

Arterio-venous and lymphatic malformation mimicking acute appendicitis in a patient. Case report and review of literature

Talal Majeed, Jennifer Allan, Conor Magee, Jeremy Wilson

From Specialty Registrar, Department of Coloproctology, Wirral University Teaching Hospital, Wirral, United Kingdom.

Corresponding to: Dr. Talal Majeed, Department of Coloproctology, Wirral University Teaching Hospitals, Arrowe Park Road, Birkenhead, Wirral CH49 5PE, United Kingdom. E-mail: talal.majeed@nhs.net.

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ABSTRACT

Arteriovenous and lymphatic malformations are a common phenomenon and can present in various clinical forms. They rarely present as a cause of acute abdomen, and in the literature, there are only a few cases where intra-abdominal arteriovenous malformation (AVM) presented as acute appendicitis in the adult population. Most of the cases are diagnosed post-operatively on histology but the previous reporting in the literature has made it possible for the radiologists to look for these anomalies and consider them as a probable cause of acute abdomen when no other obvious cause is found. We are presenting an interesting case of congenital vascular malformation mimicking acute appendicitis in a young female. She had a previous history of complications related to AVM and her complicated urological history was initially focussed causing a delay in referral to surgeons and imaging which lead to the final diagnosis.

Keywords: *Acute appendicitis, Haemangioma, Lymphatic malformation, Vascular malformation.*

Acute appendicitis is the most common cause of acute abdominal pain with an incidence of 5.7-57/100,000 population [1]. Arteriovenous malformations (AVM) are congenital in nature and usually found sporadically. While in only 2% of cases they are multiple, most of them are solitary [2]. According to a study, only 12% of AVM become symptomatic in the course of life [3]. In rare instances, AVM has been reported to cause acute abdominal pain. In the literature, there are few cases where hemorrhage caused by AVM can present as acute abdomen and all the cases were diagnosed postoperatively [4-7].

The case we are presenting is unique as hemorrhage caused by an AVM in common iliac vein and external iliac artery caused inflammation around the appendix and adjacent area presented clinically as acute appendicitis. The patient was diagnosed

preoperatively by imaging and not subjected to surgery, as the episode proved to be self-limiting.

CASE REPORT

A 23-year-old young girl presented with 3 days history of lower abdominal pain which started suddenly and then radiated to her right groin which she described in her own words felt like a 'stitch'. There were no aggravating or relieving factors for the pain. Her pain symptoms were accompanied by urinary frequency and offensive smell in her urine for the last three days. She recently had cough and coryza which settled on its own. The pain progressively worsened and forced the patient to seek medical attention. Her past medical history included congenital

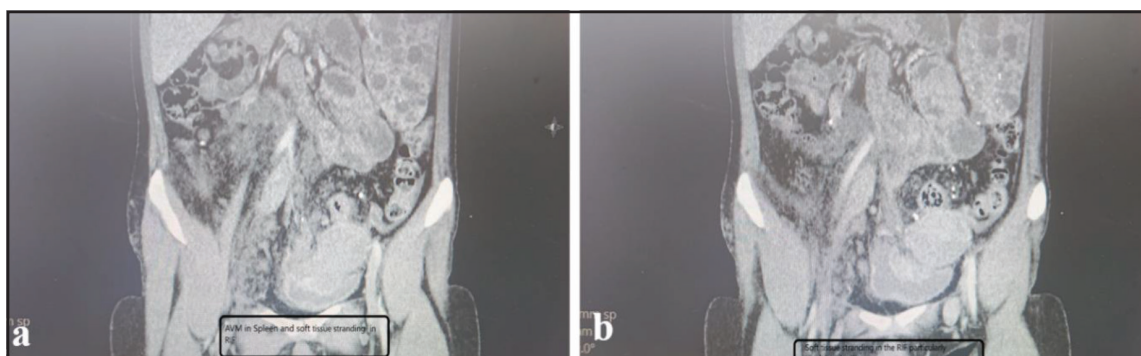


Figure 1: CT scan coronal plane image of the abdomen and pelvis showing (a) AVM/Lymphatic malformation in spleen and pelvis and (b) extensive AVM and soft tissue stranding in the pelvis around Iliac vessels and RIF close to appendix.

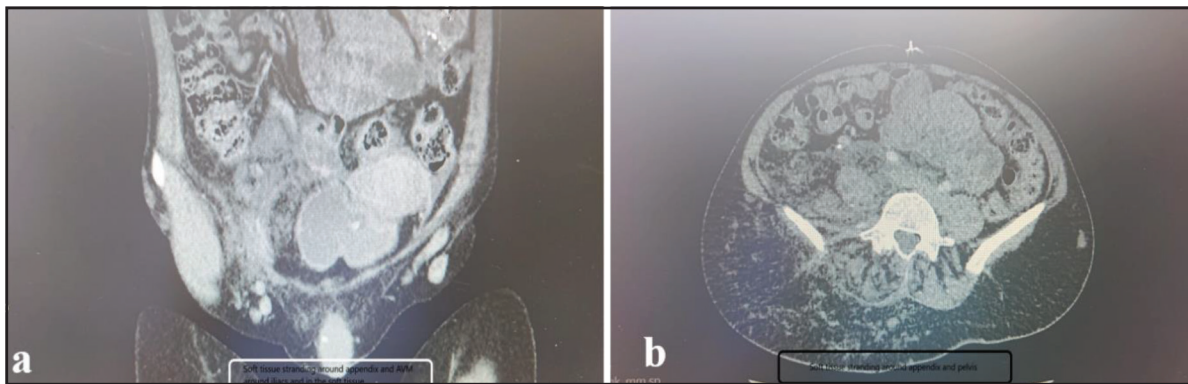


Figure 2: CT scan (a) coronal plane image of the abdomen and pelvis showing distribution of AVM in abdomen and soft tissue in the perineum; (b) transverse plane image of the abdomen and pelvis showing soft tissue stranding around appendix and pelvis

hemangiomas, previous hemorrhagic hemangioma in the urinary bladder, neurogenic bladder, a mic-key button for urinary bladder and large infected hemangioma in the left buttock. She was taking ferrous sulphate for iron deficiency anemia and cyclizine for sickness. Her social history included working as a social care worker and occasional drinker.

She was initially assessed by the urologist in view of previous complex urological history and after an initial assessment; they were not convinced that she has any urological problem and asked general surgeons to assess the patient for possible acute appendicitis. The surgical team examined her in the surgical assessment unit. The surgical team particularly made a note of her previous history of haemangiomas in the bladder and the buttock area.

On examination, her abdomen was found to be soft but markedly tender in the right iliac fossa (RIF) with signs of localized peritonism. They were able to elicit rebound tenderness in the RIF and percussion tenderness. They also examined the left buttock area for possible collection secondary to hemangioma but could not find any evidence for that. Her vitals were found to be normal except tachycardia of 116/min and a recent spike of temperature up to 37.6 F.

Her white cell count was $9.3 \times 10^9/L$, neutrophil count $7.5 \times 10^9/L$, hemoglobin 116g/L, C-reactive protein (CRP) 116mg/L and lactate of 0.9. Urinalysis showed some leucocytes and nitrites. The pregnancy test was negative on urine analysis.

A decision was made to do imaging before attempting diagnostic laparoscopy. Because of complex history, multiple procedures in the past and non-availability of magnetic resonance imaging (MRI) scanner facility in out of hours, it was decided to perform computed tomography (CT) scan of the abdomen and pelvis with contrast. CT scan was reported initially by a trainee radiologist who commented on appendix as completely normal but changes throughout the lower abdomen, pelvis and subcutaneous tissue related to vascular malformations (Fig. 1 and 2).

An amendment came later from a gastrointestinal radiologist, who indicated some linear soft tissue stranding in the RIF anterior to the common iliac vein and external iliac artery. He also mentioned that it was unclear whether this mild stranding is acute or chronic and try to explain it that it might have caused by a hematoma secondary to rupture of one of the AVM in the iliac

vessels. The radiologist also noted high CRP, temperature and signs of peritonism on notes and opined that it could probably be due to free blood in the pelvis resulting from haemorrhage from AVM. He also mentioned noticing multiple AVM, and lymphatic malformation throughout the abdomen particularly in the spleen, pelvis, soft tissue and perineum. The radiologist looked at previous MRI which was done six months ago and found that AVM was present throughout the above-mentioned locations but no other changes. Hence, a diagnosis of inflammation secondary to hemorrhage from AVM in the vicinity of the appendix was made and decided to treat her symptoms conservatively. The patient was observed for the next 48 hours as her symptoms started to settle down and no attempt was made to do diagnostic laparoscopy. The patient was discharged when the pain completely settled. The patient was followed up in a month and was found to have no abdominal symptoms.

DISCUSSION

Arteriovenous malformation is an abnormal communication between an artery and a vein which bypasses the capillary bed and because of this, shunted blood flow and autonomous growth can lead to ischemia and formation of a tangled mass of abnormal friable vessels called nidus. Nidus can easily rupture and can lead to hemorrhage. The formation of AVM is unclear and they are usually picked up on examination if they are superficial or on imaging when they are situated deep in the body. AVM rarely becomes symptomatic and there is a paucity in the literature about symptomatic AVM. Intrabdominal AVM can lead to pain, inflammation, bleeding, anemia secondary to bleeding and as in most of the cases remain innocuous.

Some authors have reported finding AVM as the cause of appendicitis, where malformation was found postoperatively in the mesoappendix. Bleeding and pressure effect lead to the presentation of symptoms of acute abdominal pain in the lower right side of the abdomen. Emil *et al.* in 2005 reported a case of acute appendicitis due to inflammation caused by Caecal AVM [8]. Parikh *et al.* in 2015 also reported a similar case [9].

Other vascular anomalies like haemangiomas can present as acute abdomen and there are several cases reported in the

literature. Geramizadeh *et al.* have reported five cases (from 1974-2015) of finding symptomatic haemangioma presenting as acute abdomen, where original presentation in three cases was of acute lower abdomen pain [10]. Two more cases were reported later, where symptomatic haemangiomas presented as acute abdominal pain [11-12]. There are two cases where haemangiomas were the cause of intra-abdominal hemorrhage, which was not life-threatening and treated conservatively [10].

Although AVM and haemangioma are completely two different pathologies, the reason to discuss them together is to stress that when looking for the cause of acute abdominal pain, radiologists should look for abnormal vasculature, as rarely vascular anomalies and their complication can mimic some of the intra-abdominal pathologies. If these vascular anomalies can pick up pre-operatively during imaging, we can save unnecessary surgery. The literature review has shown that rarely these intrabdominal vascular malformations can lead to life-threatening condition, and most of the time they are an innocent bystander.

There is a paucity of the literature where congenital AVM was reported presenting clinically as acute appendicitis. Radiologists should be looking for vascular anomalies if the patient presenting with acute abdominal pain and no cause to explain the pain is found. Early preoperative detection by imaging can save unnecessary surgery. MRI scan or CT scans with intravenous contrast are the best modalities to look for vascular malformation. The multi-disciplinary approach should be adopted by surgeons and the radiologists especially when the past medical history of the patient is suggestive of congenital vascular anomalies. The natural history of intra-abdominal vascular anomalies suggests a conservative approach to treat complications caused by them.

CONCLUSION

Intra-abdominal AVMs are rare and symptomatic AVM are very rare. Patients with multiple AVM presenting with acute abdominal pain should be approached with caution and holistic approach should be adopted to look for the cause of abdominal

pain. Preoperative imaging and multi-disciplinary discussion between a surgeon and radiologist can save unnecessary surgery.

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