Klippel-Trenaunay-Weber syndrome with intracranial dural arteriovenous fistula and spinal arteriovenous shunt

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Abstract: Our clinical image revealed a case of Klippel-Trenaunay-Weber syndrome (KTWS) accompanied by a spinal arteriovenous shunt and an intracranial dural arteriovenous fistula. There have been few reports of KTWS patients developing dural arteriovenous fistulas. This clinical image highlights the strong association between KTWS and CNS vascular malformations. It is important to note that KTWS patients are at risk of developing central nervous system vascular diseases. Therefore, early detection and screening are recommended.

Keywords: Klippel-Trenaunay-Weber Syndrome; Dural Arteriovenous Fistula; Spinal Vascular Malformation; Angiography.

A 40-year-old woman presented with a headache and weakness in the right lower limb. The physical examination revealed asymmetry in lower limb length, hypertrophy, and stains in the right lower limb (Fig. 1A). Her muscle strength in the right lower limb is level 3, and level 5 in the left lower limb. She has a history of Klippel-Trenaunay-Weber syndrome (KTWS) and has had lower extremity endovascular intervention as well as right hip replacement surgery. Digital subtraction angiography revealed vascular malformations of the right lower limb (Fig. 1B), bilateral sigmoid-transverse sinus occlusion (Fig. 2A), right transverse sinus dural arteriovenous fistula (DAVF) (Fig. 2A-C), and spinal arteriovenous shunt (Fig. 2D). The DAVF was completely occluded (Fig. 2C), and conservative treatment for the spinal arteriovenous shunt was chosen. The patient's headache had improved four days after embolization, and her muscle strength was the same as before.

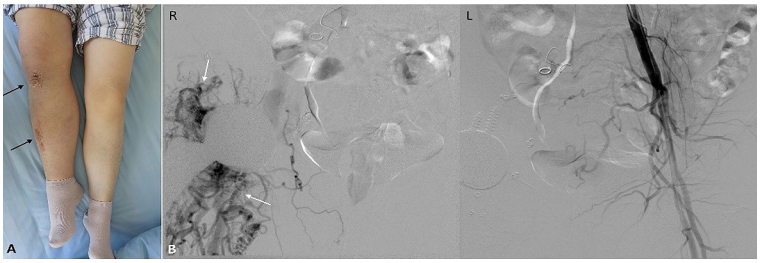

Figure 1. (A) Photograph of a 40-year-old patient with stains (black arrows) and hypertrophy in the right lower limb. Note the asymmetry in the length of the lower limbs. (B) Lower limb digital subtraction angiography revealed vascular malformations of the right lower limb (white arrows).

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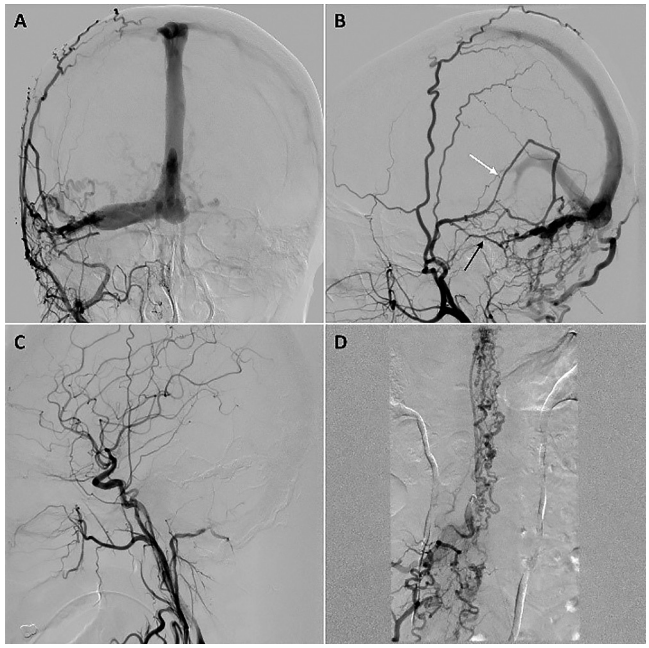

Figure 2. (A-B) The angiogram revealed a right Cognard type IIa+b transverse dural arteriovenous fistula supplied by petrosal (black arrow) and petrosquamous (white arrow) branches of the middle meningeal artery, transosseous branches of the occipital artery (orange arrow), and pial branches of the superior cerebellar artery (not shown), with superior saggital sinus and straight sinus-the vein of Galen-deep cerebral veins drainage. Note the bilateral occlusion of the sigmoid-transverse sinus. (C) The fistula was completely occluded via the superior cerebellar artery, petroquamous branch of the middle meningeal artery, and the occipital artery, using Onyx and Glubran. (D) The spinal angiogram revealed a lumbar spinal arteriovenous shunt.

Klippel-Trenaunay and Parkes Weber syndrome share similarities as complex vascular malformations. Both disorders exhibit limb enlargement, varicosities, and cutaneous capillary malformations. However, Klippel-Trenaunay syndrome is characterized by a pure low-flow condition, whereas Parkes-Weber syndrome consistently displays significant arteriovenous fistulas.[1] Some authors refer to them as the KTWS. Cavernous malformations and spinal vascular malformations are common central nervous system disorders that can occur in KTWS, although some debate exists.[2-5] However, there have been few reports of KTWS patients developing DAVF.[6]

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Informed Consent

Consent obtained from parent(s)/guardian(s).

Ethical approval

This retrospective study involved human participants and was approved by the Ethics Committee of Xuanwu Hospital, Capital Medical University [2017; 010].

Conflicts of Interest

The author declares no conflict of interest.

Data Availability

Data are available upon reasonable request.

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