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ORIGINAL ARTICLE

Segmental arterial mediolysis: Unrecognized cases culled from cases of ruptured aneurysm of abdominal visceral arteries reported in the Japanese literature

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Abstract

Segmental arterial mediolysis (SAM) is a rare nonatherosclerotic and noninflammatory arteriopathy that was proposed by Slavin et al. [Segmental mediolytic arteritis. A clinical pathologic study, Lab. Invest. 35 (1976) 23–29]. It mainly involves abdominal visceral arteries and is characterized by lytic degeneration of the media, resulting in intraabdominal bleeding. We collected 27 unrecognized cases of SAM by reviewing microscopic slides of cases of ruptured aneurysms of visceral arteries, except splenic and hepatic aneurysms, reported in the Japanese literature. This paper describes the pathological and clinical features of these cases. The symptom at onset was abdominal pain associated with intraabdominal bleeding in all cases. The most involved artery was the middle colic artery, accounting for 14 (50%), followed by gastric and gastroepiploic arteries, (6 and 5, respectively). Seventy-eight percent of aneurysms were of dissecting type and the rest of pseudoaneurysm type, except for one. Multiple aneurysms were found in 9 cases (33.3%). Pathological lesions were acute in all. The outcome of those who had surgery was good, even in those who had surgery for 1 ruptured aneurysm, leaving the others unmanaged. The relationship of SAM to fibromuscular dysplasia is discussed. Secondary changes in the wall of the accompanying vein to the affected artery are briefly described. It is emphasized that the majority of aneurysms of abdominal visceral arteries are gathered together as SAM as a definite clinical and pathological entity.

Keywords: Segmental arterial mediolysis; Abdominal visceral artery; Ruptured aneurysm; Dissecting aneurysm; Multiple aneurysm

Introduction

We were interested in this arteriopathy, which has several characteristics, and attempted to cull unrecognized cases of segmental arterial mediolysis (SAM) from cases of ruptured aneurysm of abdominal arteries except splenic and hepatic aneurysms reported in the Japanese literature on the assumption that most aneurysms, especially dissecting aneurysms, might be caused by SAM. The purpose of this paper is to describe the pathological and clinical features of these cases and to emphasize that SAM should be considered as a differential diagnosis in cases of intraabdominal bleeding.

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Materials and methods

Cases in this study were derived from a review of 27 previously reported cases of ruptured aneurysms of abdominal visceral arteries of unknown cause or other etiologies. We asked the authors for permission to review microscopic slides of their cases, and unstained sections were also available for some cases. Microscopic slides were reviewed in 26 cases, except one, which were considered to be putative SAM based on the description of histopathologic findings and attached microphotographs [14].

The diagnosis of SAM was made according to the criteria described by Slavin et al. [16–18]. They emphasized 4 distinct diagnostic lesions: first, mediolysis, which begins in outer media and results in vacuolization; second, separation of the media from the adventitia, resulting in dissecting hematoma; third, arterial gaps created by transmedial mediolysis with concomitant loss of intima and elastica, which frequently enlarge and cause rupture resulting in pseudoaneurysm; and fourth, reparative fibrosis replacing granulation tissue.

Table 1 shows the 27 cases. The first series of 13 cases (Cases 1–13) was reported in 1998 [3], the next 5 (Cases 14–18) in 2003 [2], followed by 4 (Cases 19–22) in 2005 [4]. The remaining 5 were collected thereafter [1,9,10,14].

Results

Table 2 summarizes the pathological and clinical data of the 27 patients (17 men, 10 women), who ranged in age from 44 to 88 years (av. 60.5 years). Abdominal pain associated with intraabdominal bleeding was the initial sign and symptom in all patients. The most frequently involved artery was the middle colic artery, followed by the gastric and gastroepiploic arteries. Multiple aneurysms were found in 9 cases (33.3%).

The stage of pathological lesions in all 27 cases was acute. Reparative lesions were found in 2 cases: in a nonruptured aneurysm in Case 12 and in a ruptured aneurysm in Case 14. All 3 diagnostic criteria proposed by Slavin were confirmed in all cases. Inflammatory cell infiltration around the adventitia was seen in 2 cases. The initial diagnosis of the 27 cases varied as follows: unknown (14), atherosclerosis (4), congenital (3), medial necrosis (2), fibromuscular dysplasia (FMD), inflammatory, infectious, and systemic lupus erythematosus (1 each). It was noticeable that the 13 cases who presented with dissecting aneurysm of unknown genesis were all SAM. The types of the 28 ruptured aneurysms were as follows: dissecting 22 (78.6%), pseudoaneurysm 5 (17.8%), and unknown 1 (3.6%).

Several changes such as edema, separation, and patchy loss of muscle layers were found in the wall of the accompanying vein to the involved artery. A severe change, patchy vacuolization of the cytoplasm of smooth muscle cells associated with mediolysis similar to arterial lesions, was found only in Case 3 [4,19]. These changes were found in 7 among 13 cases where original microscopic slides were available for review. Four representative cases are described.

Case 1: A 65-year-old woman abruptly developed abdominal pain followed by shock that necessitated a laparotomy. This revealed a hematoma behind the lesser omentum and pulsating bleeding from a branch of the left gastric artery that required partial gastrectomy. Histological examination of the involved arteries revealed changes of early SAM and dissecting aneurysm. Several small arteries also showed changes similar to FMD (Figs. 1–4). In addition, many arteries had an abnormal arrangement of bundles of smooth muscle cells.

Case 12: A 52-year-old man abruptly developed abdominal pain in the morning and was hospitalized. Blood pressure dropped to 68 mm Hg at night. Emergency laparotomy revealed massive intraabdominal bleeding and ruptured aneurysm of the middle colic artery. Partial resection of the transverse colon was performed. Pathological sections of aneurysm, fusiform in shape and 2.5 cm in diameter, showed a dissecting aneurysm. Postoperatively, abdominal angiography disclosed an aneurysm of the common hepatic artery, fusiform in shape, 2.1 cm in length and 1.2 cm in diameter, and it was resected on the 46th day after emergency laparotomy. Pathological sections disclosed a dissecting aneurysm in the reparative stage (Fig. 5). This was the only case in which 2 aneurysms were resected sequentially with an interval.

Case 14: A 54-year-old man underwent emergency laparotomy with ligation of a ruptured aneurysm of the accessory middle colic artery that caused intraabdominal bleeding and shock. This aneurysm had been discovered on a preoperative abdominal angiography. Four weeks later, an aneurysm of the left branch of the middle colic artery was identified – it had been missed on the previous angiogram – and was excised on the 49th postoperative day. Histological examination revealed a pseudoaneurysm due to SAM and 2 arteries showing FMD-like lesions (Figs. 6–9).

Case 20: An 88-year-old man presented with complaints of abdominal pain and distension. Abdominal paracentesis revealed blood, and angiography disclosed continuous bleeding from the greater curvature of the stomach. On emergency laparotomy, a hematoma in the lesser omentum was removed. Pathological sections of the artery revealed a dissecting aneurysm and injury to the wall of the accompanying vein (Figs. 10 and 11).

Discussion

SAM is considered a rare disease, and there have been few reports since Slavin et al. published 3 papers in the West. The first case of SAM in Japan was reported by