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Kasuistika | Case report

Midaortic syndrome in a young man: Case presentation

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ARTICLE INFO

Article history:
Received: 18. 1. 2016
Received in revised form: 25. 4. 2016
Accepted: 27. 4. 2016
Available online: 21. 5. 2016

Klíčová slova:
Aortografie
Hypertenze
Syndrom střední aorty

SOUHRN

Koarktace aorty je vrozené zúžení aorty, které je obvykle diagnostikováno v dětství. U menšího procenta pacientů je tato malformace zjištěna v dospělosti na podkladě pozdních komplikací, např. systémové hypertenze nebo ruptury intrakraniálního aneurysmatu. Popisujeme případ jednatřicetilého muže bez významné anamnézy, který se dostavil k lékaři pro bolesti hlavy a rostoucí únavu. Fyzikální vyšetření prokázalo vysoký krevní tlak a slabý femorální puls v obou dolních končetinách. Ultrazvukové vyšetření břicha zjistilo stenózu břišní aorty za odstupem celiakální větve. Aortografie potvrdila koarktaci i uzávěr horní mezenterické tepny spolu s aneurysmatem pravé renální tepny. Hypertenze byla léčena inhibitorem angiotensin-konvertujícího enzymu. Protože došlo k úpravě krevního tlaku použitou medikací a pacient nevykazoval ani digestivní symptomy, ani klaudikace, byla chirurgická revaskularizace odložena a místo ní bylo rozhodnuto o dalším sledování pacienta.

Keywords:
Aortography
Hypertension
Midaortic syndrome

ABSTRACT

Aortic coarctation is a congenital narrowing of the aorta, usually diagnosed during childhood. In a minority of cases, this malformation is detected during adulthood, by late complications such as systemic hypertension or intracranial aneurysm rupture. We present the case of a 31-year-old man, without past medical history, who presented for headache and increasing fatigue. The physical exam revealed high blood pressure values and weak femoral pulses in both legs. Abdominal ultrasonography identified stenosis of abdominal aorta, after the origin of the celiac trunk. Aortography confirmed the coarctation and revealed also an occlusion of the superior mesenteric artery, with an aneurysm of the right renal artery. The hypertension was treated with an angiotensin converting enzyme inhibitor. Because the blood pressure was controlled with medication and the patient did not have symptoms of digestive claudication, the surgical revascularization was postponed and monitoring was preferred in this patient.

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DOI: 10.1016/j.crvasa.2016.04.007

Please cite this article as: C.C. Diaconu, Midaortic syndrome in a young man: Case presentation, Cor et Vasa 59 (2017) e171–e173 as published in the online version of *Cor et Vasa* available at http://www.sciencedirect.com/science/article/pii/S001086501630039X

Introduction

Aortic coarctation is a congenital malformation of the aorta. Most often, the diagnosis is done during childhood, with subsequent surgical correction. Sometimes, it may remain undetected until adulthood. The clinical manifestations are arterial hypertension during childhood, lower extremity muscle weakness, delayed or diminished lower extremity pulses and/or signs of congestive heart failure. The physical exam may raise the suspicion of aortic coarctation, but imaging investigations are necessary in order to evaluate the exact morphology of the malformation and to diagnose other associated malformations, such as aneurysms.

Case presentation

A 31-year-old man, with unremarkable previous medical history, was referred by the family doctor to our hospital, due to increasing fatigue and headache. He had been well until one month previously. Physical exam at presentation identified high blood pressure values (170/100 mmHg in both arms), a heart rate of 78 beats/min, femoral pulses palpable bilaterally, but weak and delayed compared to the brachial pulses. The rest of the examination was within normal limits. ECG and echocardiography were normal. Abdominal ultrasound identified stenosis of abdominal aorta, after the origin of celiac trunk. Aortography showed an eccentric significant stenosis (80%) of abdominal aorta (minimum diameter 5 mm), 55 mm in length, extended from the celiac trunk to 23 mm below the emerging of the renal arteries (with pressure gradient 40 mmHg); the celiac trunk permeable, without stenosis. The superior mesenteric artery was occluded at the ostium, with origin in the aortic stenosis and late charge by Riolan's arcade and pancreatico-duodenal branches from the celiac trunk. Also, aneurysm of 14/8 mm, bilobed, with broad base of implantation, on the upper wall of the right renal artery (Fig. 1). The structure and function of the kidneys were normal (Fig. 2). Antihypertensive therapy with an angiotensin converting enzyme inhibitor (perindopril) was initiated, with good control of the blood pressure. Despite the fact that the superior mesenteric artery was occluded, the patient did not have symptoms of digestive claudication. The surgical revascularization will be difficult and complex in this patient, the superior mesenteric artery and both renal arteries (left and right) having origin in the aortic stenosis. Because the blood pressure was controlled with medication and the patient did not have symptoms of digestive claudication, the surgical revascularization was postponed and monitoring was preferred.

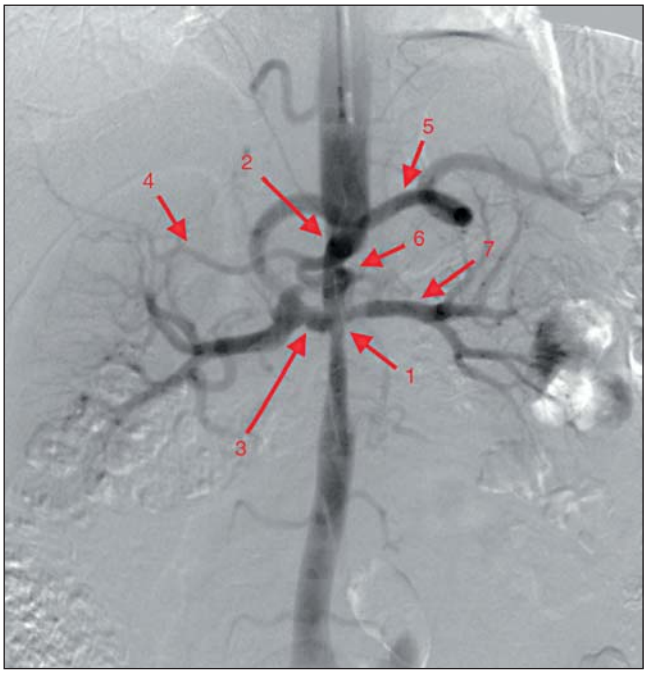

Fig. 1 – Aortography. Eccentric significant stenosis (80%) of abdominal aorta (arrow 1), extended from the celiac trunk below the emerging of the renal arteries, the celiac trunk permeable, without stenosis (arrow 2). The superior mesenteric artery occluded at the ostium, with origin in the aortic stenosis and late charge by Riolan's arcade and pancreatico-duodenal branches from the celiac trunk. Aneurysm on the upper wall of the right renal artery, with broad base of implantation (arrow 3). Arrow 4 – hepatic artery, arrow 5 – splenic artery, arrow 6 – gastroduodenal artery, arrow 7 – left renal artery.

Fig. 2 – Abdominal MRI. Kidneys with normal structure.

Conclusion

Mid-aortic syndrome usually presents with arterial hypertension in children or youths [1]. A definite diagnosis of coarctation is best made by arteriography but other imaging modalities including digital subtraction angiography, ultrasound, CT or MRI can also be used to demonstrate the pathology [2]. The treatment can be medical, endovascular or surgical. Whenever possible, surgery is the best treatment option [3]. A good control of arterial hypertension with drug therapy is mandatory before surgery [4]. If left untreated, the majority of patients die by 40 years of age, due to complications of severe hypertension and ischemia [5].

Conflict of interest
None declared.

Funding body
None.

Ethical statement
Authors state that the research was conducted according to ethical standards.

Informed consent
Informed consent was obtained from all patients participating in this study.

Acknowledgements
Prof. Pierre Francois Plouin, Hypertension Centre of Excellence, Hopital Européen Georges Pompidou, Paris, France.

Appendix A – Supplementary data
Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.crvasa.2016.04.007.

References

[1] T.W. Rooke, J.W. Joyce, Uncommon arteriopathies, in: R.B. Rutherford (Ed.), Vascular Surgery, Saunders, Philadelphia, 2000, 418–434.
[2] H. Steen, V. Schwenger, Good MRI images: to Gad or not to Gad?, Pediatric Nephrology 22 (2007) 1239–1242.
[3] C. Zabal, F. Attie, M. Rosas, et al., The adult patient with native coarctation of the aorta: balloon angioplasty or primary stenting?, Heart 89 (2003) 77–83.
[4] C.W. Lillehei, R.C. Shamberger, Staged reconstruction for middle aortic syndrome, Journal of Pediatric Surgery 36 (2001) 1252–1254.
[5] J.R. Cohen, E. Birnbaum, Coarctation of the abdominal aorta, Journal of Vascular Surgery 8 (1988) 160–164.