CASE REPORT

Surgical Repair of an Aneurysm of the Arc of Buhler in a Patient with von Recklinghausen’s Disease

J. L. Myers 1, S. V. Losseff 2 and M. N. Gomes 3

Departments of 1Surgery and 2Radiology, Georgetown University Hospital Washington, DC, USA

Key Words Neurofibromatosis; Visceral aneurysm, Arc of Buhler.

Introduction

In 1904, Buhler first described an embryologic remnant that represented an anastomosis between the coeliac artery and superior mesenteric artery (SMA).1 This remnant, or “Arc of Buhler”, may enlarge to become a major collateral in the presence of coeliac axis occlusion. To our knowledge, we report only the second aneurysm of the Arc of Buhler in the English literature between 1966 and 1997, as well as the only one resected and revascularized. It is also the first reported in association with neurofibromatosis, a disease associated with visceral artery aneurysms. Treatment options for visceral aneurysms, including the aorto-splenic artery graft with autogenous vein that we describe, are discussed.

Case Report

The patient is a 39-year-old female with a 4-month history of urinary frequency. A calcified, saccular mass consistent with a left renal artery aneurysm was noted incidentally on an intravenous pyelogram. She has a 14-year history of neurofibromatosis type 1 (von Recklinghausen’s disease) requiring resection of multiple soft tissue neurofibromas in the past. Her physical exam was unremarkable except for diffuse truncal and extremity “café au lait” spots and soft tissue nodules.

Computed tomography of the abdomen revealed an attenuated coeliac axis with enlarged retroperitoneal collaterals from the SMA and a large, partially calcified, bilobate aneurysm of unclear origin (Fig. 1). Angiography revealed an extremely small calibre coeliac artery with an atretic channel. The artery of Buhler was demonstrated from the proximal portion of a widely patent SMA to the distal coeliac trunk reconstituting an enlarged, tortuous splenic artery and small calibre hepatic and left gastric arteries. Arising near the origin of the artery of Buhler was a three-lobulated aneurysm with one area of calcification (Fig. 2). No other visceral aneurysms were noted and the left renal artery was normal.

At operation the retroperitoneum was entered and the root of the mesentery exposed, a large multilobular aneurysm was immediately encountered (Fig. 3a). The afferent and efferent arteries, which appeared to be in continuity with the superior mesenteric artery

Fig. 1. CT scan of abdomen showing the calcified aneurysm. The vessel of origin is not clear.

*Please address all correspondence and reprint requests to M. N. Gomes, Professor of Surgery, Georgetown University Hospital, 3800 Reservoir Rd 4PHC, Washington, DC 20007
and the coeliac axis respectively, were ligated and the aneurysm resected. A saphenous vein graft was placed in a retro-pancreatic position between the infrarenal aorta and the splenic artery to restore flow to the coeliac axis (Fig. 3b). Normal arterial Doppler signals were noted in the splenic artery distal to the anastomosis and a palpable pulse was noted in the graft. The left renal artery was grossly normal. The patient did well postoperatively and was discharged home.

The aneurysm was submitted to pathology and revealed areas of focal calcification of the intima and media. The coeliac artery was ligated intraoperatively and was not part of the specimen. It therefore could not be evaluated for the mesodermal dysplastic changes typical of neurofibromatosis.

**Discussion**

Neurofibromatosis (NF) is an autosomal dominant trait that has a rare but well documented association with arterial stenotic and aneurysmal disease. The renal arteries are most commonly involved, with rare involvement of intracranial arteries, coronary arteries, aorta and other visceral arteries. Vascular pathology in patients with NF depends on the calibre of the...
vessel. Large vessel alterations are a result of fibrosis secondary to intramural Schwann cell proliferation. Lesions of smaller vessels (the majority of lesions in NF) are mesodermal dysplastic lesions of the media resulting in stenosis, with the potential for post-stenotic dilatation and aneurysmal degeneration. Rupture of visceral aneurysms in NF patients has only been rarely reported, but the potential for rupture of visceral aneurysms in general is high. Stanley reported nearly one in four aneurysms presented as clinical emergencies with a mortality of 8.5%. This was confirmed by Carr’s studies in which 63% were symptomatic at presentation and 24% had evidence of rupture. While angiography remains the “gold standard” for defining visceral artery aneurysms, helical CT scanning with 3-dimensional reconstruction and magnetic resonance angiography should be appropriate in the future for defining these lesions.

Treatment of visceral aneurysms is dependent in part on the location. In general, coeliac artery lesions should be revascularized. Coeliac artery ligation without restoration of vascular continuity is infrequently considered, and only in cases where certainty exists that the hepatic circulation will not be compromised. A recent review of the literature indicates that between 65–83% of these lesions were revascularised. A single saccular aneurysm may occasionally be treated by aneurysmmorrhaphy or aneurysmectomy with reanastomosis of the coeliac trunk to the aorta. None of these options were available in this large, friable aneurysm and the large calibre feeding vessel was not amenable to embolisation. Since occlusion of the proximal coeliac artery made this collateral vessel the primary vascular supply to the coeliac circulation, vascular continuity was restored via a saphenous vein graft from the aorta to the splenic artery. The only other report of an Arc of Buhler aneurysm is a 3 cm saccular aneurysm associated with coeliac artery occlusion of unknown aetiology. Aneurysmectomy without revascularisation was successfully performed after two failed attempts at embolisation.

To our knowledge, this case reports the first documented aneurysm of the Arc of Buhler in a patient with von Recklinghausen’s disease and the first reported resection and revascularisation of such a lesion in any patient. Because the complications of visceral artery aneurysms are potentially catastrophic and include rupture and death, their existence should be considered in patients who present with abdominal pain or masses not otherwise explained. This is particularly true in patients with neurofibromatosis, since they have an increased incidence of aneurysms secondary to the vascular manifestations of the disease. Treatment may consist of trans-arterial embolisation or surgical resection. We recommend revascularising aneurysms of the Arc of Buhler or coeliac artery, although it is not mandatory to do so.

References


Accepted 11 August 1998