Acute abdominal pain: a challenging diagnosis

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Abstract

We hereby describe a case of an acutely ill 41-year-old male without any medical history who presented with an acute abdomen in the emergency department. An abdominal CT showed a dissection of the coeliac trunk and infarction of the spleen. Because of a presumed diagnosis of vasculitis he was started on high dose IV steroids. However, after additional testing the diagnosis of segmental arteriolar Mediolysis (SAM) was made. In this case report we describe the presentation, diagnosis, treatment and follow-up of this patient and provide the readers with background about common differential diagnosis and criteria for diagnosing SAM. (Acta gastroenterol. belg., **2022,** 85, **1-2**).

Keywords: Acute abdominal pain, challenging diagnosis, vasculitis, Nonatherosclerotic Abdominal Vasculopathie, SAM, Case report

Introduction

Acute abdominal pain can be caused by many common medical conditions, however sometimes diagnosing the underlying illness is not so straightforward. Segmental arterial mediolysis (SAM) is a rare disorder and the diagnosis might be very challenging. It presents it-self mainly in middle aged, healthy adults but the complications may be severe. Due to the rarity and heterogenic presentation a lot of cases are presumably missed or patients are treated for other diseases with similar presentation. This potentially exposes patients with SAM to iatrogenic risk for further ionizing or invasive tests and treatments that are not indicated. Missing the diagnosis of SAM can be prevented if clinicians are better aware of this interesting clinical entity and are more familiar with its presentation and management, as illustrated in this case report.

The case history

A 41-year-old man was admitted after he fell unwell at work. He had no previous medical or family history. He complained of acute abdominal pain (respiration and movement dependent) and nausea with one episode of vomiting. No fever, dyspnoea, chest pain, cough or urinary complaints were present. Heart- and lung examination revealed no abnormalities. The abdominal examination showed a non-distended abdomen with diffuse tenderness (more in the epigastric region and left flank) without guarding or rebound tenderness.

Laboratory tests showed a normal hemoglobin (15.8 mg/dl and NV: 14.0-18.0 mg/dl), platelets (333 x10^9/L, NV: 150-450 x10^9/L), White blood cell (WBC) counts (9.12 x10^9/L, NV: 4.00-10.00 x10^9/L), coagulation parameters, kidney function, electrolytes, liver enzymes, lipase, creatinine phosphokinase (CPK), Lactate dehydrogenase (LDH) and thyroid function. C-reactive protein (CRP) was 55 mg/dl (Upper limit of normal (ULN): 5 mg/dl) and erythrocyte sedimentation rate (ESR) was very mildly elevated (23 mm/h, ULN: 10 mm/h). Human immunodeficiency viruses (HIV), Treponema pallidum, hepatitis B and C serology were negative. Complement studies and autoimmune screening were normal. Urinary analysis (UA) and an arterial blood gas (ABG) were non-remarkable.

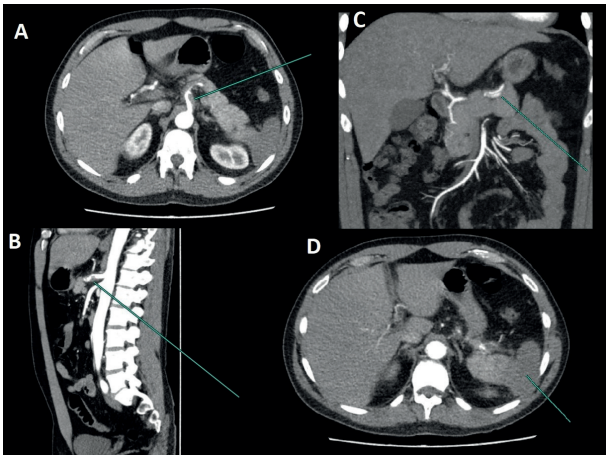

Fig. 1. — CT angiography of the Abdomen: (A) Thrombus in the Coeliac trunk in an Axial plane (B) Thrombus in the Coelic trunk in a Sagittal plane (C) The splenal artery dissection (D) The splenic infarction in the lower lobe.

Abdominale X-ray (AXR) showed no pathological signs. Additional computed tomography (CT) angiography showed a dissection of the coeliac trunk with a wall-adhering thrombus with a moderate obstructing effect and a dissection in the splenic artery that caused an infarction of the lower lobe of the spleen (figure 1). There were no signs of obstruction, bowel ischemia or pathological findings in other organs.

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As the provisional diagnosis was vasculitis, high doses of IV steroids (methylprednisolone 500mg IV, once daily (OD)) were started in combination with therapeutic dose of enoxaparin. Our main differential diagnoses were Takayasu arteritis, Polyarteritis Nodosa (PAN) and Segmental arterial mediolysis (SAM). An additional total body Positron emission tomography (PET)/CT scan showed no evidence of vasculitis. Additional Transthoracic echocardiography (TTE) and 24 hour tape were normal.

During admission the patient suffered from multiple side effects of the treatment such as insomnia, fluid retention, elevation in blood glucose levels and significant arterial hypertension. The case was discussed in a multidisciplinary meeting and based on the clinical, biochemical and radiology features the diagnosis of SAM was made. The IV steroids were safely stopped and the patient was discharged with Amlodipine (5mg oral, OD) and therapeutic Enoxaparin.

There was complete recovery of all symptoms and CT Angiography showed stable signs of dissection and partial resolution of the thrombus at 3 months follow-up. Enoxaparin was stopped and switched to lifelong Aspirin for secondary prevention.

Table 1. — **Diagnostic criteria for SAM (3,4)**

Criteria	Presentation
Patient population	Middle aged (mostly > 50 years old), both sexes are equally affected Absence of a congenital predisposition for dissections (Marfan, Ehlers-Danlos, Loeys-Dietz).
Clinical	Acute presentation with abdominal/flank pain, back or chest pain, stroke or hematuria caused by ischemia/infarctions of intra-abdominal organs or brain Severe: hemorrhagic shock from ruptured intra-abdominal aneurysm or dissecting hematoma
Laboratory	Absence of significant inflammatory markers and auto-antibodies (antinuclear, antineutrophil cytoplasmic), normal complement levels
Radiologic	Simple (wall) thickening of mesenteric or renal arteries, single or multiple aneurysm, dissecting hematomas, arterial stenosis and occlusions, no associated atherosclerosis
Pathologic	Injurious phase: smooth muscle cells of outer media layers affected with vacuolization and lysis with separation from the adventitia without significant inflammatory signs Reparative phase: granulation tissue and healing fibrosis

Discussion

SAM was first described as a disorder in 1976 and belongs to the group of Nonatherosclerotic Abdominal Vasculopathies (1). The pathogenesis is unclear but might be related to excessive response to vasoconstrictor stimuli. The clinical presentation is very heterogenic. In a review of reported cases, the major associated signs and symptoms included abdominal pain (68%), followed by hypovolemic shock (25%), neurological symptoms (12%) and sudden death (11%) (2). The confirmation of the diagnosis can be made with a vessel biopsy showing vacuolar degeneration of smooth muscle cells in the outer media layer, absence of significant inflammation and a healing fibrosis. As in most patients these biopsies are not easily available, clinical criteria are developed (Table 1) (3). In our case, there were no arguments for a congenital predisposition for dissections (eg. Ehlers-Danlos, Loeys-Dietz, Marfan), and no clinical or radiological evidence for fibromuscular dysplasia (FMD), atherosclerosis or arteritis. In addition, the laboratory results show normal auto-immune screening and complement. The two main clinical differential diagnoses are PAN and FMD. PAN classically presents itself with micro-aneurysms and infarctions, prodromal symptoms, positive hepatitis B antigen, elevated ESR, anaemia and a positive PET/CT scan (4). Arterial dissection is not a prominent feature. FMD is most commonly seen in young females and typically shows the typical "string of beads" pattern on imaging. Renovascular hypertension and CNS-symptoms are frequent (4). SAM has no specific treatment but the available data show stabilization and improvement on follow up under lifelong secondary prevention (5).

Conflict of interest

The authors have no conflict of interest to declare.

References

1. SKEIK N, HYDE JR, OLSON SL, THALER CM, ABUATIYEH W, AHMED AK, *et al.* Nonatherosclerotic Abdominal Vasculopathies. *Ann Vasc Surg.* 2019, **6**: 128-46.
2. KIM HS, MIN S, HAN A, CHOI C, MIN S-K, HA J. Longitudinal Evaluation of Segmental Arterial Mediolysis in Splanchnic Arteries: Case Series and Systematic Review. *PLOS ONE*. 2016, **11**(8): e0161182.
3. KALVA SP, SOMAROUTHU B, JAFF MR, WICKY S. Segmental arterial mediolysis: clinical and imaging features at presentation and during follow-up. *J Vasc Interv Radiol*. 2011, **22**(10): 1380-7.
4. FILIPPONE EJ, FOY A, GALANIS T, POKUAH M, NEWMAN E, LALLAS CD, *et al.* Segmental arterial mediolysis: report of 2 cases and review of the literature. *Am J Kidney Dis*. 2011, **58**(6): 981-7.
5. MICHAEL M, WIDMER U, WILDERMUTH S, BARGHORN A, DUEWELL S, PFAMMATTER T. Segmental Arterial Mediolysis: CTA Findings at Presentation and Follow-Up. *American Journal of Roentgenology*. 2006, **187**(6): 1463-9.