A 21-year-old woman presented with left ocular pain and severe pulsatile headache. She had suffered from easy bruising since she was young. On physical examination, ecchymoses and bruises were noticed on trauma sites (Figure 1A). The skin was so thin that subcutaneous blood vessels were visible (Figure 1B). Huge hematomas and ecchymoses occurred at the puncture sites after angiography (Figure 1C). There was no significant hyperelasticity of the skin. Mild hypermobility of the joints in the hands was observed.

Angiography showed a large carotid-cavernous fistula and an aneurysm at the cervical portion of the left internal carotid artery (Figure 2A). Abdominal aortography disclosed a large ovoid aneurysm of the left renal artery with delayed nephrogram (Figure 2B). The carotid-cavernous fistula was successfully occluded by detachable balloons.

This case demonstrates characteristic clinical features of type IV Ehlers-Danlos syndrome, an autosomal dominant disorder resulting from mutations in the COL3A1 gene. The gene defects alter the metabolism of type III collagen, a major constituent of the walls of blood vessels. Type IV Ehlers-Danlos syndrome is life-threatening because of its vascular manifestations, which are characterized by rupture, dissection, or aneurysm formation affecting large or medium-sized arteries.

References
Figure 1. A, Patient’s arms showed ecchymoses and bruises at needle puncture sites. B, Patient’s skin over upper thorax showed transcluence with visible vessels (arrows). C, Severe ecchymoses and hematomas occurred at puncture sites after transfemoral abdominal aortogram despite adequate compression.

Figure 2. A, Left internal carotid angiogram disclosed a direct carotid-cavernous fistula (solid arrow) with drainage into superior ophthalmic vein (arrowhead). There is a small aneurysm at anterior wall of distal cervical internal carotid artery (open arrow). B, Abdominal aortogram showed large aneurysm (arrow) of left renal artery with delayed nephrogram.
Skin Manifestations, Multiple Aneurysms, and Carotid-Cavernous Fistula in Ehlers-Danlos Syndrome Type IV
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