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SEGMENTAL ARTERIAL MEDIOLYSIS AND ITS MIMICKERS: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

This case report addresses segmental arterial mediolysis (SAM), a rare non-inflammatory vasculopathy. A 51-year-old man presented at the emergency department for epigastric and left upper quadrant pain. He had a history of arterial hypertension and had recently received methylprednisolone for knee pain. Blood tests revealed elevated C-reactive protein levels at 40 mg/l and lactate dehydrogenase levels at 496 IU/ml. Abdominal computerized tomography showed arterial thickening, arterial dilatations, and dissections of the splenic and renal arteries, leading to organ ischemia. This case emphasizes the importance of considering SAM in cases of unexplained abdominal pain or suspected arteriopathy.

KEYWORDS

Segmental arterial mediolysis, abdominal arteriopathy, arterial dissection, arterial aneurysm, vasculitis mimickers

LEARNING POINTS

- Segmental arterial mediolysis (SAM) is a rare and underdiagnosed vasculopathy.
- SAM is a challenging diagnosis and should not be confused with vasculitis.
- SAM has a good prognosis with spontaneous resolution in most cases.

CASE DESCRIPTION

A 51-year-old man presented at the emergency department for epigastric and left upper quadrant pain. He had a medical history of arterial hypertension treated with an angiotensin-converting enzyme inhibitor and calcium antagonist since 2017. Recently, the patient received methylprednisolone 32 mg/day. An eso-gastro-duodenoscopy revealed erythematous gastropathy. Methylprednisolone was suspended and a proton pump inhibitor initiated.

As the symptoms did not improve, the patient went to the Emergency Room (ER). Initial blood tests showed neutrophilic leukocytosis with elevated C-Reactive Protein, and elevated lactate dehydrogenase levels *(Table 1)*. Abdominal computerized tomography (CT) without contrast injection suggested pancreatitis of Balthazar score C. The patient was discharged with conservative treatment.

Several days later, he returned to the ER with worsening abdominal pain. Additional blood tests revealed elevated

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		Initial work-up			Follow-up	
					3 months	6 months
	Reference	21.01.2023	23.01.2023	31.01.2023	24.04.23	31.07.23
WBC	*3.5-11 x10³/mm³*	12.400	8.200	10.14	5.8	6.6
- Neutrophils	*1.5-6.7 x10³/mm³*	10.28	5.70	7.59	3.18	4.16
- Lymphocytes	*1.2-3.5 x10³/mm³*	0.93	1.22	1.06	1.59	1.54
Platelets	*150-440 x10³/mm³*	244	251	300	251	229
Hemoglobin	*12-18 g/dl*	16	14.2	12.9	14.8	14.8
CRP	*5 mg/l*	40.4	60.2	100	1.3	0.82
GOT	*< 40 U/l*	21	21	29	25	17
GPT	*< 41 U/l*	51	34	64	31	19
GGT	*10-71 U/l*	38	36	126	31	24
PAL	*40-129 U/l*		77	118	68	64
Bilirubin total	*< 1.2 mg/dl*		0.5	0.37	0.26	0.49
LDH	*220 IU/l*	496	403	283	184	159
Lipase	*75 IU/l*	35	1511	36		
Urea	*16.6-48.5 mg/dl*	31	40	14.4	37.1	37.8
Creatinine	*0.7-1.2 mg/dl*	1.1	0.9	0.9	0.97	1.12
Na+	*136-145 mmol/l*	136	134	135	141	140
K+	*3.5-4.5 mmol/l*	4.2	4.5	3.7	4.5	4.5
Cl-	*98-107 mmol/l*	101	100	99	105	105
Bicarbonate	*23-29 mmol/l*	25	30	25	25	26
ANA	*Negative*			Negative		
ANCA	*Negative*			Negative		
C3	*72-156 mg/dl*			128		
C4	*10-46 mg/dl*			54		

Table 1. Laboratory results

lipase levels *(Table 1)*. A CT with contrast injection revealed segmental circumferential thickening and dissection of the splenic and renal arteries associated with left renal infarction and mild pancreatic edema *(Fig. 1A, B)*.

Prophylactic anticoagulation with low-molecular-weight heparin (LMWH) was initiated, and the patient was transferred to a university hospital. A follow-up abdominal CT scan revealed an extension of arterial wall thickening and bilateral renal infarcts. Angiography confirmed the dissections and aneurysmal dilatations in the renal arteries' medial segment *(Fig. 1C)*.

Extensive investigations excluded differential diagnoses: Immunological work-up and 18F-Fluorodeoxyglucose Positron Emission Tomography - Computed Tomography

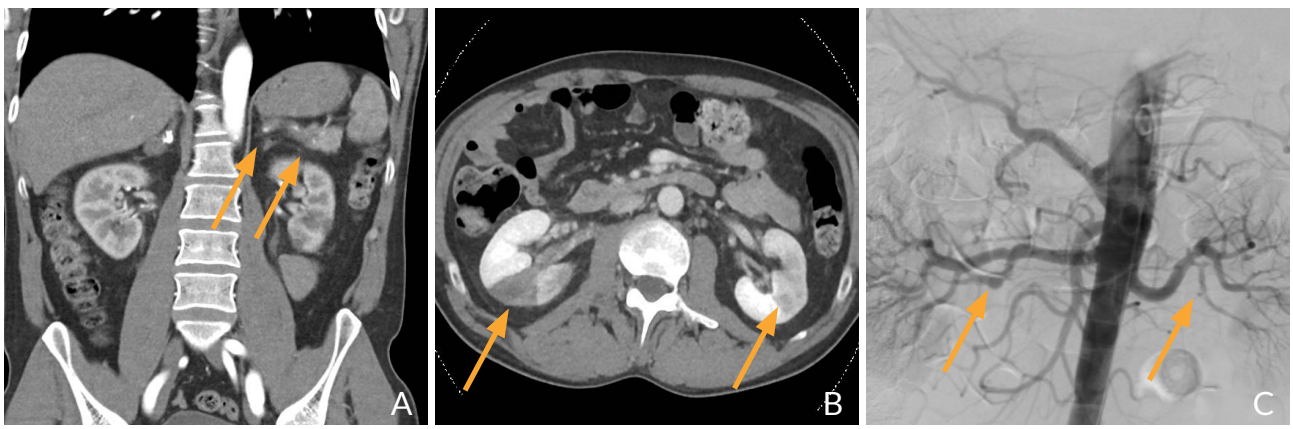

Figure 1. A: Early arterial phase CT shows the major circumferential hypodense thickening of the splenic artery (arrows). B: Late arterial phase CT reveals infarction of both kidneys (arrows). C: Arteriography demonstrates an alternation of dissections and aneurysmal dilatations of the renal arteries in their medial portions (arrows).

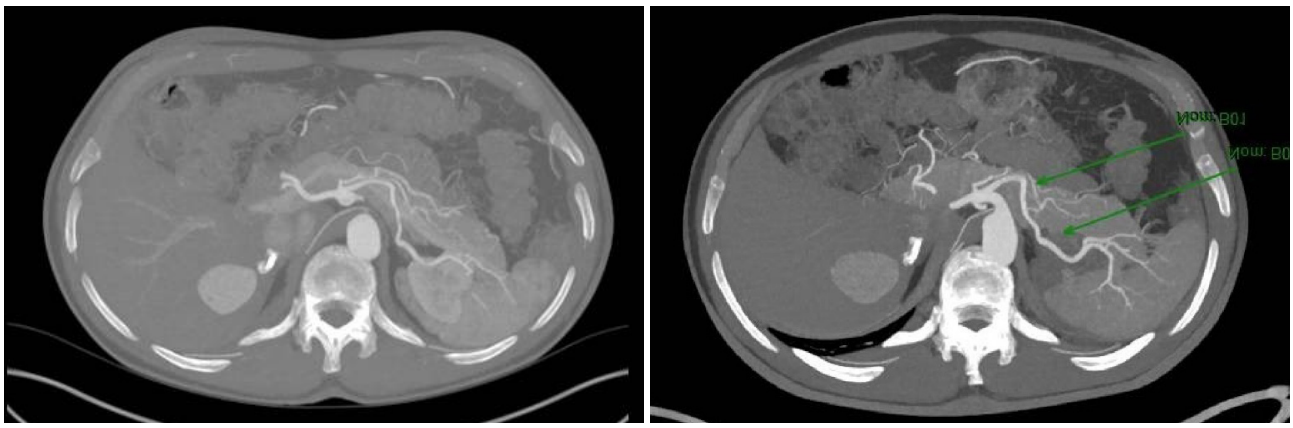

Figure 2. Three- and six-months follow-up. CT shows progressive improvement of the splenic arterial caliber and decreased arterial wall circumference (arrows).

Clinical criteria	Imaging criteria	Serologic criteria
Absence of: - Common disease (gastroenteritis, ulcer, appendicitis, or pancreatitis) - Collagen diseases - Fibromuscular dysplasia - Vascular disorders (atherosclerosis, or arteritis) Acute or Chronic presentation by: - Abdominal/Back/Chest pain - Transit disorders - Melena/Hematochezia - Hematuria - Intra-abdominal hemorrhage or shock - Cerebrovascular symptoms	Absence of associated contiguous aortic dissection or atherosclerosis. Presence of: - Dissection - Fusiform aneurysm - Occlusion - String of beads - Wall thickening - Rupture - With or without organ infarction	Absence of inflammatory markers: - Antinuclear antibodies - Antineutrophil cytoplasmic antibodies - Erythrocyte sedimentation rate - C-reactive protein*

* CRP level may falsely exclude a diagnosis of SAM, as organ infarction caused by SAM can secondarily induce elevated CRP.

Table 2. Criteria for diagnosis of segmental arterial mediolysis, adapted from Naidu et al.[1]

(^{18F}FDG-PET-CT) to rule out vasculitis; blood cultures and a transthoracic echocardiography to exclude mycotic aneurysm; cerebral and supra-aortic trunk imagery to rule out collagen disorders. Moreover, the French Vasculitis Study Group (FSVG) diagnostic criteria for Polyarteritis nodosa (PAN) were not fulfilled. A final diagnosis of segmental arterial mediolysis (SAM) was established, and the patient was shifted to therapeutic doses of LMWH. Arterial hypertension management was reinforced with beta-blockers.

Significant clinical, biological, and radiological improvement was observed during follow-up *(Table 1; Fig. 2)*. No relapse was observed after six months of follow-up.

DISCUSSION

SAM is a non-inflammatory vascular disorder affecting medium-sized vessels. First described by RE Slavin and JC Gonzalez-Vitale in 1976, SAM was initially classified as a vasculitis but later reclassified based on histopathological studies[1,2]. The reported incidence of SAM is 1 in 100,000 per year[3]. The pathophysiology of SAM remains unclear. Experimental studies suggest environmental exposure factors promoting vasoconstriction and local hypoxemia, leading to mediolysis[2].

Three large case series of SAM are discussed below and detailed in the Supplementary data[1-3]. SAM affects men in their fifties. Arterial hypertension has been associated with SAM, but its role is not well established. The most common clinical presentation is abdominal pain (69.2 - 82%), which may progress to hemorrhagic shock (4.2%). Asymptomatic patients are anecdotic (4.9 - 10.3%). The splanchnic network, including the superior mesenteric artery (43.6 - 53.1%), celiac trunk (35.7 - 54.7%), and renal arteries (25.9 - 52%), is most often affected, but cerebrovascular arteries can also be involved (13%). Radiological presentations of SAM include segmental dissections (60.8 - 95%) and aneurysms (52.1 - 76.2%), while stenosis (18.9 - 26%) and "string of beads" (14.7 - 31%) appearances are less frequent. Multiple vascular involvement is the most common presentation. CT angiography provides best accuracy for vascular diagnosis. In challenging cases, angiography may be used for both diagnosis and therapeutic purposes[1].

While histology remains the gold standard for diagnosing SAM, current practice relies on a combination of non-invasive evidence[1,3]. Naidu et al. proposed non-invasive diagnostic criteria *(Table 2)*.

Differential diagnosis encompasses some rare conditions, described below.

Fibromuscular dysplasia (FMD) is a non-inflammatory, segmental pathology of small and medium-size vessels, particularly in the renal and carotid arteries. FMD patients are often asymptomatic and typically involve women in their third decade[1,3]. Radiologically, FMD is characterized by focal or multifocal stenosis, often resembling a "string of beads". Additional radiologic features, such as aneurysms, dissections, or tortuous arteries are insufficient for an FMD diagnosis[3,4]. The etiology of FMD remains unclear, though genetic and environmental factors (tobacco and hormones) have been suggested[4]. Antiplatelet treatment (aspirin 75-100 mg/day) is recommended to prevent thromboembolic complications[4].

Since SAM mainly affects medium-size vessels, we focus on PAN, the main medium-size vasculitis in adults. PAN is mostly idiopathic but secondary PAN has been reported in 28% of cases[5]. PAN shows a slight predilection for males, with a median age of 53 years. PAN is suspected based on a classic triad of cutaneous signs, mononeuritis, and constitutional signs. Histology reveals arteritis with fibrinoid necrosis of the media, intimal proliferation, and lesions of varying ages. PAN does not harbor antineutrophil cytoplasmic antibodies nor histological granulomas. Renal involvement presents with renal artery microaneurysms, cortical infarction or reno-vascular hypertension. ^{18F}FDG-PET-CT is a useful imaging tool for exploring large vessels involvement. However, it may not be effective to investigate smaller vessels involvement seen in PAN. The differential diagnosis between SAM and vasculitis is crucial due to the distinct management and prognosis of these conditions.

Little is known about management of SAM. Corticosteroids are not recommended as they have not been proven effective and may even be detrimental[2]. Conservative treatment is advised for hemodynamically stable patients (pain relief, antihypertensive drugs, and avoidance of vasoconstrictors such as tobacco, cocaine, and pseudoephedrine). We preferred carvedilol, an alpha1-adrenergic blocker, to other beta-blockers for its relaxing action on smooth muscle vessels. For hemodynamically unstable patients, endovascular procedures are prioritized over surgery whenever possible[2]. The roles of antiplatelet therapy and anticoagulants remain uncertain, but they may be considered in the presence of organ ischemia or compressive thrombus[1].

Contrary to earlier case series reporting 25% mortality, the prognosis of SAM is favorable, with clinical improvement in 90.6% and radiological improvement in 71.5% to 83.8% of cases. Overall survival rates range from 93% to 100% at 1 year, and 95.7% at 3 years[1-3]. Clinical and radiological follow-up is recommended at 1, 3, and 9 months after onset and annually thereafter[1].

In conclusion, increased awareness of SAM can help avoid lengthy and sometimes invasive investigations and management errors.

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APPENDIX

	Naidu et al.[1] n (%)	Skeik et al.[3] n (%)	Peng et al.[2] n (%)
Clinical features			
- Age	51 (23-87)	55 (48-63)	52
- Men	79 (71%)	67.8%	79 (67.5%)
Medical background			
- Arterial hypertension	50 (55%)	42.7%	42 (35.9%)
- Hyperlipemia	32 (36%)	11.9%	35 (29.9%)
- Diabetes	3 (3%)	1.4%	5 (4.3%)
- Active smoking.	33 (37%)	11.9%	51 (43.6%)
Current medications			
- Aspirin	-	-	26 (22%)
- Clopidogrel	-	-	0 (0%)
- Anticoagulation	-	-	6 (7%)
- Antihypertensive	-	-	34 (29.6%)
- Corticosteroids	-	-	4 (3.4%)
- Statins	-	-	23 (19.7%)
Clinical presentation			
- Abdominal pain	74 (82%)	79.7%	81 (69.2%)
- Back pain	9 (10%)	-	23 (19.7%)
- Chest pain	6 (7%)	-	14 (12%)
- Diarrhea	5 (6%)	4.9%	-
- Nausea - Vomiting	4 (4%)	16.1%	-
- Hematuria	2 (2%)	-	10 (8.6%)
- None	6 (7%)	4.9%	12 (10.3%)
- Intra-abdominal hemorrhage	-	49.7%	-
- Melena - Hematochezia	-	5.6%	-
- Shock	-	4.2%,	-
- Cerebrovascular symptoms	-	5.6%.	-
Territory affected			
- Renal	47 (52%)	25.9%	58 (49.6%)
- Superior mesenteric	46 (51%)	53.1%	51 (43.6%)
- Coeliac trunk	46 (51%)	35.7%	64 (54.7%)
- Hepatic	23 (25%)	44.8%	30 (25.6%)
- Iliac	18 (20%)	-	18 (15.4%)
- Splenic	14 (16%)	24.5%	24 (20.5%)
- left gastric	5 (6%)	-	-
- inferior mesenteric	1 (1%)	10.5%	3 (2.7%)
- Cerebrovascular	-	13%	-
- Multiple.	-	62.2%	-
Number of arteries affected			
- 1	43 (48%)	-	-
- 2	32 (35%)	-	-
- 3	13 (14%)	-	-
- 4	5 (6%)	-	-
- 5	5 (6%)	-	-
- 6	2 (2%)	-	-
Image			
- Dissecting	86 (95%)	60.8%	93 (79.5%)
- Aneurysm	57 (63%)	76.2%	61 (52.1%)
- String of beads	28 (31%)	14.7%	18 (15.4%)
- Obstruction	19 (21%)	16.9%	26 (22.2%)
- Thickening of the wall	15 (17%)	7.7%	16 (13.7%)
- Rupture	-	45.5%	-
- Stenosis	-	18.9%	31 (26.5%)
- Thrombosis	-	14.7%	-
- Infarction	-	-	49 (41.9%)
Treatment			
- Coil – Embolization	1 (1%)	27.9%	-
- organ surgery	-	23.5%	-
- Artery Repair Surgery	-	20.6%	-
- Antihypertensive	-	19.9%	-
- Anticoagulation	35 (36%)	11.8%	-
- Antiplatelet therapy	46 (47%)	10.3%	-
- Angioplasty – Stenting	-	8.1%	-
- Conservative treatment	-	8.1%	-

Table S1. Comparative data from literature.

	Naidu et al.[1] n (%)	Skeik et al.[3] n (%)
Survival	100%	93%
Clinical evolution Improvement Steady Degradation	 - - -	 90.6% 5.2% 4.2%
Radiological evolution Improvement (I) Steady (S) Degradation	 I + S = 76 (80%) 19 (20%)	 58.8% 66.3% 17.5% 16.3%

Table S2. 12-months follow-up of SAM in the literature.

	Peng et al.[2] n (%)
Survival	95.7%
Clinical evolution Improvement Steady Degradation	 - - -
Radiological evolution Improvement (I) Steady (S) Degradation	 I + S = 71.5% 28.5%

Table S3. 36-months follow-up of SAM in the literature.