Cardiovasc Intervent Radiol (1986) 9:139–141

CardioVascular and Interventional Radiology

Isolated External Iliac Artery Aneurysm Secondary to Cystic Medial Necrosis

Madeline S. Crivello, David H. Porter, Ducksoo Kim, Jonathan F. Critchlow, and Leslie Scoutt
Departments of Radiology and Surgery, Beth Israel Hospital and Harvard Medical School, Boston, Massachusetts 02215, USA

Abstract. The computed tomographic and angiographic findings of an isolated external iliac artery aneurysm secondary to cystic medial necrosis in a patient without Marfan's disease are demonstrated. A review of the differential diagnosis and surgical treatment of iliac artery aneurysms is presented. The dramatic surgical sequelae in this patient underscore the importance of preoperative consideration of this rare diagnosis.

Key words: Artery, external iliac—Aneurysm—Angiography—Computed tomography—Cystic medial necrosis

Aneurysms of the iliac artery are much less common than aneurysms of the abdominal aorta. They can occur in conjunction with other aneurysms, particularly of the aorta, or, less frequently, as isolated lesions. They are most often secondary to atherosclerosis and thus are usually seen in older patients. Other etiologies are mycotic aneurysms, trauma, and pregnancy [1]. To our knowledge, this case of an isolated iliac artery aneurysm secondary to cystic medial necrosis (CMN) is the first report in the literature. In addition, it is unusual because this patient lacked the findings of Marfan's disease. Preoperative consideration of this diagnosis might have changed the surgical approach in this case and might have facilitated a difficult operation.

Case Report

Three weeks prior to this admission, this 27-year-old Vietnamese man noted the sudden onset of right grain swelling and pain without preceding trauma. Past history was notable for a perforated appendix 1 year previously. A contrast-enhanced CT scan of the pelvis at another hospital was remarkable for a high attenuation rim surrounding the right external iliac and common femoral arteries and veins with some lower attenuation material between the opacified vessels and the rim (Fig. 1). Several needle aspirations were performed in the right groin; cultures were negative. The patient was discharged home with a diagnosis of hematoma of unknown etiology.

The symptoms persisted and he was seen at this hospital. Physical exam was remarkable for a slight, emaciated male. Blood pressure was 100/70 mm Hg. The patient weighed 40 kgs. and was 160 cms. tall. Hands and joints were normal. There were no ocular abnormalities. No cardiac murmurs were noted. There was induration of the right groin with absent femoral and distal pulses.

A contrast-enhanced CT scan demonstrated a soft-tissue mass in the expected positions of the external iliac and common femoral vessels (Fig. 2). The vascular structures were no longer clearly seen as they had been on the outside scan from 3 weeks earlier. Arteriography demonstrated a saccular aneurysm of the right external iliac artery at its origin (Fig. 3). A flush abdominal aortogram and a left iliofemoral runoff showed no other vascular abnormalities.

The preoperative diagnosis was a mycotic aneurysm with local hemorrhage. The patient went to the operating room for aneurysmectomy and cross-femoral anastomosis via a retroperitoneal approach. Friability of the arteries and veins was noted. During the dissection the right common iliac vein was injured, resulting in uncontrollable bleeding and hypovolemic shock. The peritoneal cavity was entered to control the inferior vena cava. Gentle traction on the aorta tore it in half. After attempts to graft the aorta were unsuccessful, it was finally ligated. The pelvis was packed to control venous bleeding. When the left axillary artery was carefully mobilized to perform an axillofemoral graft, it tore in half. It was repaired after several attempts with a Gortex graft. The operation took 18 h, and the patient received over 100 units of blood products. Autotransfusion was not performed because of the possibility of a mycotic aneurysm.

The pathologic examination of the excised right external iliac artery segment showed an aneurysm with extensive mucoid degeneration (cystic medial necrosis) of the arterial wall. Cultures of the arterial wall were negative. Cardiac echo demonstrated a normal-appearing aortic root and thoracic aorta. The mitral and aortic valves appeared normal.

Discussion

Isolated iliac artery aneurysms are uncommon, comprising less than 2% of all aneurysms. The most

Address reprint requests to: Madeline S. Crivello, M.D., Department of Radiology, Beth Israel Hospital, 330 Brookline Avenue, Boston, MA 02215, USA

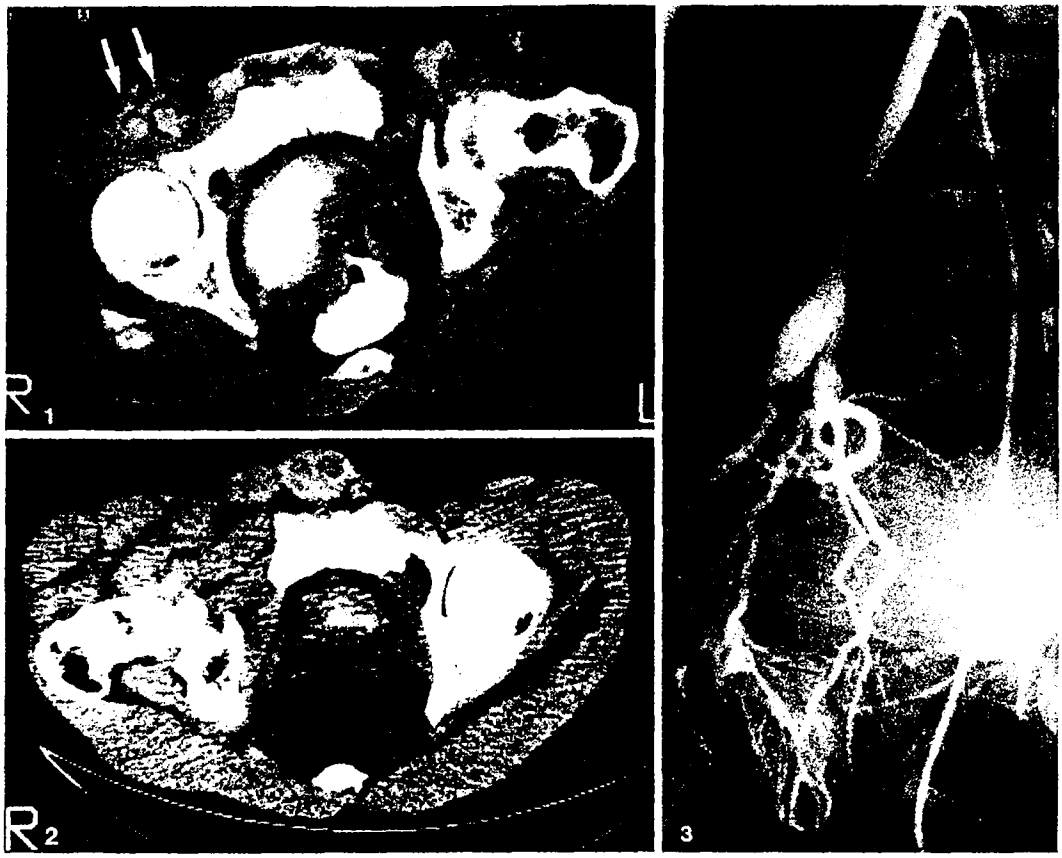

Fig. 1. Enhanced CT scan. The opacified common femoral artery and vein lumens are surrounded by lower-attenuation areas, and an outer, high-attenuation rim (arrows). **Fig. 2.** Enhanced CT scan. The right common femoral artery and vein are no longer seen as separate structures. There is now an ill-defined soft-tissue mass in the right groin. **Fig. 3.** Arteriogram. The saccular aneurysm of the external iliac artery causing high-grade narrowing of the adjacent arterial lumen with smooth encasement of the distal external iliac artery is demonstrated. A small arteriovenous fistula, presumably secondary to the previous needle aspirations, is noted (arrows).

common cause is atherosclerosis. In a Mayo Clinic series, all of the 40 isolated iliac artery aneurysms examined histologically were atherosclerotic in origin [2]. Iliac artery aneurysms have also been reported after pregnancy, trauma, and infections. To our knowledge, there has been no reported case of an isolated iliac artery aneurysm secondary to CMN.

The term cystic medial necrosis (CMN) was introduced by Gsell in 1928. Since that time, use of elastic tissue stain has indicated that the dominant histologic finding in the aorta is massive degeneration of elastic fibers, with replacement by collagen fibers and mucoid material, rather than true cyst formation [3]. Arteries with CMN are prone to aneurysm formation, dissection, and rupture. CMN is characteristically seen in aortas of patients with the Marfan syndrome. It has been reported in the aorta of some patients without Marfan's syndrome features and rarely in the pulmonary arteries of patients with Marfan's syndrome and in some patients with long-standing pulmonary hypertension [4].

CMN is but one feature of Marfan's syndrome which has characteristic familial, ocular, cardiovascular, and skeletal features [5]. It is inherited as an autosomal-dominant with a prevalence of the classic syndrome being 4–6/100,000 people. However, the expressivity of the classic Marfan's gene varies widely. Our patient had none of the classic features.

A variety of cardiovascular lesions have been observed in the heart and vessels of patients with Marfan's syndrome [6]. Most common is fusiform dilatation of the ascending aorta, but involvement of the descending and abdominal aorta has been reported. It is estimated that less than 10% of the expected number of elastic fibers are present in the media of the wall of aortic aneurysms in patients with Marfan's syndrome [3]. Analysis of 151 previously reported necropsy patients with Marfan's syndrome found that 8 (5%) had normal hearts and ascending aortas [6].

Iliac artery aneurysms are often asymptomatic unless rupture occurs [7]. Because they lie adjacent to the pelvic structures, symptomatic patients often present with genitourinary or gastrointestinal symptoms. The natural history of iliac aneurysms appears to be progressive enlargement with a high rate of rupture, often resulting in death due to retro- or intraperitoneal hemorrhage [2]. Lowry and Kraft [8] reported that 75% of their patients presented with ruptured aneurysms. The mortality rate in patients undergoing emergency iliac aneurysmectomy is high, 46–80% [1, 8]. It is recommended that patients with isolated iliac artery aneurysms undergo elective repair which is associated with low morbidity and mortality for atherosclerotic aneurysms [8].

The extreme surgical difficulties encountered in our patient suggest that the rare iliac artery aneurysm due to CMN can present significantly higher operative risks. In retrospect, a clue to this patient's vascular fragility was that during his arteriogram, despite a clean, one-pass arterial puncture, smooth arterial dilatation, and placement of a 5-F polyethylene catheter, immediate bleeding around the catheter necessitated exchange for an arterial side-arm sheath and prolonged compression for hemostasis. When a young patient, without a history of trauma, presents with an isolated iliac artery aneurysm, cystic medial necrosis, although rare, should be considered in the differential diagnosis.

References

1. Markowitz AM, Norman JC (1961) Aneurysms of the iliac artery. Ann Surg 154:777–787
2. McCready RA, Pairolero PC, Gilmore JC, Kazmier FJ, Cherry KJ Jr, Hollier LH (1983) Isolated iliac artery aneurysms. Surgery 93:688–693
3. Waller BF, Reis RL, McIntosh CL, Epstein SE, Roberts WC (1980) Structure-function correlations in cardiovascular and pulmonary diseases (CPC). Chest 77:533–540
4. Natelson EA, Watts HD, Fred HL (1970) Cystic medionecrosis of the pulmonary arteries. Chest 57:333–335
5. Ryeritz RE, McKusick VA (1979) The Marfan syndrome: Diagnosis and management. N Engl J Med 300:772–777
6. Roberts WC, Honig HC (1982) The spectrum of cardiovascular disease in the Marfan syndrome: A clinico-morphologic study of 18 necropsy patients and comparison to 151 previously reported necropsy patients. Am Heart J 104:115–135
7. Silver D, Anderson EE, Porter JM (1967) Isolated hypogastric artery aneurysm. Arch Surg 95:308–312
8. Lowry SF, Kraft RO (1978) Isolated aneurysms of the iliac artery. Arch Surg 113:1289–1293