml/h; serum aldosterone supine 230 pg/ml with n.v. 30–150 pg/ml; serum aldosterone standing 393 pg/ml with n.v. 70–350 pg/ml), in spite of normal values of both serum and urinary sodium and potassium in two collections (142.2 mEq/l and 4.14 mEq/l respectively).

To investigate the presence of vascular and/or renal abnormality, we performed a magnetic resonance angiography of the abdomen and found a proximal stenosis of both renal arteries with parietal thickening and luminal stenosis of submesenteric trait of abdominal aorta (approximately 3 cm long). Arteriographic study confirmed the presence of the bilateral proximal stenosis of the renal arteries and involvement of the abdominal aorta (Figure 1). The radiological aspect of proximal critical stenosis was suggestive for Takayasu’s syndrome. Other segments of the aorta such as the aortic arch and major arteries were studied during the angiographic examination and were found not to be involved.

The diagnosis of Takayasu’s syndrome was based on clinical and radiological criteria determined by the Aortitis Syndrome Research Committee of Japan [1]. Gail et al. [2] proposed the ‘Criteria for active disease in patients with Takayasu’s arteritis’ for the diagnosis of new onset of TA: (1) systemic features (no other cause identified), (2) elevated ESR, (3) typical angiographic features, (4) vascular ischaemia; the minimum required for the diagnosis is two of the above criteria. In our case the diagnosis of activity of Takayasu’s syndrome was based on the presence of three of the four criteria (hypertension, elevated ESR, and typical angiographic features). Vascular ischaemia may be considered positive since the kidney were clearly ischaemic (high PRA values).

The patient was then treated with oral glucocorticoid (prednisone) at a dose of 1 mg/kg/day. One month later ESR was back to normal values (13 mm/h).
According to previously reported criteria the disease was considered inactive since the ESR was back to normal and a percutaneous transluminal renal angioplasty (PTRA) was successfully performed in the right renal artery, restoring renal vascular flow. In the left renal artery the PTRA was unsuccessful. The blood pressure values remained high after PTRA. To preserve the renal function in the left kidney we decided to perform an arterial reconstruction. During the surgery the involvement of both aorta and renal artery was found to be more severe than shown by the arteriography. The involvement of the arterial tree prevented any anastomotic procedure. The only possibility was to perform a heterotopic renal autograft transplantation in the pelvis to the internal iliac artery, using autologous saphenous vein inverted for reconstruction of the renal artery (Figure 2). MR angiography performed a few days later showed that both the renal arteries were patent with a normal conformation and normal vascularization of both kidneys. In the following days the blood pressure values decreased but did not reach normal values (24 h monitoring daytime SBP 145/DBP 98 mmHg; night-time SBP 132/DBP 86 mmHg). We related this mild hypertension to the glucocorticoid treatment. Angiotensin-converting enzyme inhibitor (enalapril 10 mg), and antiaggregant therapy were started. The blood pressure reached normal values in 5 days (130/80 mmHg) while renal function did not change. The patient was dismissed from the hospital a week later.

Renal function, blood pressure, and ESR were measured every month and remained normal for 5 months. We then decided to discontinue oral prednisone treatment, but during the tapering of glucocorticoids the erythrocyte sedimentation rate increased again. We then reintroduced a minimal oral dose of prednisone (5 mg/day). With such a low dose of steroid the antihypertensive therapy was no longer necessary. MR angiography performed 7 days after the surgical treatments, and angiography performed 6 months and 1 year later, confirmed the patency of renal arteries (Figure 3) with no radiological sign of Takayasu’s syndrome activity.

Discussion

The prevalence of secondary hypertension in young people is directly related to blood pressure level and inversely related, to age as reported in several epidemiological studies [3]. Thus the finding of severe hypertension in a young girl requires a thorough medical evaluation to exclude the presence of secondary hypertension. The major cause of hypertension in Western young people is renal parenchymal diseases (43–84%) [4]. In our case the renal function and the urinalysis were normal, as well all other investigations aimed at

**Fig. 1.** Angiography of renal arteries before treatment. IADSA shows severe bilateral stenosis of both renal arteries with poststenotic dilatation (minor in the right renal artery).

**Fig. 2.** Surgical image of left kidney after the autograft transplantation: anastomosis between left renal artery and internal iliac artery using autologous saphenous vein inverted. The polar superior artery has been clamped.
Nephrovascular hypertension in Takayasu syndrome

Since the risk of surgical procedure failure is increased during active disease [7,8] we treated the active phase of Takayasu’s arteritis with glucocorticoid for 30 days, as recommended in the literature [2,9]. During this period the blood pressure values remained high, but we did not start any antihypertensive treatment since the high systemic blood pressure could be of help in maintaining some renal blood flow in spite of the tight renal artery stenosis. The response to steroid treatment was excellent, with normalization of ESR. The titres of anticardiolipin antibody decreased and the C4 increased, but both of them remained abnormal.

In a recent study of the mid-term results of PTRA in children with Takayasu’s syndrome the procedure was technically successful in 95% of stenoses and the predicted cumulative patency rate at 5 years was 71% [5]. The authors observed that the presence of a long stenosis beginning at the origin of the artery (as in our case) predisposes to restenosis. After 1 month of steroid treatment PTRA restored renal vascular flow in the right renal artery. On the left side renal autograft transplantation in the pelvis was the only technically possible alternative. Beale et al. [9] reported nine cases, five of which were successful. The internal iliac artery was selected since it is infrequently affected by TA.

As one can see in Figure 3 PTRA on the right renal artery was not morphologically perfect, although the blood flow is normal. The patient will routinely undergo MR angiography every 6 months to evaluate the status of the renal arteries and to be able to undertake any needed technical correction as soon as possible.

The following conclusion can be drawn:

Takayasu’s syndrome must be considered in the differential diagnosis of renovascular hypertension in young individuals.

Bilateral stenoses are common in Takayasu’s disease. During the acute phase of the disease anti-inflammatory treatment is a priority and surgical treatment should be deferred.

In selected cases PTRA and autologous transplantation are sensible approaches to treatment.

References