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Giant Congenital Pelvic AVM Causing Cardiac Failure, Diplegia, and Neurogenic Bladder

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Summary

Background:

Pelvic arteriovenous malformations (AVMs) are uncommon lesions and only a rare number of male cases have been reported. Their clinical presentations are variable and imaging modalities have an important place in diagnosis and treatment planning.

Case Report:

We present the imaging findings of a giant congenital pelvic AVM that was diagnosed in a 30-year-old male patient eight years ago and which progressed despite follow-up and treatment, causing cardiac failure, diplegia, and neurogenic bladder.

Conclusions:

Pelvic AVMs are uncommon lesions and they can present with various symptoms based on their locations and sizes. Delays in the diagnosis and treatment can cause local and systemic complications. Imaging is very important in the diagnosis of pelvic AVM.

MeSH Keywords:

Angiography • Arteriovenous Malformations • Pelvis • Tomography Scanners, X-Ray Computed

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Background

Arteriovenous malformations (AVMs) are uncommon vascular lesions that result from multiple abnormal connections between arteries and veins [1]. Pelvic arteriovenous malformations (AVMs) are uncommon lesions and only a rare number of male cases have been reported in the literature [2]. Pelvic AVMs are generally acquired lesions and they are caused by tumors, traumas, or surgical procedures to that area, and congenital pelvic AVMs generally remain asymptomatic. [2,3]. They have variable clinical presentations and their imaging modalities play a vital role in making a diagnosis, evaluating the spread of the lesion, and planning and guiding the treatment. The purpose of this case report was to present the imaging findings of a giant congenital pelvic AVM in a male patient with cardiac failure, diplegia, and neurogenic bladder. To our knowledge, our case has the largest pelvic AVM in the literature and there are no AVM cases in the literature that caused diplegia and neurogenic bladder.

Case Report

A 30-year-old male patient was admitted to our hospital with a complaint of weakness in both legs that had begun

two months before. The patient's anamnesis showed that he had been diagnosed with pelvic AVM eight years ago and he had undergone embolization 14 times during the follow-up period. For the last five years, he had complaints of apnea associated with severe cardiac failure that developed secondary to AVM. For the last 6 months, he had a complaint of urinary incontinence. Upon physical examination he had abdominal tenderness. A manual muscle test showed muscle weakness in the hip flexors (3/5 on the right; 4/5 on the left), the knee extensors (3/5 on the right; 3/5 on the left), the ankle extensors (1/5 on the right; 2/5 on the left), and the muscles of the big toe (0/5 on the right; 1/5 on the left). There was sensory loss in the right L4-L5 and S1 dermatomes. A significant spasticity was present in both lower extremities. The following vital signs were obtained upon triage: a temperature of 36°C (oral), a heart rate of 102 beats per minute, a respiratory rate of 23 breaths per minute, and a blood pressure of 90/60 mmHg. Laboratory tests revealed a total leukocyte count of $5.3 \times 10^9/L$, a hemoglobin level of 7.9 g/dL, a serum bilirubin level of 1.54 mg/dL; the patient's INR level was 1.45. We performed a computed tomography angiography examination with a 16-MSCT scanner (Aquilion 16 CT system, Toshiba Medical Systems Corporation, Japan). CT angiography showed an increase in the diameter of the vena cava inferior, and hepatic congestion

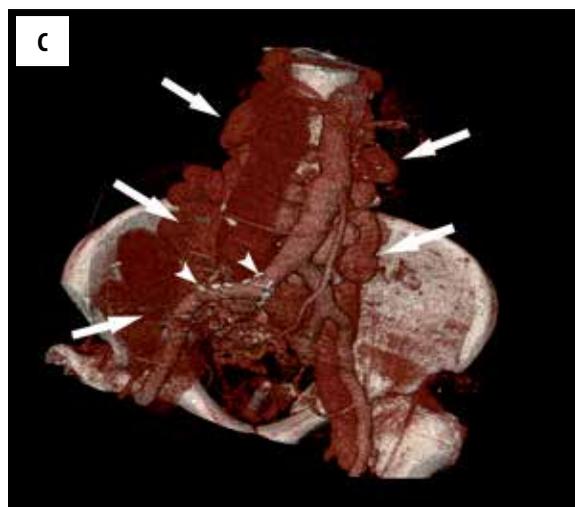


Figure 1. CT angiography demonstrated an increase in the diameter of the vena cava inferior (*curved arrows*), and hepatic congestion accompanied by hepatic veins and vascular structures, starting from the retrocrural space and continuing down to the pelvic area, that reached up to 38 mm diameter at the level of the iliac vascular structures (**A–C**) (*arrows*), and which formed a glomus along the spinal canal through the spinal cord (**A**) (*arrows*), causing bone destruction. Coils along the right internal iliac artery were also seen (*arrowheads*).

accompanied by hepatic veins and vascular structures, starting from the retrocrural space and continuing down to the pelvic area, that reached up to 38 mm diameter at the level of the iliac vascular structures, and which formed a glomus along the spinal canal through the spinal cord, causing bone destruction (Figure 1A–1C). Diplegia and neurogenic bladder were thought to be caused by the cauda equina and root compressions of the pelvic AVM. The patient was treated in our hospital for cardiac failure and for cardiac hepatic failure that developed secondary to cardiac failure.

Discussion

AVMs can be acquired or congenital. Congenital AVMs develop as a result of a spontaneous failure in focal vascular development between the fourth and tenth week of embryonic life [4]. While many AVMs are sporadic, mutations such as ENG and ALK-1 in hereditary hemorrhagic telangiectasia, RASA-1 in familial capillary malformation-AVM, or PTEN in Bannayan-Riley-Ruvalcaba or Cowden syndrome, have also been described [5]. In general, pelvic AVMs are more often acquired lesions and their etiologic factors include trauma, tumors, and surgery [2,3]. Congenital pelvic AVMs develop slowly and they become symptomatic late in their growth period due to infiltration and destruction of the neighboring tissues [1].

Almost 20% of the patients with pelvic AVM are asymptomatic [6]. Symptomatic patients have different presentations. The literature has reported rare cases in which patients felt pelvic uneasiness or pelvic pain caused by local effects; rectal pain and tenesmus; genitourinary complaints, such as hematuria, hydronephrosis, hemospermia, impotence, and orchitis; and sciatic nerve pressure-induced sciatic neuralgia. [2,3,6–8]. Very large AVMs result in cardiac failure caused by hemodynamic changes [3]. The symptoms started in our patient when he was in his 20s. At that time, diplegia and neurogenic bladder caused by spinal invasion and cardiac failure caused by hemodynamic changes developed. Our patient had the largest pelvic AVM in the literature and there are no pelvic AVM cases in the literature that caused diplegia and neurogenic bladder.

Imaging plays a very important role in the diagnosis of pelvic AVM. Digital subtraction angiography (DSA) is considered to be the gold standard in the diagnosis and treatment of AVM [2,6]. Gray-scale imaging with ultrasonography and color Doppler imaging are useful for the first examination, showing and following the vascular component of the lesion. In gray-scale imaging, a lesion can be seen to have a multicystic pattern or tortuous anechoic tubular structures [9,10]. Color Doppler imaging is useful in showing the lesion's vascular origin and in showing the high velocity – low resistance flow in vessels that feed and drain AVM [9,11]. CT angiography shows tortuous enlarged vascular structures typically associated with pelvic veins in the pelvic area [7,9]. MRI is the preferred method for differentiating slow-flow malformations (venous) from high-flow malformations (arteriovenous) [5].

Follow-up is recommended for patients with asymptomatic pelvic AVM, rather than treatment [6]. Surgical treatment is preferred in small lesions that have not spread to the neighboring tissues. Endovascular treatment, in conjunction with surgery or on its own, in conjunction with embolic and sclerosing agents, is the preferred therapeutic option when factors, such as intraoperative hemorrhage risk, insufficient resection of AVM, and post-surgery relapse, are considered [2,5,6]. However, relapses after embolization caused by rechannelization and neovascularization are also frequent. Embolization can be repeated in case of a relapse [2,6]. Multiple agents, such as coils, sclerosants (e.g. alcohol), detachable silicon balloons, ethylene-vinyl alcohol co-polymers (Onyx) (Covidien, Plymouth, MN, USA), rapidly polymerizing acrylic adhesives (e.g., n-butyl cyanoacrylate or isobutyl cyanoacrylate), and polyvinyl alcohol foam particles are used in the treatment of AVM [5]. Access to AVM or complications caused by endovascular treatment varies based on the location of the lesion. Transarterial, transvenous, direct percutaneous, or combined approaches can be preferred. Embolo/sclerotherapy, which uses a combination of coil and ethanol, has been found to be effective in

providing complete remission. In order to prolong the period of contact between the sclerosing agent and the vascular malformation endothelium, lower extremity tourniquets or an intravascular occlusion balloon can be used; embolization of the vein that drains AVM can also be used. In complex patients, multidisciplinary surgery and endovascular approach are generally the best options [5].

Conclusions

Pelvic AVMs are uncommon lesions and they can present with various symptoms based on their locations and sizes. Delays in diagnosis and treatment can cause local and systemic complications. Imaging is very important in the diagnosis of pelvic AVM, in assessing its spread and its association with neighboring anatomical structures, and in planning treatment and follow-up for patients.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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