

Received: 2012.07.26  
Accepted: 2013.02.18

## Giant congenital malformation of the perirectal plexus in computed tomography imaging – case report

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### Summary

**Background:**

Congenital arteriovenous malformation (AVM) in the pelvic area is uncommon in males.

**Case Report:**

The described case is of a giant lesion of this type that caused recurrent hemorrhaging in the lower part of the gastrointestinal tract. Preliminary diagnosis of vascular pathology was made on the basis of an endoscopic examination that revealed numerous pulsating protuberances of the rectal wall, in which blood flow was identified by means of transrectal ultrasonography. Complementing the diagnostics with a CT revealed a considerable extent of malformation, as well as its morphology and anatomical relations with the surrounding tissues.

**Results:**

Following a two-year follow-up period, the malformation did not progress or demonstrate any intensification of clinical symptoms, therefore the patient continues to undergo conservative treatment.

**Key words:**

**Pelvic arteriovenous malformation (pAVM) • lower gastrointestinal tract bleeding • spiral computed tomography (CT) • computed tomography angiography (angio-CT) • 3D reconstructions**

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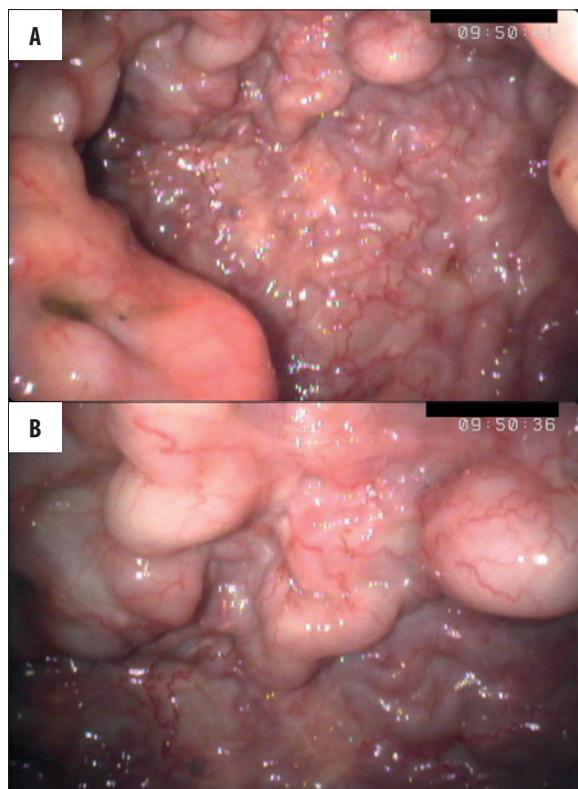
### Background

Arteriovenous malformations are rare vascular lesions caused by numerous abnormal connections between veins and arteries bypassing the capillary system. They are fast-flow malformations, contrary to the more frequent venous, lymphatic and capillary malformations [1]. They may be congenital or acquired, and are located in various body parts, usually the limbs, head and neck. Their pelvic location is uncommon, especially in males [2]. In most cases of pelvic AVMs described in literature, the main vessel supplying the *nidus* is one of the internal iliac arteries [2,3].

The case presented here is that of a male patient suffering from abdominal pain and recurrent bleeding from the lower part of the gastrointestinal tract, caused by congenital malformation of the perirectal plexus, supplied by a considerably dilated inferior mesenteric artery.

### Case Report

A 34-year-old male, suffering from recurrent bleeding in the lower part of the gastrointestinal tract, was admitted to the Clinical Gastroenterology Department of the 4th Military Hospital in Wrocław, Poland, in September 2010, for diagnostic follow-up purposes. The patient had previously undergone colonoscopy at a different medical center. He had been diagnosed with rectal tumor, but the histopathological examination of the tissue failed to determine the character of the pathology. On admission, it was established that the patient had mild hypochromic anemia, iron deficiency and slightly prolonged APTT. Following a colonoscopy performed at our center, numerous pulsating protuberances of the rectal wall were identified, protruding 1–2 cm into the lumen. The protuberances were covered with smooth, glossy mucous membrane with a visible dilated network of submucosal vessels (Figure 1). The endoscopic image of rectal lesions implied their vascular nature, which

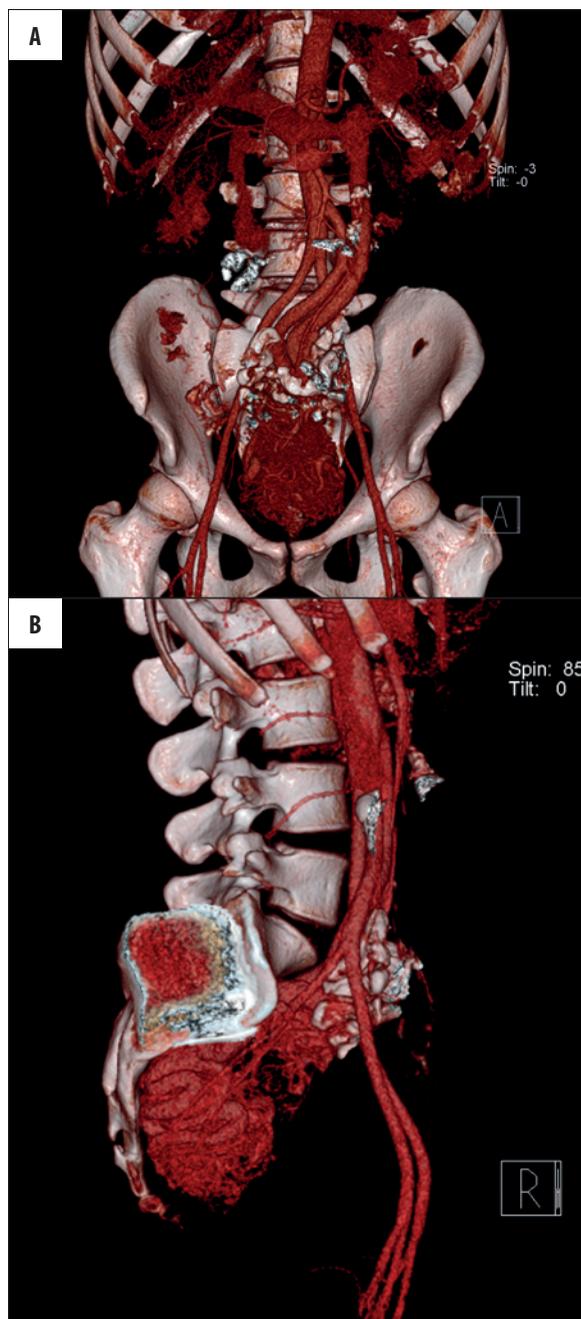


**Figure 1.** Colonoscopy: visible numerous, pulsating protuberances in rectal wall covered with smooth, glossy mucous membrane with a widened network of submucosal vessels.

was confirmed by ultrasound endoscopy, revealing blood flow in the pathologically dilated blood vessels. In order to precisely establish the extent and character of the lesions, diagnostic process was complemented by computed tomography angiography (angio-CT).

The CT examination was performed by means of a 128-slice dual source scanner and covered the abdominal and pelvic areas, from the supradiaphragmatic region to the level of ischial tuberosity. The AVM was revealed in the small pelvis, with a thick network of vessels surrounding the rectum, ranging in overall size from 6×7.5 cm in transverse plane to 11.6 cm in the long axis. The lesion started directly below the promontory of the sacrum and ended at the level of the sphincters; leaning against the front surface of the sacrum, it completely filled the space of the small pelvis (Figure 2). The veins in the malformation were up to 13 mm wide, arteries – up to approximately 11 mm and with sinuous courses. The lesion was supplied by the inferior mesenteric artery, dilated to approximately 12 mm, and the outflow was through the mesenteric vein, dilated to approximately 13 mm. The hepatic portal vein was also revealed to be dilated.

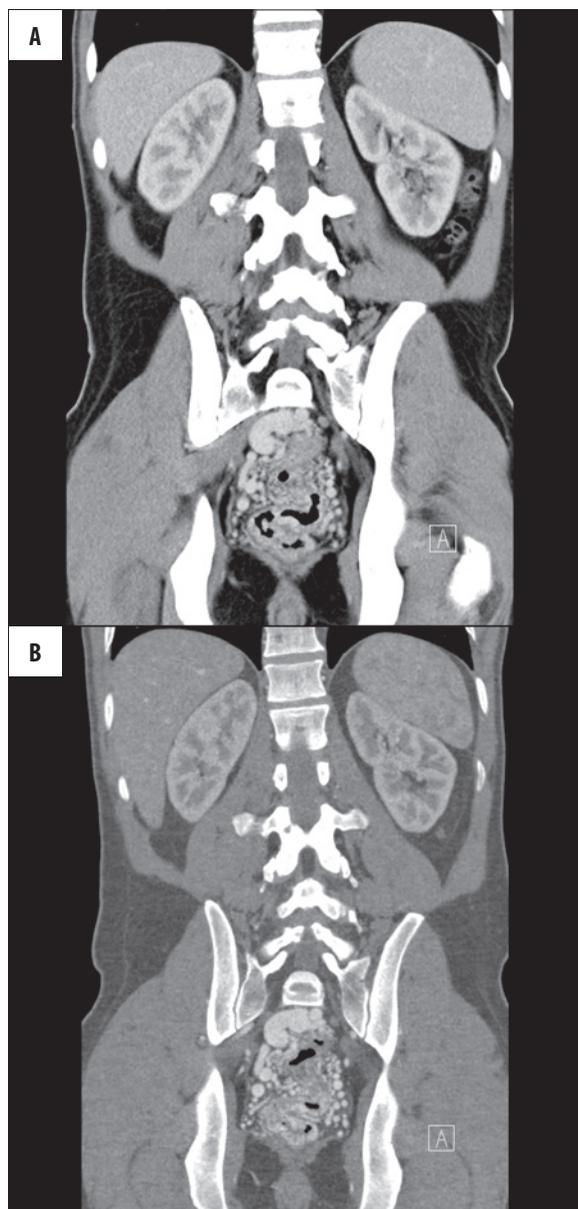
The patient was consulted at a vascular surgery and interventional radiology center. After performing classical angiography and assessing previous examinations, it was agreed upon with the patient to abandon the surgical and intravascular treatment. When making this decision, the size of the lesion was taken into consideration, as well as the lack of possibility to determine the complications



**Figure 2.** CT examination, VRT reconstructions, anterior (A) and right-hand view (B): visible arteriovenous malformation filling a considerable part of the small pelvis.

related to a possible ischemia of organs in the pelvis. The patient remained in observation and the treatment was limited to iron supplementation.

In February 2012, the patient was hospitalized again in the Clinical Gastroenterology Department of the 4<sup>th</sup> Military Hospital in Wrocław in order to have the progression of the lesion and possible complications assessed. During two years of observation, only minor instances of rectal bleeding connected with considerable physical exertion were observed in the patient. Iron supplementation administered orally allowed for hemoglobin levels to remain within the



**Figure 3.** CT examinations, MPR reconstruction, frontal plane. The image of pelvic AVM in CT examination performed approximately two years later (B) shows no essential changes in comparison with the initial examination (A).

norm. Neither physical examination, nor ultrasonography of the heart revealed any indication of heart failure caused by the AVM. An angio-CT examination revealed no progression of the described lesion (Figure 3A, 3B). The patient was advised to avoid hard physical work, undergo regular complete blood count examinations and supplement possible iron deficiencies.

## Discussion

Arteriovenous malformations (AVMs) are usually congenital, yet those located in the pelvic area tend to be acquired, secondary to cancer, trauma or surgery [2,4]. Congenital forms usually develop slowly, over extended periods of time, before becoming symptomatic. Their natural course

is characterized by mild onset and late expansion of the lesions, ultimately leading to infiltration and destruction of neighboring soft tissue and bones [1].

In about a fifth of patients with pelvic AVM, the malformation has an asymptomatic course. In the remaining cases, it may be identified by means of palpation of the abdomen or transrectally, as a pulsatile mass. It may result in pain or discomfort in the abdomen or pelvis, rectal pain and tenesmus, disorders of the genitourinary system, such as hematuria, dysuria, polyuria, hydronephrosis, hematospermia, impotence, dyspareunia, orchitis, sometimes also edema of the lower limb. Large AVMs may lead to congestive heart failure [2–4]. There are also cases described in literature in which AVMs are known to damage part of the sacrum and, by pressing on the nerve, cause sciatic neuralgia [5]. Hammad [6] describes a case of pAVM manifested by ischuria resulting from pressure on the bladder.

In the patient presented here, the vast tangle of vessels was located around the rectum, therefore the first manifestation of the lesion was a hemorrhage from the lower part of the gastrointestinal tract, leading to symptoms of anemia. The bleeding was most probably caused by dilated blood vessels in the rectal wall [3], which the performed colonoscopy revealed as polyp-like protuberances of mucosa, initially interpreted as some type of indeterminate tumor.

Imaging plays a key role in AVM diagnostics. It is possible to make a preliminary diagnosis by means of transrectal ultrasonography with the color flow Doppler function, often used in patients with disorders of the lesser pelvis. It is also a valuable method used in further monitoring of detected lesions [7]. However, classical angiography is still considered the “golden standard” in diagnosing AVMs. On the other hand, it does not always achieve the expected results. Rapid flow of blood through the dilated vessels of the malformation often makes it difficult to fill them completely and get a precise image by means of this technique. CT becomes helpful in such cases – without overdosing contrast or radiation, modern scanners show both the extent of the lesion and (thanks to their excellent 3D definition) the tiny vessels of the malformation, as well as its connections with other vessels of the circulatory system.

According to the recommendations proposed by Calligaro [3], pelvic AVMs which are asymptomatic, mild or non-progressive should not be treated, but rather monitored every 6 to 12 months. Therapeutic intervention should be indicated – apart from the enlargement of the lesion – by heart failure caused by large malformation, serious disorders in the pelvic region or hemorrhage from the malformation [8]. Surgical excision may only be effective in case of minor lesions that are well isolated from the neighboring tissue and that can be removed entirely, and the use of pre-surgery embolization contributes to the reduction of malformation size and the amount of blood loss during the procedure [3].

Currently, the method of choice for treating symptomatic malformations is an endovascular embolization with the use of various kinds of materials, which allows the control of symptoms for a certain period of time. Unfortunately,

relapses are common and require further procedures. They are caused by the processes of recanalization and neovascularization as a result of impermanent closure of the *nidus* [9]. Some alternative concepts have appeared, though. Intraoperative transvenous embolization, preferably with ethylene glycol [10], proves effective in some cases. Another novel method is stent-graft implantation combined with alcohol sclerotherapy [11].

The breakthrough came with the use of absolute alcohol as an obliteration agent during selective catheterization or direct puncture of the lesion. The procedure leads to a complete, permanent closure of the vascular lumen, resulting in good long-term effects, not only clinically but also

radiologically. However, possible complications connected with the use of concentrated alcohol must be considered, including local tissue damage with nervous necrosis and paralysis, renal failure, thrombus or contraction of pulmonary arteries [12].

## Conclusions

1. Congenital arteriovenous malformation located in the pelvic region is uncommon in males and may reach considerable sizes, remaining clinically dormant.
2. CT angiography is useful for the assessment of the extent of the vascular malformation, its relation with neighboring tissue, and morphology of the lesion itself.

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