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Segmental arterial mediolysis with 5 splenic artery aneurysms. A rare finding of a rare disease: Case report and literature review



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ABSTRACT

INTRODUCTION: Splenic artery aneurysms (SAA) are uncommon findings. They are usually single and isolated; however they can be multiple; hence vasculopathy and segmental artery mediolysis may be considered.

PRESENTATION OF CASE: In our manuscript we present a case of a 54 year old multiparous lady who was discovered incidentally to have a diseased splenic artery containing five SSAs. The largest aneurysm was close to the takeoff of the vessel and the smallest was distal embedded in the splenic hilum. Endovascular option was technically not feasible. Therefore the patient underwent a complete splenic artery resection with splenectomy and the histopathologic examination was suggestive of segmental arterial mediolysis (SAM).

DISCUSSION AND CONCLUSION: Multiple SAAs remains a rare finding of a rare disease. Complications can be crucial and high index of suspicion is important. Segmental arterial mediolysis can be considered in patients with several aneurysms on one anatomic site; Angiography is the gold standard diagnostic and therapeutic method. Complete splenic artery resection with splenectomy is the best treatment option for solitary vessel involvement.

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1. Introduction

Splenic artery aneurysms (SSA) are the most common splanchnic aneurysms. They account for 60% of all visceral aneurysms [1] and appear to be the third most common intra-abdominal aneurysms following the aorta and iliac arteries [2], in spite of this, they are considered uncommon findings especially when they present as many aneurysms in one or different vessels. Etiologic factors can be related to fibromuscular dysplasia, portal hypertension, pregnancy and atherosclerosis; however segmental artery mediolysis is rarely seen [3]. We are reporting a case of five SAAs with underlying segmental artery mediolysis which was referred to our hepatobiliary unit at Amiri university hospital in Kuwait city for further evaluation and management.

2. Case presentation

A 54 year old multiparous Kuwaiti lady G8P8, who is known to have diabetes, hypothyroidism and nephrolithiasis was discovered

incidentally to have 2.5 × 2.5 cm splenic artery aneurysm during a CT scan that was done for left sided abdominal pain mainly to evaluate her renal stone disease.

The patient has no past surgical history relevant to this finding or any family history of vasculopathy. Physical examination was normal. Initially we started with angiography as a diagnostic and possible therapeutic method that showed a diseased splenic artery with multiple SSAs, the biggest was measuring 2.5 cm in diameter, sacular in shape and located in the proximal segment of the splenic artery close to the celiac axis and the smallest was distal in the hilum nearly embedded in the parenchymal tissue of the spleen (Fig. 1). No other vascular pathology was found in different vessels in the abdomen.

The case was discussed in the multidisciplinary meeting with our interventional radiologists and due to technical reasons such as extensive vessel involvement and presence of multiple proximal and distal aneurysms, we elected to exclude the endovascular option. The patient was informed about the plan and agreed to proceed with the surgery. She was given post-splenectomy vaccinations two weeks prior to surgery and VTE prophylaxis preoperatively. She underwent open complete splenic artery resection along with splenectomy (Fig. 2a and b), no complications noted during her surgery. Microscopic examination revealed non-

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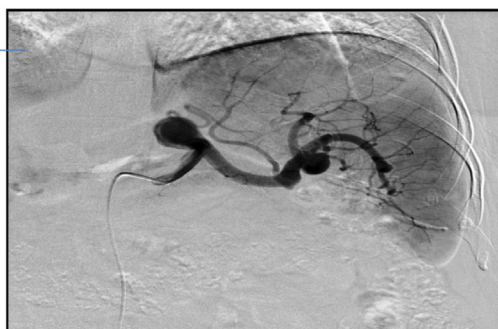


Fig. 1. Angiography showing string of beads representing multiple SAAs.

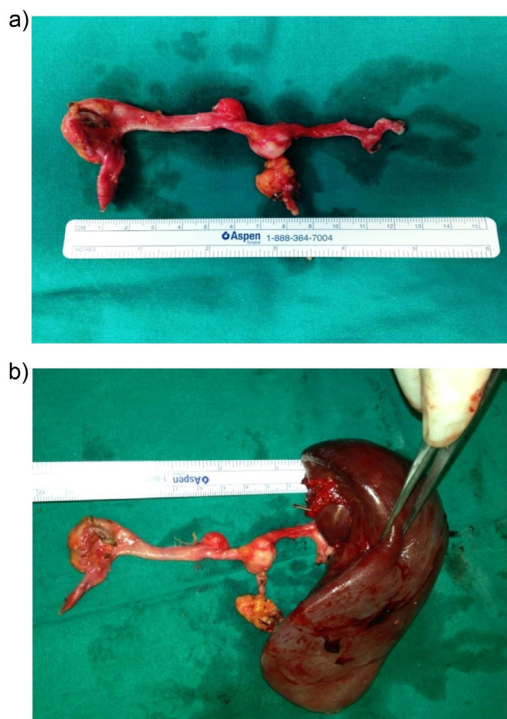


Fig. 2. a. Complete splenic artery resection with splenectomy. b. Splenic artery aneurysms with variable sizes.

arteriosclerotic, non-inflammatory vasculopathy with areas of smooth muscle fibers loss (mediolysis) and rupture of elastic lamina; those histopathological changes were suggestive of segmental arterial mediolysis (SAM) (Fig. 3). The patient had an uneventful postoperative recovery. Follow up CT angiography of the abdomen and pelvis did not show any vascular abnormalities.

3. Discussion and conclusion

Splenic artery aneurysms (SAA) have a female predominance on contrary to all arterial aneurysms. They are usually seen in the fifth and sixth decades with a mean age of 52 years [4]. Generally, SAAs manifest in one of three ways:

- I Incidental finding on imaging, during laparotomy or on post-mortem examination [5]
- II Vague abdominal pain in 20%, mainly epigastric and left upper quadrant [2,6]
- III Bleeding secondary to rupture in 3–9.6% which is usually associated with high mortality of 36% that is increased with pregnancy up to 70% maternal death [7].

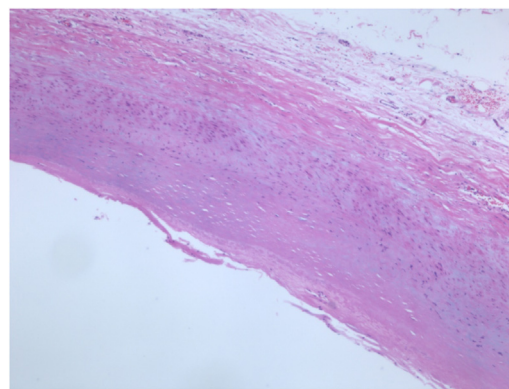


Fig. 3. Histopathological features representing segmental arterial mediolysis.

Most aneurysms are single, small saccular and located in middle or distal portion of the splenic artery [4,8]. The etiology of SAA is unknown; however local failure of the arterial wall connective tissue to maintain the vessel integrity leads to fragmentation of the elastic fibers and loss of smooth muscle [7]. It is also believed that there is a strong association between SAA and pregnancy [7,9].

Treatment is recommended for all patients with symptomatic or inflammatory aneurysms, pregnant patients or if they have the intention for conception, aneurysms measuring more than 2 cm and expanding in diameter and for those undergoing liver transplant [6,8,10]. CT arteriography is the gold standard for diagnosis in suspected unruptured aneurysms [11,12]; however angiography remains the most valuable modality to precisely locate the bleeding site and to provide an initial management option. The interventional endovascular procedures are considered the first choice of treatment when it is available and technically feasible. When the aneurysm is located in the distal portion, patient may ultimately require splenectomy [13].

Our case was challenging as the patient was asymptomatic and we attempted initially to go by endovascular option. We started with angiography that revealed an extensive involvement of the whole integrity of the splenic artery represented by tortuosity of the vessel and 5 aneurysms (2 of them were larger than 2 cm). We also noticed the presence of a big aneurysm close to the celiac axis and one at the hilum nearly embedded in the splenic parenchyma (Fig. 1). Due to the above findings we decided to exclude the endovascular method and proceed with a complete resection of the splenic artery along with splenectomy (Fig. 2a and b). We chose to proceed with an open technique due to the lack of laparoscopic experience in dealing with this rare finding.

Microscopic examination of the resected splenic artery showed multiple aneurysms with thickened intima, focal atheromatous plaques and duplication of the internal elastic lamina. The media (even away from the aneurysms) shows degenerative changes in the muscular fibers, (constriction pyknosis and cytoplasmic eosinophilia as well as myxoid changes in the background) with areas of muscle fibers loss (mediolysis) (Fig. 4). Rupture of the elastic lamina was shown with Orcein stain Shikata (Fig. 5) and elastic Van Giessen stain (Fig. 6). These histopathological features were adequate enough to establish this rare and difficult diagnosis despite the similarity with other vasculopathy.

Segmental arterial mediolysis (SAM), also known as segmental medial arteriolysis (SMA), is a rare vasculopathy characterized by non-inflammatory degeneration of the medial layer of muscular arteries [14]. It is an iatrogenic arteriopathy caused by alpha-1 adrenergic agonists or Beta-2 agonists able to release norepinephrine from the peripheral nervous system. Causative agents include adrenergic agonists used to control blood pressure, B-

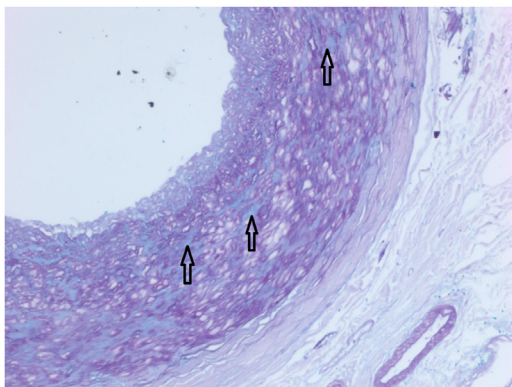


Fig. 4. Alcian blue positive staining, areas of smooth muscle fiber loss filled with myxoid material (Arrows).

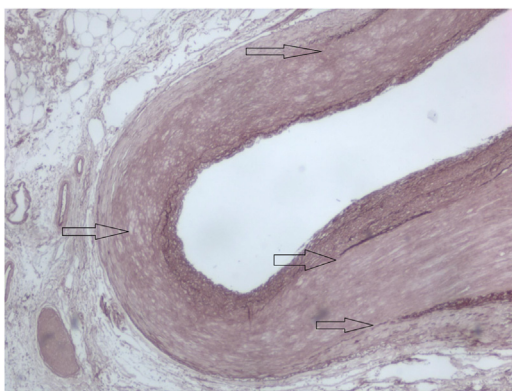


Fig. 5. Shikata stain, rupture of elastic laminae in the affected area (arrows).

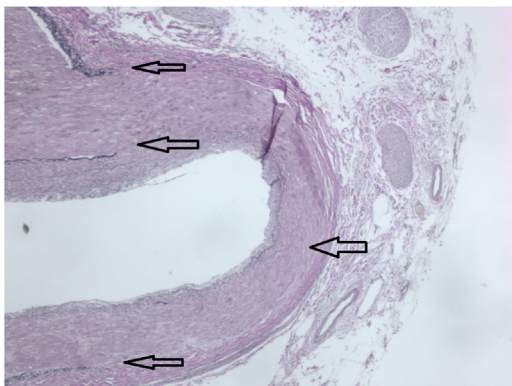


Fig. 6. Van Giessen stain showing interruption of elastic lamina (arrows).

2 tocolytic agonists, and ractopamine used as a repartitioning agent in animal husbandry. The liberated norepinephrine both injures and stimulates a robust reparative response in the muscular arteries in the abdomen, brain base, and coronary arteries. This response may be augmented by endothelin-1 formed in the arterial adventitia. Originally described by Slavin and Gonzalez-Vitale as “segmental mediolytic arteritis” in 1976, Slavin and colleagues later proposed a change in the name to “segmental arterial mediolysis” due to lack of consistent evidence of true inflammation in both the clinical presentation and the histologic features of the disease [15].

Pathologically SAM involves two phases, injurious and reparative.

The injurious stage contains three types of arterial lesions: 1) apoptotic induced mediolysis, 2) separation of the outer media from the adventitia and 3) the formation of arterial gaps that develop into vascular aneurysms which are the most frequent angiographic feature of SAM [15,16].

The reparative can develop sequelae (thrombosis, stenosis, ischemia or regaining normal appearance) that resemble fibromuscular dysplasia. Slavin and coworkers have proposed that SAM is the precursor of FMD a hypothesis derived from the morphologic appearance of SAM in its reparative phase. However, this hypothesis has been contested for the following reasons.

1) SAM in adults is a disease principally affecting the elderly while FMD occurs in younger adults. 2) FMD being much more frequent in females while SAM occurs with equal frequency in both sexes. 3) The clinical presentations differ, SAM presenting with profuse bleeding from intestinal arteries while FMD is announced by ischemic changes causing hypertension or strokes. 4) FMD is reported in large arteries supplying the extremities a finding not definitively found in SAM [17,18].

The differential diagnosis of SAM is wide and complicated. However, some clinical details may be highly significant [Table 1] [19]. The principle difference may appear only in the histopathology, like in periarthritis nodosum (PAN) since this entity may resemble SAM on angiograms. In contrast to SAM, the lesions in this arteritis are systemic, preferably occur at branching sites, the medial muscle loss is caused by fibrinoid necrosis that generally begins in the arterial inner coat, its inflammation is significant, and arterial injury frequently results in intimal scarring with luminal narrowing, dissections rarely occurs and laboratory findings support an immunologic etiology. The other differential diagnosis is muscular artery cystic necrosis, a newly named non-inflammatory muscular artery arteriopathy which may be confused with SAM both clinically and pathologically. This arteriopathy represents the muscular artery equivalent of cystic media necrosis of the elastic arteries since it exhibits similar morphologic features and can occur concomitantly with this entity [17,18].

The definitive diagnosis of SAM requires certain histological confirmation. In our case, the presence of degenerative changes of the muscle fibers (constriction, pyknosis and cytoplasmic eosinophilia as well as myxoid changes in the background) along with areas of smooth muscle loss should raise the diagnosis of SAM, especially in the presence of multiple aneurysmal formations.

SAM is typically limited to vessels in only one anatomic site [16]. This rare vasculopathy mainly involves the splanchnic, coronary and cerebral territories. The most common sites are celiac axis, splenic, superior mesenteric, renal, and inferior mesenteric arteries. The involvement of cerebral vasculature is less frequent and occurs in adults, while the coronary artery involvement usually develops in younger population, children and infants [20].

Usually, the immediate small branches of the affected arteries also show features of SAM [16]. Although damaged arteries may be completely remodeled by the reparative granulation tissue and appear normal in the subsequent angiographic studies. Arterial dissection and hemorrhage tend to be more common in SAM than in other vascular diseases. That is why periodic surveillance is prudent. Occasionally, abdominal SAM may not present with massive bleeding but with complicating arterial thrombosis.

Treatment usually involves embolization, stenting, surgical bypass or resection of the injured arteries [21]. It is a particular challenge to distinguish SAM from systemic inflammatory vasculitides since corticosteroids and immunosuppressive agents, which are crucial in the management of the inflammatory vasculitides, have no proven benefit in SAM [22].

Table 1
Clinical and laboratory features distinguishing SAM from its mimics.^a

	SAM	FMD	PAN	AAV	GCA	TA	BD	KD	Mycotic aneurysm	Type IV Ehlers-Danlos syndrome	Marfan's syndrome	NF	PXE	Atherosclerosis
Demographics														
Sex predisposition	Equal	Female (3:1)	Equal	Equal	Female (2:1)	Female (8:1)	Equal	Male (1.5:1)	Equal	Equal	Equal	Equal	Female (2:1)	Male
Age at onset of symptoms, years	Any; 40s–60s most common	20s–30s	40s–60s	Any	>50	15–30s	30s–40s	<5	Any	<10	Any	<18	<18	>60
Ethnic predisposition	None	White	None	White	White	Asian	Asian, Middle Eastern	Asian	None	None	None	None	None	None
Clinical Features														
Constitutional symptoms	No	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No	No	No	No
Hypertension	Occas	Yes	Yes	Occas	Occas	Yes	No	Occas	No	No	No	Yes	Yes	Yes
Cutaneous manifestations	No	No	Yes	Yes	Occas	Occas	Yes	Yes	Occas	Yes	Yes	Yes	Yes	Occas
Arthralgias/arthritis	No	Occas	Yes	Yes	Yes	Yes ^b	Yes	Yes	Occas	Occas	Occas	Occas ^c	No	No
Ocular manifestations	No	Occas	Occas	Yes	Yes	Occas	Yes	Yes	No	Occas	Yes	Yes	Yes	Yes
Cerebrovascular accidents	Occas	Occas	Occas	Occas	Yes	Occas	Occas	No	Occas	Occas	Occas	Occas	Occas	Yes
Pulmonary infiltrates	No	No	No	Yes	No	Occas	No	No	Occas	No	No	No	No	No
Pulmonary hemorrhage	No	No	No	Yes	No	No	Yes ^d	No	No	Occas	No	Occas	No	No
Cardiac manifestation	Occas	Yes	Occas	Occas ^e	No	Occas	No	Yes	Occas	Occas	Yes	Occas	Yes	Yes
Abdominal pain	Yes	Occas	Yes	Occas	Occas	Occas	Yes	Occas	Occas	Yes	Occas	Occas	Yes	Occas
GI bleeding	Yes	Occas	Yes	Occas	Occas	Occas	Occas	No	Occas	Occas	Occas	Yes	Yes	Occas
Renal manifestations	No	Yes	Yes	Yes	Occas	Yes	No	Occas	Occas	Occas	Occas	Yes	No	Yes
Laboratory Findings														
Leucocytosis	No	No	Yes	Yes	Occas	Occas	Occas	Yes	Yes	No	No	No	No	No
Anemia	Occas	No	Yes	Yes	Yes	Yes	Occas	Occas	Yes	No	No	Occas	No	Occas
Elevated ESR/CRP level	No	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No	No	No	Occas
ANCA positivity	No	No	Occas	Yes	No	No	No	No	Occas	No	No	No	No	No
Complement levels	Normal	Normal	Low (occas)	Normal	Normal	Normal	Normal	Normal	High or low	Normal	Normal	Normal	Normal	Normal
HBsAg	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg
Blood culture positivity	No	No	No	No	No	No	No	No	Yes	No	No	No	No	No
Abnormal urinalysis	No	Occas	Yes	Yes	No	Yes	Occas	Occas	Occas	No	No	Occas	No	No
Immunotherapy	No	No	Yes	Yes	Yes	Yes	Yes	Yes	No	No	No	No	No	No

^a Other rare mimics include relapsing polychondritis, Cogan's syndrome, aortitis of tertiary syphilis, and the aortitis associated with seronegative spondyloarthritis. SAM = segmental arterial mediolysis; FMD = fibromuscular dysplasia; PAN = polyarteritis nodosa; AAV = ANCA-associated vasculitis; GCA = giant cell arteritis; TA = Takayasu arteritis; BD = Behçet's disease; KD = Kawasaki disease; NF = neurofibromatosis; PXE = pseudoxanthoma elasticum; occas = occasionally; GI = gastrointestinal; ESR = erythrocyte sedimentation rate; CRP = C-reactive protein; ANCA = antineutrophil cytoplasmic antibody; HBsAg = hepatitis B surface antigen.

^b Upper extremity claudication and thoracic back pain are relatively common.

^c Charcot joints can develop as a result of peripheral neuropathy. Also, bone lesions such as pseudoarthrosis and bone dysplasia are common.

^d Due to vasculitis of the pulmonary arteries.

^e In Churg-Strauss syndrome.

Our decision to proceed for surgery was based on the extensive splenic artery disease, though SAM was not initially suspected. But as our patient was multiparous with typical age with pathognomonic histopathological features SAM was strongly confirmed. Later on the patient underwent a brain CT angiography and cardiac evaluation that revealed absence of cerebral arteries abnormalities and any coronary artery disease [21]. Enhanced abdominal CT scan on 2 year follow up showed no involvement of the celiac trunk vessel.

Segmental arterial mediolysis with Multiple SAAs remains a rare finding of a rare disease. It should be suspected in patients with diseased vessels and the presence of several aneurysms. Abdominal angiography and CT brain should be performed to rule out both adjacent and distant vascular involvement, in order to plan further management. In our case complete splenic artery resection with splenectomy was the best treatment option because of the increased risk of potentially life-threatening sequelae.

Conflicts of interest

The authors declare no conflict of interest.

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Ethical approval

The case report was approved by the local ethic committee. of the Amiri teaching hospital. LEC-project number 28-2014.

Consent

Written informed consent was obtained from the patient and is available upon request. No patient identifying material was used in this manuscript.

Authors contribution

Data collection: Salah Termos* and Mohammad Alali.

Analysis and case management: Salah Termos, Ali Taqi and Hussein Hayati.

Writing and revising of the final version of the manuscript: Salah Termos*, Ameera JMS Alhassan and Ayman Adi.

Registration of research studies

UIN: researchregistry1241.

Guarantor

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The work has been reported in line with the SCARE criteria [23].

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