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Spontaneous Hemorrhages from Two Arteries Preceding Rupture of a Hepatic Artery Aneurysm: A Possible Case of Multiple Congenital Visceral Aneurysms

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Summary. **A 20-year-old man with two episodes of spontaneous hemorrhage occurring about 1 month before a hepatic artery aneurysm rupture is presented. The first hemorrhage appeared as intraperitoneal bleeding from a subserosal hematoma of the ascending colon, but soon after surgery the patient required transcatheter embolization for bleeding from a branch of the left iliac artery. The hepatic artery aneurysm was identified at emergency laparotomy a few weeks later. In retrospect, the earlier hemorrhages probably similarly represented small congenital visceral aneurysms.**

Multiple visceral aneurysms of a congenital nature involving different arteries are extremely rare. We here report a case with a unique combination of arterial sites.

Key words—congenital multiple visceral aneurysms, rupture, transcatheter arterial embolization.

CASE REPORT

A 20-year-old man visited the Emergency Unit of Shirone Kensei Hospital on January 23, 1996 with abdominal pain which had worsened 48 to 50 hrs after onset; trauma was denied. His past history included two attacks of spontaneous pneumothorax at ages of 16 and 17 years, and he was negative for tuberculosis, diabetes mellitus and syphilis. In recent years, he had been in good health and experienced no symptoms suggestive of collagen-vascular disease such as pyrexia or joint pain. His family history was negative for syndromes such as hereditary hemorrhagic telangiectasia (Osler's disease), Marfan syndrome, and Ehlers-Danlos syndrome.

The abdominal pain gradually worsened after admission, and was followed by progressive anemia. Abdominal ultrasonography and computed tomography (CT) demonstrated intraperitoneal fluid accumulation and localized thickening of the wall of the ascending colon. Abdominal CT did not disclose a visceral aneurysm. At emergency surgery, 450 mL of intraperitoneal blood from a fist-sized subserosal hematoma of the ascending colon was aspirated. Blood was oozing from the bulging serosal surface, and right hemicolectomy including the lesion was performed. The resected specimen showed the subserosal hematoma measuring 9×7 cm at the middle of the ascending colon (Fig. 1). Most of the lesion was located on the antimesenteric side, and only erosion was evident on the mucosal aspect.

Although the immediate postoperative course was uneventful, blood emerged from an intraperitoneal drain later on the first postoperative day. Abdominal exploration was repeated on January 25, revealing a huge hematoma occupying the left anterior part of the true pelvic cavity and extending toward the left groin. Blood was oozing from the posterior peritoneal defect resulting from the previous ascending colon mobilization. The Abdominal wall was closed and the patient was transfused with approximately 3000 mL of blood prior to transfer to the Emergency and Critical Care Unit, Niigata University Medical Hospital.

On transfer, physical examination revealed a thin

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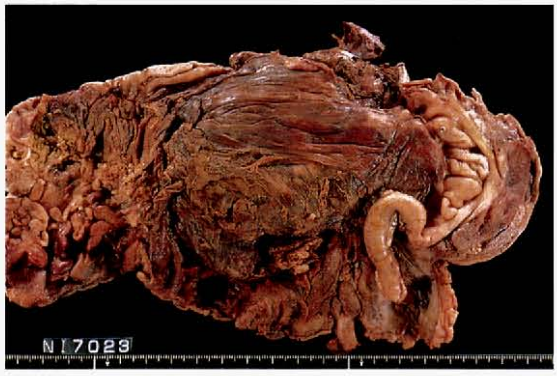

Fig. 1. Resected specimen showed subserosal hematoma measuring 9×7cm at the middle of the ascending colon. The lesion was mainly located at the antimesenteric side. Only erosion was seen on the mucosal aspect. Ruptured wall of the aneurysm can not be identified.

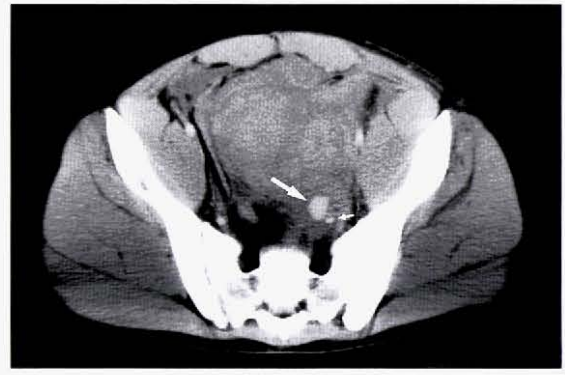

Fig. 2. Pelvic computed tomography reveals a huge mass shadow with several densities occupying the true pelvic cavity. A round high density area *(big arrow)* by the extravasated contrast material is visualized near the left internal iliac artery *(small arrow)*.

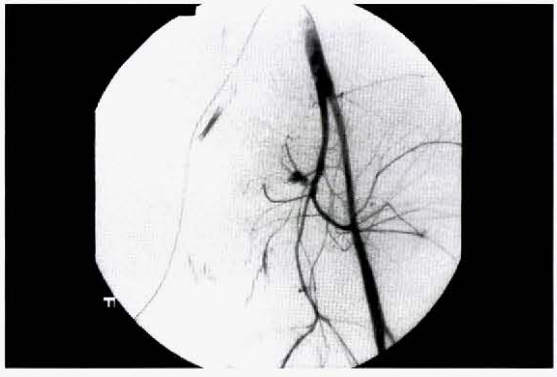

Fig. 3. Selective digital subtraction angiography of the left common iliac artery demonstrates the extravasation of the contrast material from a small branch of the left internal iliac artery which is located proximal to the left superior and inferior gluteal arteries. Of the 2 round shadows, the smaller one may delineate the shape of the ruptured aneurysm, while the larger one may be formed by the extravasated contrast material. No findings suggestive of atherosclerosis, vasculitis, or fibromuscular dysplasia are demonstrated.

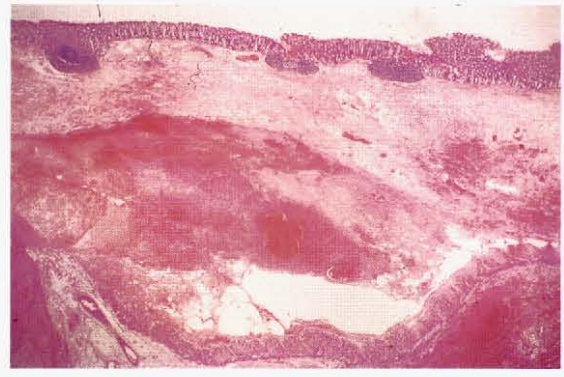

Fig. 4. Histopathologically, the specimen shows a massive hemorrhage from the subserosa to submucosa. Reactive cellular inflammation with vascular congestion existed in the mucosa. (Hematoxylin and eosin stain. ×5)

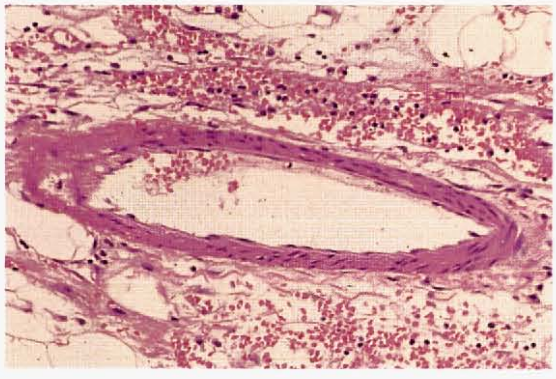

Fig. 5. No evidence of specific vasculitis or atherosclerotic changes can be found in the arterial wall. (Hematoxylin and eosin stain. ×23)

young male (weight, 55 kg; height, 172 cm). The skin was slightly pale, moist and cool; findings suggestive of telangiectasia were not observed. Abdominal examination showed a massive, diffuse, and tender swelling including subcutaneous hematoma extending from the left lower quadrant to the left inguinal region and the frontal aspect of the left thigh. A hematoma was confirmed by CT (Fig. 2), and aortic angiography demonstrated the branch of the left iliac artery located proximally to the left superior and inferior gluteal arteries as the bleeding artery (Fig. 3). Direct embolization of the bleeding artery was attempted but insufficient to stop the hemorrhage, and required embolization of the left internal iliac artery. The clinical course after the intervention was completely uneventful, being free of complications such as acute renal failure or infection of the hematoma. He was transferred back to the previous hospital on February 6 and admitted for another 16 days before discharge.

However, the patient was admitted on emergency with hemorrhagic shock and operated upon on March 4 at Juntendo University Hospital during a trip to Tokyo. Pulsatile bleeding was found from a ruptured oval aneurysm of the proper hepatic artery, about 3 cm in the average diameter at the hepatoduodenal ligament. Both the gastroduodenal and proper hepatic arteries were ligated just proximal to the aneurysm, and the aneurysm wall was sutured. Cholecystectomy also was performed. The postoperative course was uneventful and the patient was discharged on the 49th postoperative day. Postoperative angiography disclosed no additional visceral aneurysms in the abdomen. Aneurysms in the upper half of the body, including cerebral aneurysm, were not investigated. The patient has been well for 7 months after discharge.

DISCUSSION

This is a report of young man with repeated spontaneous major hemorrhages beginning about 1 month before rupture of a hepatic artery aneurysm. The first issue for discussion is the cause of the repeated bleeding episodes. Initially, intraabdominal bleeding occurred from a subserosal hematoma of the ascending colon. The lesion was mainly located at antimesenteric side, suggesting a peripheral site of the vasa recta as the bleeding point. Therefore, presumed aneurysm was judged as being very small. Subserosal hematoma formation is a common sequel following rupture of a small aneurysm of a branch of a jejunal, ileal, or colic artery.[1-5]

Although macroscopic and histopathologic examination (Figs. 4 and 5) of the resected specimen could not identify a ruptured aneurysm wall, most mesenteric branch aneurysms are known to be small, often measuring a few millimeters in diameter,[4,6,7] and it is not uncommon that aneurysms on a branch of the mesenteric arteries are not recognized at surgery or autopsy.[3,5] Therefore, the aneurysm may have been obliterated when it ruptured.

In the second bleeding episode, the bleeding point was identified by selective angiography as a small branch of the left iliac artery proximal to the superior and inferior gluteal arteries. Quite possibly a small aneurysm ruptured at the bleeding point. Findings from angiography were mildly suggestive of a small ruptured aneurysm byt did not directly show it (Fig. 3). Again this difficulty could be related to the small aneurysm size; Reuter et al. have reported small aneurysms 3-4 mm in diameter,[7] and such a small ruptured aneurysm may be inherently difficult to visualize in angiograms.

The final hemorrhage due to a ruptured hepatic artery aneurysm strongly suggests that the preceding hemorrhages were caused by ruptures of aneurysms. Presumably, the asymptomatic hepatic artery aneurysm had already existed before the beginning of the episode, although the first abdominal CT could not demonstrate it. It is unlikely that the aneurysm appeared and grew so rapidly after the first hemorrhagic episode.

The second issue is the etiology of the patient's visceral aneurysms, most likely multiple ones. Major etiologies of "true" visceral aneurysms, which exclude pseudoaneurysms arising from trauma, pancreatitis, or intraperitoneal infections, include atherosclerosis, fibromuscular dysplasia, collagen-vascular diseases, gestational alterations, mycotic embolization, and "congenital" aneurysms.[2,4,8] Rarer etiologies are hereditary conditions like Osler's disease,[9-11] Marfan syndrome,[12] and Ehlers-Danlos syndrome.[13]

The most common locations of visceral aneurysms, in order of frequency, are the splenic, renal, and hepatic arteries, followed by the superior mesenteric and celiac arteries.[2] Aneurysms of the gastroduodenal, pancreaticoduodenal, gastric, gastroepiploic, jejunal, ileal, and colic arteries have been reported in limited numbers.[2,8]

Of considerable interest is the fact that visceral aneurysms of different arteries have specific etiologic associations. In females, splenic artery aneurysms often are gestational,[14] while fibromuscular dysplasia is the main etiology of renal aneurysms. Hepatic artery aneurysms are usually atherosclerotic or traumatic.[15]

Table 1. Laboratory data

(January 24)					
WBC	$9000/mm^3$	Bleeding Time: 4 min. 0 sec.			
RBC	$386\times10^4/mm^3$	PT: 11.7 sec (Cont: 11.8 sec)			
Ht	32.7%	APTT: 28.9 sec (Cont: 29.4 sec)			
Hb	11.0g/dl				
Plt	$19.7\times10^4/mm^3$				
(January 29)					
WBC	$6600/mm^3$	T. Bil.	2.3mg/dl	CRP	10.2 mg/dl
RBC	$378\times10^4/mm^3$	GOT	33 IU/l	RF	< 5.0 IU/ml
Ht	33.4%	GPT	47 IU/l	IgG	826.1 mg/dl
Hb	11.7g/dl	LDH	496 IU/l	IgA	191.5 mg/dl
Plt	$21.7\times10^4/mm^3$	ALP	294 IU/l	IgM	78.5 mg/dl
T.P.	5.3 g/dl	ChE	117 IU/l	C3	90.0 mg/dl
Alb	3.0 g/dl	CK	135 IU/l	C4	40.0 mg/dl
STS	(−)	BUN	10 mg/dl	CH50	57 HU/ml
TPHA	(−)	Creat	0.6 mg/dl	anti-ENAab:	(−)

PT; prothrombin time, APTT; activated partial thromboplastin time, STS; serological tests for syphilis, TPHA; treponema pallidum hemagglutinin test, CRP; C reactive protein, RF; rheumatoid factor, CH50; hemolytic complement activity, anti-ENAab; anti-extractable nuclear antigen antibodies(anti-Sm, anti-RNP, anti-SS-A, anti-SS-B, antiScl-70, and anti-DNA antibodies).

Table 2. Cases of multiple "congenital" visceral aneurysms

Author and year	Age (yr.) and sex	Cause of aneurysm stated	Artery involved
Levy DF[24] 1941	31M	unknown	gastric a. (1) splenic a. (4 or 5) r. renal a. (2) l. renal a. (m†) sup. mesenteric a. (m)
Hoehen JG et al.[22] 1968	60F	idiopathic	jejunal branch of sup. mesenteric a. (1) splenic a. (1)
Pais SO et al.[23] 1968	49F	congenital	r. renal a. (3) l. renal a. (2) r. accessory hepatic a. (2) splenic a. (1) superior pancreatico-duodenal a. (1)
Noyes PF et al.[25] 1979	49F	unknown	r. hepatic a. (m) l. hepatic a. (l) splenic a. (1) gastroduodenal a. (1) sup. mesenteric a. (m)
Present case	20M	congenital	proper hepatic a. (1) branch of a. supplying the ascending colon (1) branch of l. internal iliac a. (1)

( ): number of aneurysms, m†: multiple, no statement of number.

The present patient's presumed multiple visceral aneurysms most likely are "congenital". No signs or symptoms suggestive of specific hereditary syndromes were observed. No complaints of joint pain or fever, suggestive of collagen-vascular disease such as polyarteritis nodosa and rheumatoid arteritis, were present.[16-18] Relevant serologic tests also were negative for collagen-vascular disease on January 29. Elevated CRP level was considered to be the result of its accerelated hepatic synthesis induced by the acute insult, and this decreased to 1.3 mg/dl on February 21. Incidentally, laboratory data on admission at the Shirone Kensei Hospital were negative for hemorrhagic disorders (Table 1). In addition, histopathologic examination of the resected colon disclosed no changes of systemic vasculitis such as transmural degeneration, inflammatory cell infiltration in the wall, thickening of the wall, and loss of the internal or external elastic lamina (Fig. 5). Incidentally, histopathological evaluation of the hepatic artery aneurysm was impossible because no specimen was obtained.

The patient had been normotensive, and angiography after the final surgical intervention revealed normal renal, celiac, superior and inferior mesenteric arteries. Earlier iliac angiography also demonstrated no abnormal findings suggestive of fibromuscular dysplasia,[19,20] vasculitis, or atherosclerosis (Fig. 3). Additionally, 94% of cases of fibromuscular dysplasia are reported to occur in female patients.[19] Hereditary conditions and collagen diseases, therefore, represent unlikely etiologies of aneurysms in this case, as do atherosclerosis and fibromuscular dysplasia.

Shallow et al.[21] describe congenital aneurysms as showing "frequent multiplicity, small size, location at the bifurcation angle of a medium-sized artery, and association with a defect in the muscularis". In most other reports,[1,6,22,23] however, aneurysms are labeled as "congenital" because of the patient's age or the absence of the other know etiologies.[4] Certainly from the latter viewpoint, the present patient's aneurysms would be considered "congenital" with his age of 20 being supportive of the supposition.

A third aspect of the case warranting discussion is the very uncommon multiplicity of the visceral aneurysms, with single visceral aneurysms being by far the rule. Multiple "congenital" visceral aneurysms are exceedingly rare, compared with the less uncommon multiple visceral aneurysms of collagen-vascular disease, particularly polyarteritis nodosa.[17,18] Previous reports of multiple "congenital" visceral aneurysms are summarized in Table 2.[22-25] Considering cases reported in English over the last few decades, the present case is unique of the combination of arteries involved.

In summary, successive hemorrhagic episodes in the present patient are best explained by the ruptures of the three aneurysms of different small-to-medium-size arteries: a branch of colic artery supplying the ascending colon, a branch of the left iliac artery, and the proper hepatic artery. Multiple aneurysms in this patient represent a somewhat ill-defined "congenital" category, though long-term follow-up may reveal a specific underlying condition.

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