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Ehlers-Danlos syndrome with a spontaneous caroticocavernous fistula occluded by detachable balloon: case report and review of literature

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Abstract We report a caroticocavernous fistula (CCF) treated by an endovascular procedure in a young woman with Ehlers-Danlos syndrome type IV, with severe bilateral carotid and vertebral artery dysplasia. The CCF, which appeared after minor trauma, was successfully occluded by a detachable balloon introduced into the venous side by an arterial approach. Six previously published cases are reviewed, five successfully treated. The difficulties and risks of the endovascular procedure due to the vascular changes, are emphasised. The possibility of the venous approach is discussed.

Key words Ehlers-Danlos syndrome · Caroticocavernous fistula · Endovascular procedure

Introduction

The diseases of connective tissue include various congenital or acquired disorders resulting from abnormal synthesis, organisation or degradation of connective frame tissue. The Ehlers-Danlos syndrome (EDS), first described in 1688 by Van Meek'ren [1], includes various hereditary changes in collagen, with different molecular determinants, of dominant or recessive genetic transmission. Ten types are currently described, the most frequent being types I–IV, of which cutaneous hyperelasticity and vascular fragility are the basic clinical signs. The risk of vascular rupture is not the same for all variants of EDS, but characterise type IV. In this type, an abnormality of type III collagen is responsible for haematological disorders and severe blood vessel changes such as rupture, dissection and aneurysm, affecting large or medium-size peripheral and cervical arteries, caroticocavernous fistula (CCF) and intracranial aneurysms. These complications can be treated, by coils for saccular aneurysms or detachable balloons for CCFs, with increasing safety of approach and a low postoperative risk of bleeding at the point of puncture. We report a patient with EDS type IV who developed a CCF, successfully treated by endovascular procedure.

Case report

In 1994, following minor cranial trauma, a 40-year-old woman complained of reduced visual acuity in the right eye and a pulsatile intracranial bruit. A past family history of cardiovascular disease was discovered; her elder brother had a history of myocardial infarct and died aged 40 years after spontaneous dissection of a subclavian artery aneurysm. The patient noticed thinness of the skin, with a tendency to subcutaneous bruising after small injuries. Neurological examination was normal except for dilatation of the right retinal vessels and a marked right temporal murmur. CT before and after intravenous contrast medium was interpreted as normal. Angiography (Fig. 1) showed dysplasia of both vertebral and internal carotid arteries, extending down to C5. There was also a direct high-flow CCF on the right draining into the sphenoparietal petrosal vein, the inferior petrosal sinus and the contralateral cavernous sinus via the coronary sinus (Fig. 2). Occlusion of the fistula with preservation of carotid artery flow was obtained via the femoral route using a detachable balloon attached to a 1.8 F catheter introduced through an 8 F coaxial catheter. The anatomical result at the end of the procedure was good (Fig. 3) and the murmur had disappeared. A paralysis of the right abducens nerve regressed 6 months later. On the basis of the family and personal history and the radiological findings, the diagnosis of an arterial form of type IV EDS was made.

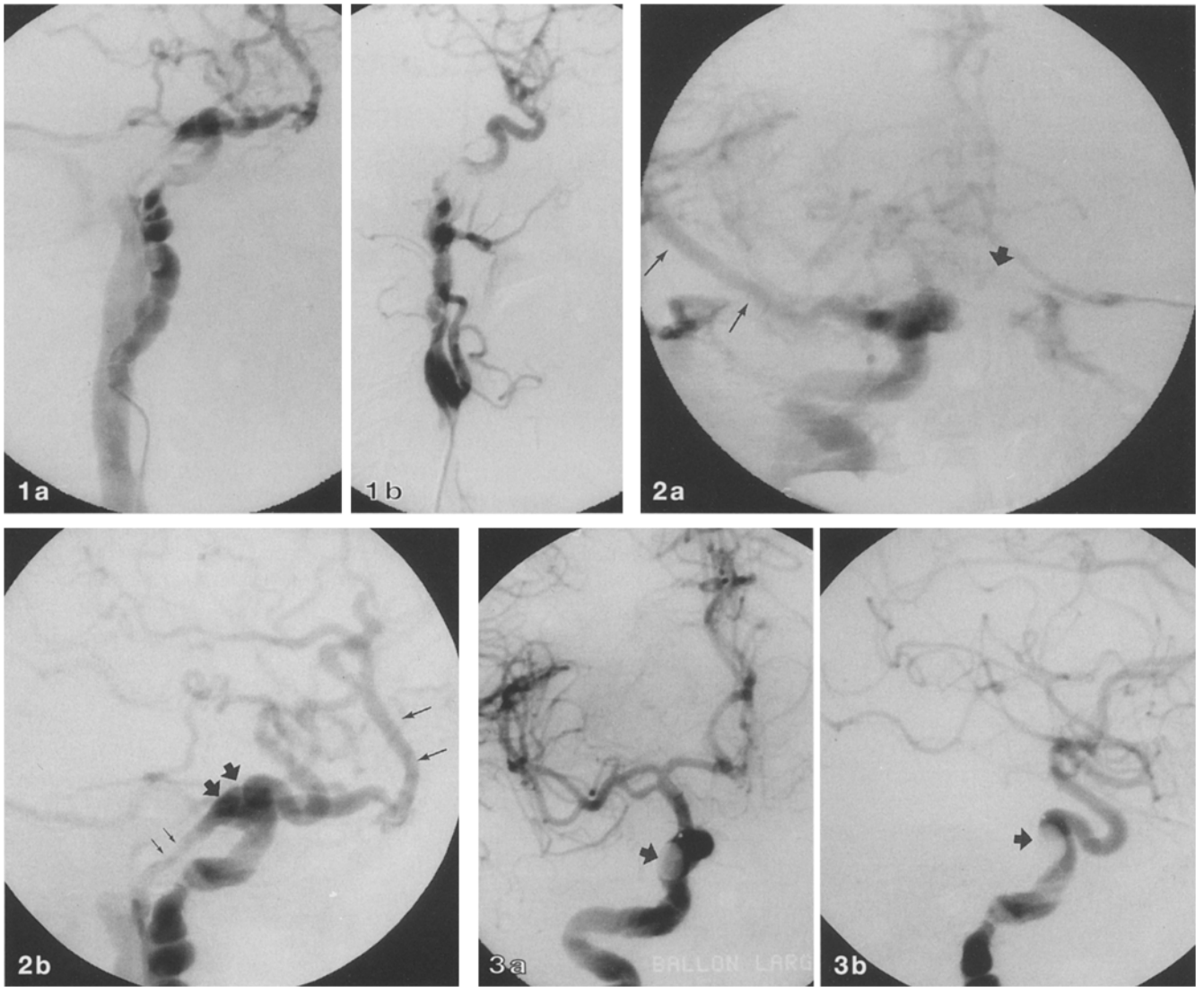

Fig. 1 Preoperative (**a**) right (**b**) left lateral carotid arteriograms demonstrating ectasia and tortuosity of the extracranial internal carotid arteries

Fig. 2 Right internal carotid angiogram (**a** anteroposterior, **b** lateral projections) demonstrates a right caroticocavernous fistula (*double broad arrows,* **b**) to the inferior petrosal sinus *(small arrows),* sphenoparietal veins *(medium arrows)* and contralateral cavernous sinus (*broad arrow,* **a**)

Fig. 3 Right internal carotid angiogram (**a** anteroposterior, **b** lateral views), after detachement of the balloon *(arrow)* in the cavernous sinus, demonstrates complete closure of the fistula with preservation of the internal carotid artery

Discussion

The (EDS) is a hereditary dysplasia of connective tissue due to abnormalities of collagen. Clinically, genetically and biochemically the syndrome is heterogeneous [2].

Clinical manifestations of type IV (increased skin elasticity, bruising and vascular ruptures), correspond to the arterial form of the disease described by Barabas [3]. They are determined by a major decrease of type III collagen, one of the three major fibrillar collagens, a component of dermal papillae, intestine and approximately 40 % of blood vessel walls [4]. This decrease could be the result of, for instance, defective synthesis of collagen or overly-rapid degradation [5–7]. In the arterial wall, the lack of collagen affects, in decreasing order, the internal elastic lamina, the media and the adventitia. Disturbances of metabolism of type III collagen during the early stages of haemostasis should also explain the frequent bleeding and bruising [8].

Genetically, transmission can be recessive or dominant, this last form being the most severe. A CCF in the EDS was first described in 1955 by Francois et al. [9]. Wouter et al. [4] noted (pp 993–994) that only sixteen

cases had been reported by 1991. Unlike our case, these CCF were spontaneous. It has been suggested that these CCF are secondary to rupture of a saccular aneurysm in the cavernous sinus or spontaneous dissection of the intracranial internal carotid artery [3, 10]. However, spontaneous rupture due to the changes in the arterial and venous walls cannot be ruled out, and is suggested by the presence in three patients, at the time of diagnosis, of bilateral fistulae [3, 11].

Given the weakness of the walls of the cervical internal carotid artery and the risk of traumatic vascular rupture, the fistula can be reached by the venous access, as described by Farley et al. [12]. The fistula can also be reached via the inferior petrosal sinus, with uncertain success, given the risk of fatal haemorrhage secondary to venous rupture or possible septation of the sinus preventing correct placement of the balloon. The superior ophthalmic vein represents another possible approach but requires as surgical access [11], and is contraindicated in fistulae with posterior drainage or those less than 3 months old, where the vein is not yet arterialised.

The arterial approach is well documented [13–15]. Despite the large gauge (8 F) of the catheter used to insert detachable balloons, staff used to supraselective catheterisation are ready for possible complications and can modify the initial interventional protocol, particularly when they use the femoral approach. Because of the vascular fragility this approach must be recommended as the only adequate access since the distance between the site of the fistula and the point of arterial puncture increases the safety. Despite the risk of rupture during the procedure, patients with high-flow CCF must be treated, since these fistulae are unlikely to resolve spontaneously. These patients can have ophthalmoligical complications and have a high risk of spontaneous rupture or of the development of a complex fistula which will become difficult to treat. The risk of rupture during the procedure and the technical difficulties explain that, of the 16 patients treated by venous or arterial endovascular access, only 4 had their fistula definitively occluded. At 5-year follow-up, only 3 are still alive [16]; 10 (60 %) died: 4 after a diagnostic or therapeutic angiographic procedure, 5 after a spontaneous vascular rupture 2 weeks to 6 months after diagnosis, and 1 after an intestinal perforation 7 months after successful treatment of his CCF.

References

1. Van Meek'ren J (1988) On a special elasticity of the skin. Commelijn, Amsterdam, pp 170–172
2. Beighton P, Paepe A de, Danks D (1988) International nosology of heritable disorders of connective tissue. Am J Med Genet 29: 594–598
3. Barabas AP (1972) Vascular complications in Ehlers-Danlos syndrome (with special reference to the "arterial type" or Sack's syndrome). J Cardiovasc Surg 12: 160–167
4. Wouter I, Schevink WI, Piepgras DG, Earnest F, Gordon H (1991) Spontaneous carotid-cavernous fistulae in Ehlers-Danlos syndrome type IV. J Neurosurg 74: 991–998
5. André JM (1973) Les dysplasies vasculaires systématisées. Expansion Scientifique Française, Paris, pp 292–307
6. Beylot C, Bioulac P, Doutre MS (1983) Les manifestations artérielles du syndrome d'Ehlers-Danlos. Ann Med Int 134: 451–457
7. Julien J, De Boucaud D (1971) Fistule carotido-caverneuse spontanée et maladie d'Ehlers-Danlos. Presse Méd 79: 1241–1242
8. Lapière CM (1978) Les maladies du tissu conjonctif à expression cutanée. Ann Dermatol Vénérol 105: 905–911
9. François PW, Woilliez M, Maillet WP (1955) Maladie d'Ehlers-Danlos avec anévrysme artério-veineux intra-crânien. Bull Soc Ophtalmol Fr 5: 392–395
10. Fox R, Pope FM, Narcisi P, Nicholls AC, Kendall BE, Houridan MD, Compston DA (1988) Spontaneous carotid-cavernous fistula in Ehlers-Danlos syndrome. J Neurol Neurosurg Psychiatry 51: 984–986
11. Halbach V, Higashida R, Dowd CF, Barnwell SL, Hieshima G (1990) Treatment of carotid-cavernous fistulas associated with Ehlers-Danlos syndrome. Neurosurgery 26: 1024–1027
12. Farley MK, Clark RD, Fallor MK, et al (1983) Spontaneous carotid-cavernous fistula and the Ehlers-Danlos syndromes. Ophthalmology 90: 1337–1342
13. Debrun GM, Lacour P, Viñuela F, Fox AJ, Drake CG, Caron JP (1981) Treatment of 54 traumatic carotid cavernous fistulas. J Neurosurg 55: 678–692
14. Debrun GM, Viñuela F, Fox AJ, Davis KR, Ahn HS (1988) Indications for treatment and classification of 132 carotid-cavernous fistulas. Neurosurgery 22: 285–289
15. Pierot L, Moret J, Boulin A, Castaings L (1992) Traitement endovasculaire par voie artérielle des fistules carotido-caverneuses complexes post-traumatiques. J Neuroradiol 19: 79–87
16. Halbach V, Higashida R, Hieshima G, Hardin C, Yang P (1988) Transvenous embolization of direct carotid-cavernous fistulas. AJNR 9: 741–747