Spinal Arteriovenous Epidural Fistula with Acute Paraplegia

Diagnosis and Neurointerventional Emergency Treatment

A Case Report

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Summary

We report a case of a 68-year-old woman with an acute paraplegia due to venous congestion of the spinal cord caused by an exclusive epidural arteriovenous fistula. Diagnosed by MRI and selective spinal angiography the fistula was embolized during emergency treatment via transarterial access. Immediately after the intervention the paraplegia declined and the patient recovered completely. Epidural AV fistulae are a very rare and therefore relatively unknown cause of vascular myelopathy. They may require emergency management to avoid permanent neurological deficits.

Introduction

Solely epidural fistula are a very rare cause of vascular myelopathy resulting in paraplegia. The diagnosis is even more demanding than that of a typical dural AV fistula.

Our report of a woman suffering from total paraplegia due to an epidural fistula presents the patient's complete restitution after neurointerventional treatment. Furthermore the potential etiology and pathophysiology are analyzed.

Case Report

History and Examination. A 68-year-old woman was transferred to a urologic department

with suspected kidney colic. She suffered from acute paravertebral back pain. On admission she reported some unspecific disturbances while walking which she had noticed for the last two weeks. Her history showed no diseases and no neurologic abnormality was detected at the time of admission, namely no signs or symptoms of any spinal disorders.

Hospital course. She was treated with spasmolytics and analgetic drugs. At night she suddenly developed an acute and severe paraplegia. She was unable to move either of her legs, she suffered a sensory loss of all qualities and had a bladder dysfunction.

An MRI was performed and interpreted as a spinal hemorrhage (figure 1). For spinal angiography the patient was transferred to neuroradiology. Because of doubts concerning the spinal hemorrhage, the MRI was repeated. First MRI was done with ultrafast sequences with low resolution and many susceptibility artifacts. We performed standard SE and TSE T1 pre- and post contrast media application, and T2 weighted sequences in sagittal and axial planes.

The images showed a longspread centromedullary edema at mid-thoracic level as well as some minimally enlarged veins around the conus medullaris. On the surface of the thoracic spinal cord a dural AV fistula was not seen clearly enough, but in context with the in-

Figure 1 Spinal MRI with T2 and T1 weighted ultrafast sequences misinterpreted as an intraspinal hemorrhage

Figure 2 MRI T1 weighted pre- and post contrast and T2 Image with centromedullary edema. Only slightly enlarged veins in the contrast enhanced T1 image.

tramedullary changes its suspicion was raised. No hemorrhage was seen (figure 2).

Spinal intra-arterial angiography was performed as emergency treatment. The segmental spinal arteries of both sides beginning at Th5 down to L2 were selectively documented. The main anterior spinal artery was originating at Level Th10 on the left side. The angiography of the segmental artery L2 right showed a pathological opacification of the epidural venous space with a very late reflux in an ascending dorsal medullary vein. One small feeding artery arising from the lumbar artery L1, distal from the origin of the radiculo-meningeal artery was identified (figure 3).

Consequently a spinal epidural arteriovenous malformation with venous reflux and congestion and consecutive centromedullary edema was diagnosed.

A 0.010 inch microcatheter was navigated into the feeding artery close to the fistula point. Using a mixture of NBCA and Lipiodol, the fistula was closed by a single injection. The glue passed the fistula point to the venous side closing the fistula completely (figure 4). The control DSA showing all neighbouring segments proved total occlusion of the fistula. Next day's control MRI showed the partial disappearance of the enlarged veins, but the medullary edema could still be seen.

The patient's clinical condition improved markedly and she was able to move her legs the next day. The next but one day she regained her ability to walk and climb stairs. Three days later she went home without any remaining neurological deficit.

Discussion

Spinal epidural AV fistula can cause acute paraplegia due to reflux into the medullary veins with venous congestion and consecutive intramedullary edema. Compared to the rare dural/intradural AV fistula, the epidural fistula seems to be even more rare, but this may be a selection artifact due to the diagnostic management performed. A review of the literature revealed only very few articles on epidural AV fistula, most of them focusing on those in the thoracolumbar spine^{1-3,6}. There are different reasons for this moderate result.

First, those fistulas can occur without any pathologically enlarged pial veins. Only showing the centromedullary edema, can they easily

be interpreted wrongly as myelitis or ischemic infarction and will therefore miss consecutive diagnostic by spinal DSA.

Secondly, angiographic proof can be difficult, because the epidural venous pouch and the arising medullary vein are opacified quite late due to a slow flow and might go unnoticed for this reason. That is why in our opinion every patient with an obscure centromedullary edema and paraplegia should undergo selective spinal angiography.

The etiology of this kind of fistula is unheard of. Dural fistulae are well known causes of chronic progressive paraparesis. In a few cases, the development progresses rapidly in days or hours. The typical dural AV fistula is fed by radiculomeningeal feeders and drains into medullary veins^{7,8}. In contrast, epidural fistulae are fed by osseous branches of the segmental arteries without an anatomical relationship to the meningeal arteries³.

Normally, the epidural venous plexus is a communicating system with septations and pouches, but it still has multiple outflows at nearly every segmental level. Thus, a reflux into medullary veins requires a special patho-anatomical condition as a blockade of the outflow at a circumscribed level. We hypothesize that a venous thrombosis of parts of the epidural venous plexus with incomplete recanalisation and some shunts due to an insufficient collateralisation may lead to an increase in venous pressure and might cause a reflux into the intradural medullary venous system because of the occluded outflow. The patho-physiological mechanisms could to some extent be compared to those of the intracranial dural fistulas arising after cerebral sinus thrombosis^{4,5}. It is hypothesized that recanalisation processes in thrombosed sinuses induce an angiogenesis in the dural wall as well as the opening of preexisting dural shunts inducing the growth of the fistula⁵. The same mechanism may account for the typical spinal dural fistula. The epidural fistula, however, seems to be the result of a thrombosis-recanalisation process, but in contrast to this, it is away from intradural angiogenic changes.

With regard to this supposition the patient's history with some foregoing symptoms could be interpreted as a manifestation of epidural thrombosis. Such a thrombosis may induce a disruption of the barrier which normally inhibits retrograde flow from the epidural plexus

Figure 3 A,B: Angiography: A-P view of the lumbar artery L1 right in immediate arterial (A) and late arterial phase showing the opacification of the epidural plexus and the reflux into the ascending medullary vein (B)

to perimedullary veins. In a recently published paper valve-like mechanisms were discussed as the basis for such a barrier. These mechanisms physiologically should inhibit reflux from the

Figure 4 Spinal angiography at Level L1 right after glue embolization of the fistula with complete occlusion of the epidural shunt.

Figure 5 MRI T2 weighted confirms the improvement of the intramedullary signal.

epidural plexus to perimedullary veins. Probably other pathological circumstances may disturb the barrier leading to a reflux from epidural to perimedullary veins⁷, but a thrombosis is the most likely cause in most of the rare cases.

Dural fistulae usually occur in elderly men and are seldom detected in women⁷. Our patient was female, but a single case may not allow speculations on gender differences but could be a hint to observe the matter further.

The MRI changes can persist and have a longer latency than the clinical symptoms. Interestingly, the MR findings continued to be pathological when the patient was already free of symptoms. Another remarkable point is the presence or absence of enlarged intradural veins suspicious for an AV fistula. In dural fistulae, these varicose veins seen in MRI led to further diagnostics and detection of the fistula. In epidural fistula, the veins were less prominent and could be missed or interpreted as usual veins. Thus, even a centromedullary edema without enlarged intradural veins is suspicious for an arteriovenous fistula and should lead to a spinal angiography.

The evolution of the disease in dural AVF is chronic and deterioration occurs over years in most cases⁸. As an absolute exception, a more rapid course is seen and an emergency treatment must be performed right away. In our case, there was a desolate course with acute deterioration and progression to an acute severe paraplegia in hours, suggesting that epidural fistulae may have a dynamic different from that of dural fistulae.

Conclusions

Solely spinal epidural arteriovenous fistulae are rare malformations which differ from dural AV fistulae in many ways. The diagnosis is difficult, MRI can be misleading because varicose veins cannot be seen easily.

The course of impairment can be very rapid, so emergency management by spinal angiography and neurointerventional therapy has to be considered in such cases. Pathophysiologically, a subacute thrombosis of the epidural plexus may be responsible for the development of the lesion.

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