A RARE CASE OF TREATMENT OF GIANT PARAVERTEBRAL ARTERIOVENOUS MALFORMATION COMBINED WITH IDIOPATHIC SCOLIOSIS

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The paper describes a case of successful treatment of a 32-year-old female patient with idiopathic scoliosis combined with giant paravertebral arteriovenous malformation in the lower thoracic region, causing the T11 vertebral body destruction, circular epidural compression, and myelopathy. The treatment was performed using 3D printing technology and hybrid surgery. The follow-up period after the first surgery was 26 months. The case report is accompanied by a literature review that analyzes world experience in the treatment of spinal arteriovenous malformations.

Key Words: arteriovenous malformation, scoliosis, 3D model, 3D printing, hybrid surgery.

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Spinal arteriovenous malformation (AVM) is usually a relatively rare pathology. The most common malformations are spinal dural arteriovenous fistulas; AVMs with the intramedullary nidus are less common. The diagnosis, management, and prognosis for them are formulated quite clearly. Paravertebral AVMs are the rarest and least studied lesions. Accumulated experience in the treatment of these malformations is represented by reports of individual cases or small series. Paravertebral AVMs, being often asymptomatic and incidentally diagnosed, can cause serious neurological disorders. The treatment of these patients is a challenge for the surgeon due to the management uncertainty, variable angioarchitectonics of the lesion, risks of massive blood loss, neurological deficit worsening (ischemia and venous infarction), and other factors [1–3].

A 32-year-old female patient with idiopathic scoliosis in the thoracic and lumbar spine felt sensory disturbances and progressive weakness in her lower extremities for 6 months before admission to the hospital. She had a history of scoliosis since childhood. On primary examination, CT revealed scoliosis in the thoracic and lumbar regions with destruction of the T11 vertebral body. At admission to the hospital, a neurological examination revealed signs of lower paraparesis of up to 2–3 points in various muscle groups, hypesthesia in the lower extremities and perineum (ASIA class C), pain in the thoracic and lumbar spine (VAS score of 6), and urinary retention. MRI showed an epidural space-occupying lesion that circularly compressed the spinal cord with signs of myelopathy (Fig. 1). The situation was considered as a possible tumor process. The patient underwent a percutaneous biopsy; after removal of a mandrin, high pressure arterial bleeding developed.

At the next step, CT angiography of the thoracic and lumbar spine was performed, which revealed a giant paravertebral AVM with multiple afferent vessels, up to 1.5 cm in diameter, extending from the thoracic aorta, and efferent vessels, the largest of which drained into the *vv. cava inferior, azygos*, and *hemiazygos*. The vasculature was filled during the arterial phase. The malformation was partially located in the T11 vertebral body with its destruction and drainage through the epidural veins; multiple vessels perforating posterior parts of the T11 and T12 vertebrae were also identified (Fig. 2).

Superselective angiography provided analogous information. In addition, enhancement of the anterior spinal artery through several afferents (Adamkiewicz artery) was detected. For this reason and due to a large number of high-flow vessels, total intravascular occlusion was impossible. Embolization with hemostatic sponge was performed; microcoils were placed into several large branches (Fig. 3).

Given potential progression of the neurological deficit to the lower paraplegia level, there was a decision to perform open surgery with fixation of the spine, possible partial correction of scoliosis, and an attempt to isolate and exclude AVM afferents at the compression level. A 3D model of the spine with vessels was prepared for better preoperative planning and intraoperative visualization (Fig. 4).

Open surgery was performed 4 days after angiography with embolization. During standard posterior median approach to the spine, multiple AVM vessels perforating the posterior structures, mainly on the left, were transected, which was accompanied by intense bleeding from muscles and bones. Hemostasis was achieved using mono- and bipolar coagulation (Surgicell and Sur-

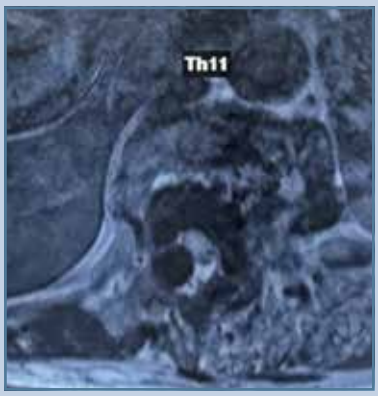

Fig. 1
MRI scan of T11 vertebra destruction with an epidural space-occupying lesion and a hyperechoic T2 signal from the spinal cord in the 32-year-old female patient

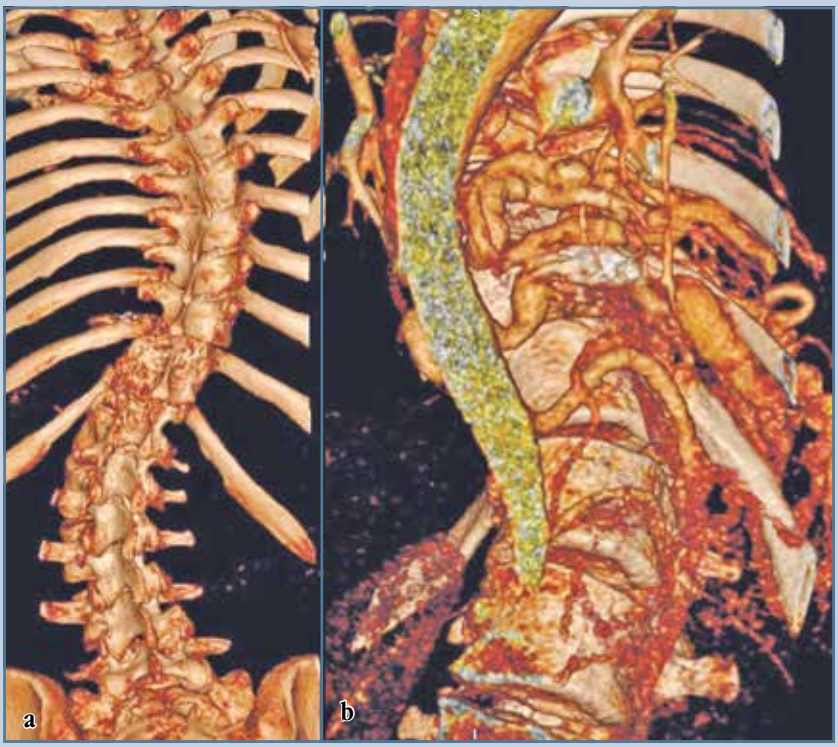

Fig. 2
CT angiography of the 32-year-old female patient: **a** – 3D reconstruction of the thoracic and lumbar spine; **b** – anterior portion of the malformation (multiple afferent vessels arising from the aorta and efferent vessels coming from the spinal canal and draining into vv. azygos and hemiazygos are seen)

giFlo) and tamponade of bone perforation areas with a large amount of wax. An estimated volume of blood loss at the approach stage amounted to 2,000 mL.

At the next step, transpedicular T5–T12–L1–L4 fixation (monolateral stabilization at the T10–T12 level on the right) was performed. Then, a posterolateral extrapleural approach to the T11 body with isolation and ligation of afferent vessels and subsequent laminectomy were planned. However, during right-sided costotransversectomy at the T10–T11 level and an attempt to open the spinal canal, the surgical team faced permanent intensive bleeding with failure to visualize individual vessels. So, further decompression was refused. After achieving hemostasis, scoliosis was partially corrected without osteotomy. An approximate volume of blood loss was 9,000 ml; replenishment was achieved by reinfusion of the patient's blood and donor components. The surgery time was 6 h 35 min.

On day 7, the patient developed a complication – exudative pleurisy, which required placement of a pleural drain. The patient was discharged on the 24th day. Control CT angiography revealed a decrease in the paravertebral AVM component, which was achieved, in our opinion, due to transection and coagulation of multiple vessels during surgical access. Filling of the epidural veins at the myelopathic lesion level was also reduced, which was probably due to remodeling of blood flow in the AVM. There was neurological regression (ASIA class D) with recovery of the pelvic functions.

After a 3-month improvement period, the patient again felt progressive symptoms of a spinal cord lesion with paresis of up to 2–3 points, sensory disturbances (ASIA class C), and urinary retention. CT angiography showed recurrent epidural compression. Re-surgery with correction of scoliosis, coagulation of paravertebral vessels, and possible laminectomy was decided. The second operation was performed in a hybrid operating room, which enabled partial AVM embolization with hemostatic sponge immediately before the open stage. During approach,

Fig. 3
Spinal angiogram demonstrating filling of several afferent vessels and enhancing of the anterior spinal artery (indicated by the arrow)

more attention was paid to excluding a posterior paravertebral portion of the malformation by means of more extensive lateral dissection and coagulation of large afferents. After removal of rods, osteotomy (Schwab type 2) was performed at the T4–T9 level. Opening of the spinal canal at the T8 level resulted in intense arterial epidural bleeding, which was stopped by wax tamponade. A similar situation occurred when T11 laminectomy was undertaken. Correction of scoliosis was achieved by segmental derotation; conditions for spinal fusion were created by decortication of the posterior structures with capping using autologous bone and osteoconductor.

On day 9, the patient developed signs of *S. aureus* wound infection, which required wound debridement and antibiotic therapy. The patient was discharged with a neurological deficit at the preoperative level on the 21st day after surgery.

In the postoperative period, the patient gradually recovered movements and sensation in the lower extremities, with complete regression being achieved 4 months after surgery (ASIA class E). The patient underwent frontal and lateral radiography before and after surgical treatment to assess the degree of deformity correction (Fig. 5). Table 1 shows parameters of the frontal and sagittal balances before and after surgery.

Control CT angiography at 7 months revealed filling of the residual malformation with drainage through epidural veins, with smaller AVM filling at the T11 and T12 levels (Fig. 6).

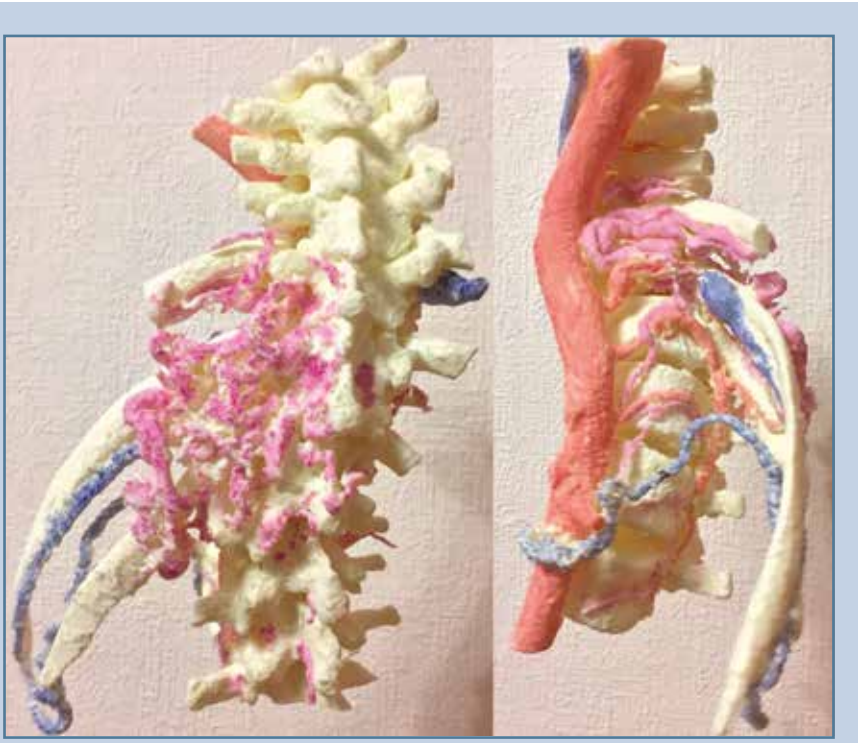

Fig. 4
Personalized 3D model with the largest vessels of a arteriovenous malformation

Discussion

Symptomatic paravertebral AVMs are extremely rare pathological processes described in the literature as individual cases or small series (Table 2). The causes for myelopathy development include compression by epidural veins, epidural hematoma, and venous hypertension due to dural arteriovenous fistulas with spinal perimedullary venous drainage. Paravertebral AVMs can be represented by single or multiple direct dural arteriovenous shunts or have a nidus. In most of the known spinal AVM classifications, this type of malformation is not mentioned [5–9]. Among vascular malformations of the spinal cord, Bao and Ling [10] classified not only intramedullary AVMs and intradural and dural arteriovenous fistulas but also paravertebral AVMs and Cobb syndrome.

Gandhoke et al. [14] reported a case of venous malformation located epidurally in the thoracic region and mediastinum. The malformation was accompanied by the formation of a syrinx; the symptoms were represented by progressive tetraparesis, paresthesia, and frequent headache attacks. At the first stage, the patient underwent osteoplastic C6–T5 laminectomy with resection of an epidural portion of the AVM (blood loss – 2,000 mL); the second stage, 2 months later, included percutaneous sclerotherapy of a mediastinal fragment.

The largest series of 10 cases was reported by Goyal et al. [15]. Nine patients had different neurological disorders. In three cases, a hyperintense T2-weighted signal was observed; of these, there was drainage into the perimedullary veins in two cases and compression by the epidural veins in one case. Three patients underwent microsurgery; seven patients underwent transarterial or transvenous embolization. Neurological symptoms regressed in all cases.

Paravertebral AVMs are not prone to rupture, in contrast to intramedullary AVMs that are characterized by a rupture rate of 54% [8]. The only rupture case reported in the literature was that described by Kitagawa el al. [16] in a 12-year-old child with acute symptoms of thoracic spinal cord compression caused by an epidural hematoma. Laminectomy with evacuation of the hematoma provided partial regression of symptoms. A month later, the patient underwent superselective angiography that revealed a paravertebral AVM located in the T2–T3 intervertebral foramen. After endovascular treatment (Onyx embolization), the patient developed a neurological deficit,

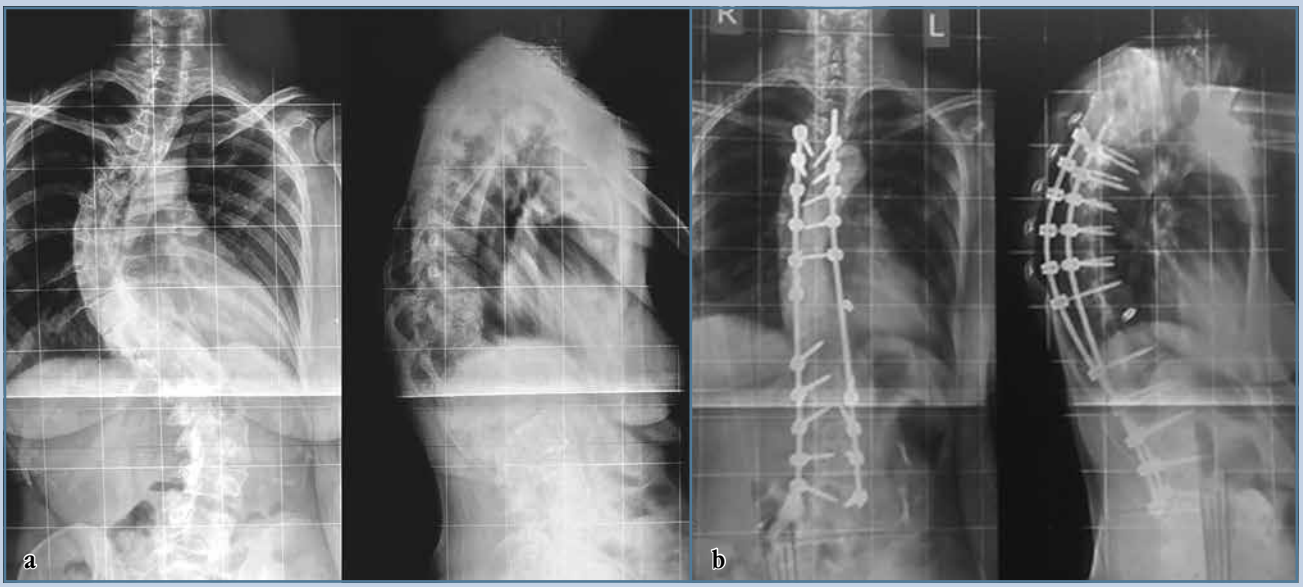

Fig. 5
Frontal and lateral radiography before **(a)** and after **(b)** surgical treatment

presumably due to spinal cord edema caused by increased venous hypertension. The same authors reviewed 16 previously described cases of paravertebral AVMs in children [16].

One of the rarest variants of paravertebral AVMs is malformation with an intraosseous component. In 1997, a group of Italian authors [17] reported a case of an intraosseous spinal AVM in a combination with juvenile scoliosis in a child. The authors suggested that it was the malformation that caused the development of scoliosis. There are no other cases reporting combination of a paravertebral AVM and idiopathic scoliosis.

In 2014, Louis Jr. el al. [18] reported another case of an intraosseous spinal AVM with epidural compression of the spinal cord and myelopathy, indicating that this case, according to the literature, was the second.

Farhat el al. [19] described 2 cases of a paravertebral AVM in children. The clinical presentation in these cases included only systolo-diastolic murmurs during auscultation in the paravertebral area in one child and in the 2nd intercostal space in the other. Given the lack of cardiovascular and neurological symptoms, delayed surgery was decided. The authors also indicated that, according to the literature, the location of malformation anterior to the vertebral body may cause orthopedic disorders in the future. In addition, the authors indicated the possibility of compression of intracanal nerve structures by dilated epidural veins [19].

The approach to treatment of paravertebral AVMs is not defined due to a rare occurrence and variability of the pathological process. Unlike cerebral AVMs where the main goal is complete exclusion of the malformation to prevent its rupture, the main task in the case of spinal AVMs is to stop progression of myelopathy, which is achieved by reducing the filling of epidural veins in combination with uncoupling of dural arteriovenous fistulas in the presence of drainage into the perimedullary veins [1, 20].

According to some reports, microsurgical treatment of isolated dural arteriovenous fistulas is more effective than

Table 1
Parameters of the sagittal and frontal balances of the patient before and after correction of spinal deformity

Parameter	Before correction	After correction
Pelvic tilt, deg.	43.3	43.3
Pelvic inclination, deg.	9.8	7.2
Sacral slope, deg.	33.5	36.1
Lumbar lordosis, deg.	60.5	44.5
Difference between pelvic tilt and lumbar lordosis, deg.	-17.2	-5.0
Thoracic kyphosis, deg.	53.0	47.2
Cervical lordosis, deg.	6.0	8.2
Sagittal vertical axis C7, mm	20.7	11.5
Cobb angle, deg.	43.2	17.6

intravascular treatment [2, 3, 21]. However, in most of the reported cases, paravertebral AVMs were treated endovascularly through a transarterial or transvenous approach. It is worth noting that one or several dominant afferents were identified in the presented cases, whose exclusion from circulation led to a positive clinical outcome [4].

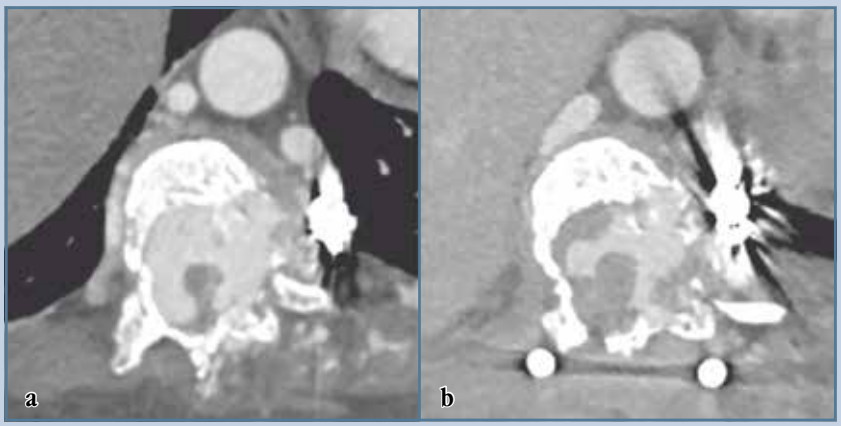

Fig. 6
CT angiography of the 32-year-old female patient before the 1st surgery **(a)** and 7 months after the 2nd surgery **(b)**

In our case, the AVM was represented by a plexus of high-flow vessels with afferents extending directly from the thoracic aorta and penetrating the spinal canal from all sides. In this situation, isolation of dominant vessels as objects for embolization or microsurgery was not possible. Manifestation of the clinical picture at 32 years of age probably occurred because of vertebral body destruction caused by perforating high-flow vessels and circular compression of the spinal cord. We suggest that the positive clinical outcome in this case was achieved due to transection of the afferent vessels penetrating the spinal canal through the posterior structures, coagulation of vessels in the paravertebral muscles on the left, and, possibly, correction of the deformity. Therefore, AVM remodeling was performed, which was sufficient to

Table 2
Analysis of publications on treatment of paravertebral arteriovenous malformations (AVMs)

Publication	Number of cases, gender	Age	Localization	Symptoms	Treatment	Outcome
Cognard et al. [11]	1, f	17 years	L4–L5, L5–S1	Lower back pain	Balloon occlusion	Complete obliteration, regression of back pain
Fotso et al. [12]	1, m	3 years	T11	Murmur in the paravertebral area	Microspiral occlusion	Complete obliteration without deficit worsening
Hui et al. [13]	1, f	10 years	C6–C7	Paraparesis	Microcoils + NBCA	Complete obliteration, regression of symptoms
	1, m	9 months	T1–T2	Heart failure	Glue	
	1, f	7 months	C3–C7	Dysphagia	NBCA	
Goyal et al. [15]	7, m 3, f	1–73 years	–	–	Microsurgery – 3 Embolization – 7	Worsening i n 1 case
Kitagawa et al. [16]	1, f	12 years	T1–T3	Paraplegia	Emergency laminectomy with hematoma removal, followed by Onyx embolization	Improvement after decompression, worsening after embolization
Molina et al. [17]	1, m	8 years	L4 with an intraosseous component and scoliosis	Back pain	En bloc removal	Improvement (pain regression)
Louis Jr. et al. [18]	1, f	59 years	T5 AVM + an intraosseous component	Back pain	Combined (embolization + resection)	Improvement
Gandhoke et al. [14]	1, f	18 years	C6–T5, venous malformation	Tetraparesis, migraine	Combined (resection + sclerotherapy)	Improvement

reduce filling of the epidural veins and their compression effect on the spinal cord with regression of myelopathy and complete recovery of lost neurological functions. The initially proposed management for decompression of the spinal cord via isolation and exclusion of afferent vessels through the posterolateral extrapleural approach and laminectomy turned out to be inapplicable due to permanent intense arterial bleeding.

In our opinion, the use of a 3D model of the spine with a reconstructed AVM was an additional useful tool of intraoperative imaging, which is especially important in cases of atypical anatomy. Visualization of the AVM vessels with reference to the bone structures enabled prediction of intense bleeding areas and preventive measures to stop bleeding. The main goal of re-surgery was deformity correction, which was not performed at the first intervention due to a large volume of blood loss, additional exclusion of AVM afferents, and an attempt to perform laminectomy. Performing this stage at a hybrid operating room allowed embolization of the AVM branches by hemostatic sponge immediately before the open stage within one anesthesia session.

Conclusion

The presented case is unique because it combines an extremely rare pathology in the form of a giant paravertebral AVM with an intraosseous component, compression myelopathy, and idiopathic scoliosis. In this case, surgical treatment options previously used for paravertebral AVMs, which included isolated endovascular or microsurgical treatment, were inapplicable. To achieve a positive clinical outcome, it was sufficient to remodel the AVM without its complete exclusion by embolization, open exclusion of the malformation vessels, and correction of scoliosis.

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The authors declare no conflict of interest.

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