Treatment of renal fibromuscular dysplasia in an adolescent male: A case report

Dennis M. Fry, Ojas A. Pradhan

ABSTRACT

Introduction: Fibromuscular dysplasia (FMD) is a rare vascular disease that causes stenosis in the major arteries such as the renal and carotid arteries. The exact prevalence of FMD in the general population is not known. It is often presented with symptoms of hypertension. Mainly reported in middle-aged females, the diagnosis of FMD is often delayed. There are very few case reports of FMD in males and children in literature.

Case Report: We report the case of a 13-year-old asymptomatic boy found to have FMD in the right renal artery (RRA). An angiography revealed high grade stenosis that was corrected by surgery. A hepatorenal bypass using anastomosis of the gastroduodenal artery (GDA) to RRA was deemed appropriate. An angioplasty at the anastomosis six weeks after this procedure corrected an upward trend in postsurgical blood pressure readings and RRA blood velocities. Two years after the surgery, the patient has well-controlled blood pressure, managed with low dose of ACE-inhibitors.

Conclusion: Hepatorenal bypass of RRA through anastomosis of GDA was a viable surgical option in this case. The monitoring of blood pressure readings allowed for early detection and correction of any post-surgical stenosis.
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Keywords: Adolescent male, Fibromuscular dysplasia (FMD), Hepatorenal bypass, Renal hypertension,
blood test for renal panel and CBC, duplex ultrasound of abdominal organs and vessels, and echocardiography. With the exception of elevated right renal artery (RRA) blood velocities and elevated blood aldosterone level, no other results occurred outside the normal ranges. The patient was administered increasing dosages of various anti-hypertensive medications including beta-blockers, ACE-inhibitors, calcium-channel blockers and central alpha agonists (CAA) over next three weeks. These medications poorly controlled the blood pressure.

High velocities in the RRA suggested stenosis in excess of 60%. An abdominal aortic angiography followed, which indicated high-grade 95% stenosis just beyond the origin of the RRA and significant poststenotic dilation with two small aneurysms in the dilated segment. The high stenosis and beaded aneurysms led to the diagnosis of FMD (Figure 1). A large peri-ureteric collateral flow extending into right renal hilum was detected. The left renal artery showed no signs of stenosis. The superior mesenteric, celiac, and gastroduodenal arteries (GDA) were widely patent. Due to more than 95% stenosis of RRA, the case was deemed unsuitable for balloon angioplasty and was referred for surgery.

Within two weeks, a hepatorenal bypass of the RRA was completed using anastomosis of the GDA to the RRA. Post-surgical Doppler readings showed adequate blood flow in the hepatorenal artery bypass (Figure 2). The patient was discharged after three days, advised to continue beta-blockers to control blood pressure, and record daily blood pressure measurements.

Blood pressure readings initially declined following the bypass surgery, but gradually increased over a period of six weeks (Figure 3). Relatively higher renal velocities from a duplex ultrasound in this period supported the possibility of re-stenosis at the site of anastomosis (Table 1). A second angiography suggested 50% stenosis at the anastomosis and concurrently a PTA was performed using a 4 mm balloon. Within a week, the blood pressure readings decreased noticeably and the patient was put on ACE-inhibitors to control the blood pressure (Figure 3). Subsequent, continued monitoring over the next two years showed a steady decrease in the blood pressure readings and renovascular duplex velocities (Figure 4, Table 1). The patient continues to be monitored through regular check-ups while on a decreasing dose of ACE-inhibitors.

**DISCUSSION**

The prevalence of FMD in the general population is not known [4]. However, Plouin et al. report that the prevalence of symptomatic renal FMD is about 4 in 1000. Renal FMD accounts for nearly 58% of all reported FMD cases [5]. FMD is thought to be more prevalent in females than in males; representing 91% of patients in the US FMD registry. Patients in this registry had a mean age of 51.9 years with averages of 4 to 9 years separating the initial symptoms of FMD and its diagnosis [1]. Literature review also indicates limited studies of FMD in males or in pediatric cases [1, 5]. The most common symptom of FMD is hypertension with a prevalence of 63.8% in the U.S. FMD registry [1]. Secondary symptoms of hypertension include headaches, dizziness, pulsatile tinnitus, neck pain, and chest pain. In this case, the patient was completely asymptomatic for hypertension. Elevated blood pressure was the only indication of any problem. As such, this study is unique for the presentation of FMD in an adolescent male that was diagnosed and treated in relatively short period of time.

Although angioplasty remains the gold standard for correcting FMD, Trinquart et al. reported that salutary
blood pressure responses were more likely among younger patients after surgical revascularization for renal FMD [2, 6]. Contemporary surgical treatments of pediatric renovascular hypertension suggest aortic implantation of normal renal artery beyond the stenosis [7]. This case, however, demonstrates that GDA could be used for revascularization of RRA in an adolescent with

CONCLUSION

This case shows that gastroduodenal arteries (GDA) can be used for revascularization of right renal artery in adolescent patients of fibromuscular dysplasia (FMD). The remodeling of arterial lumen, possibly due to natural growth and increased blood flow demand by the kidney, allowed for the viability of this revascularization. Regular postsurgical blood pressure monitoring aided in early detection of postsurgical stenosis.

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Author Contributions

Dennis M. Fry – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ojas A. Pradhan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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