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Case Report

Giant carotid pseudoaneurysm amenable to pipeline stenting in a patient with Ehlers–Danlos type IV

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Abstract

Background: Ehlers–Danlos type IV primarily affects collagen synthesis in the vasculature, increasing the risk of these patients to have dissection and pseudoaneurysm formation. Due to friable vessels, antiplatelet or anticoagulation has been the treatment of choice. However, newer intravascular surgical devices may be promising for future management.

Case Description: A 24-year-old man with a history of Ehlers–Danlos type IV with multiple vascular and bleeding complications presented after recurrent, unprovoked presyncopal episodes. Patient was found to have dissection of bilateral internal carotid arteries (ICA) and right vertebral artery. Left ICA pseudoaneurysm was found in the proximal cervical segment. Patient was stabilized as an inpatient and discharged with outpatient follow-up with neurointerventional surgery. Follow-up imaging showed growth of the left ICA aneurysm. Patient elected to have pipeline stenting of the left ICA pseudoaneurysm. The procedure was performed without complication. Patient was discharged on dual antiplatelet therapy. At 7-month follow-up appointment, patient noted no neurological deficits. Follow-up digital subtraction angiogram at 7 months documented near-complete resolution of the pseudoaneurysm secondary to pipeline stenting.

Conclusion: Pipeline stent implantation may be a viable corrective surgical option for patients with connective tissue disorders (specifically Ehlers–Danlos type IV) who present with pseudoaneurysm formation.

Key Words: Aneurysm, Ehlers–Danlos type IV, flow diverter, neck, stent

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INTRODUCTION

Ehlers–Danlos syndrome (EDS) is a rare hereditary disorder resulting from defective collagen synthesis. There are several subtypes of this disorder – type IV being most relevant to this case. EDS type IV results in inflicted individuals being unable to synthesize type III collagen, which is utilized in blood vessel formation.[4] As a result, these individuals have vessel-wall fragility.

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Coupling this with high pressure blood flow can lead to complications of dissection, rupture, and aneurysm/pseudoaneurysm.[3] Conservative measures are generally recommended for dissection and resultant stenosis which is mild.[3] Pseudoaneurysm formation confers lower likelihood of healing with conservative measures. Rapid increases in size confer an increased likelihood of rupture, which may necessitate the need for surgical intervention.[1] Aggressive surgical approaches (conventional angiogram, endovascular intervention, vascular surgery) may be undertaken for the aforementioned complications, but may confer a higher morbidity and mortality risk perioperatively and thus should be used with caution.[2,3] If well tolerated, patients can generally have positive outcomes with minimal stroke risk and good resolution of the pseudoaneurysm.[1] Due to the rarity of the genetic disorder and pseudoaneurysm concomitantly, there have been a paucity of reports examining the effects of aggressive surgical techniques in this population. With this in mind, our case illustrates the success, which can be achieved with a more aggressive surgical technique in these rare, high-risk patients.

CASE PRESENTATION

Background

A 24-year-old man with a history of presyncopal episodes for 3 years, Ehlers–Danlos type IV, aortic dissection, gluteal artery pseudoaneurysm, possible platelet dysfunction with multiple spontaneous bleeding episodes, and chronic daily headache on sumatriptan presented after recurrent, unprovoked presyncopal episodes followed by chest tightness and headache. Given the patient's significant vascular and headache history, carotid and vertebral dissections were entertained as a possible etiology. Patient was stabilized, admitted, and advanced imaging was performed.

Investigations

Magnetic resonance imaging (MRI) of the brain and angiography (MRA) of the brain and neck were performed. MRI and MRA brain were unremarkable. MRA neck showed a right internal carotid artery (ICA) dissection with 50% stenosis, right vertebral artery (VA) dissection with 70% stenosis, and suspicion of left ICA dissection. Computed tomographic angiography (CTA) neck was performed to further evaluate. CTA redemonstrated the right ICA and VA dissections. Left ICA dissection was also noted with minimal luminal narrowing and a 2-mm proximal pseudoaneurysm [Figure 1a]. Since this pseudoaneurysm was small and patient was at baseline, medical management and outpatient follow-up with neurointerventional surgery were recommended. He was placed on aspirin 81 mg daily at discharge. Anticoagulation was avoided as patient developed hemoptysis within 1 day of starting heparin drip during this admission, as well as having a history of bleeding events.

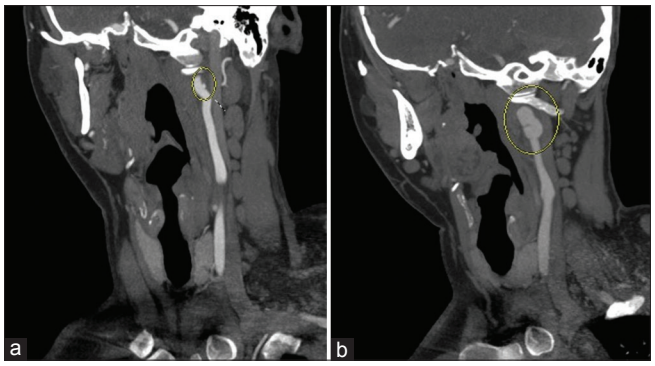

Figure 1: (a) Baseline CT angiogram (left oblique) showing small left internal carotid pseudoaneurysm. (b) Three-month posthospital follow-up CTA neck (left oblique) demonstrating left ICA pseudoaneurysm measuring 10 mm × 11 mm × 25 mm

Three-month posthospital follow-up with neurointerventional surgery was rather uneventful. However, CTA neck was repeated during this visit, which demonstrated a marked increase in size of the left ICA pseudoaneurysm – now 10 mm × 11 mm × 25 mm [Figure 1b]. Digital subtraction angiography (DSA) confirmed this finding. Medical and surgical options were discussed with the patient. Due to the rapid increase in size of the pseudoaneurysm, the patient's genetic vascular comorbidity, and risk of rupture, it was felt to be in the patient's best interest to proceed with surgical intervention. Patient opted to undergo elective pipeline stenting of the pseudoaneurysm in lieu of medical management. Aspirin was continued and he was prescribed clopidogrel 75 mg daily to begin 1 week prior to stenting procedure. On the day prior to the procedure, P2Y12 assay was performed and indicated that the patient was an adequate responder to clopidogrel.

Treatment

During the stenting procedure, a 5-Fr micropuncture technique was utilized for femoral artery access. A 6-Fr introducer sheath was placed into the vessel and a 6-Fr Aeroflex catheter was introduced into the ascending aorta. A 4-Fr Berenstein catheter was then introduced into the guide catheter and, over a 0.038 Glidewire, the aeroflex catheter was introduced into the common and ICA. 3D images were obtained during angiography, which aided in stent selection.

A Phenom 0.027 microcatheter was advanced across the pseudoaneurysm over a 0.014 Synchro 2 soft microwire. A 5.0-mm × 35-mm pipeline stent was then placed, followed by overlapping of a 5.0-mm × 25-mm variation. Postpipeline stent DSA indicated no intraluminal thrombus or luminal irregularities with good contrast filling and runoff through the

stent [Figure 2]. Catheter was removed and femoral site was sealed with angioseal.

Outcome and follow-up

Patient had no complications following procedure. He was at preoperative baseline on postoperative day 1 and was deemed stable for discharge. Discharge medications included aspirin 81 mg daily and clopidogrel 75 mg daily to prevent in-stent thrombosis. He was seen in follow-up 7 months after procedure. At that time, he noted that he stopped clopidogrel 3 months after procedure and was only taking aspirin currently, as per the recommended postoperative plan. Only complaint was fatigue. Examination revealed no neurological deficits. Follow-up DSA was performed at 7 months and showed near-complete resolution of the left ICA pseudoaneurysm [Figure 3] with minimal contrast stasis in late arterial phase [Figure 4].

DISCUSSION

Ehlers–Danlos type IV with dissection and aneurysm formation outside of the head and neck has been studied in case series, but carotid pseudoaneurysm is a rather rare problem. As a result, there have been very few reported cases. One available case documented successful aneurysm stenting, which was followed by intracranial hemorrhages and, later, fatality from rupture of intra-abdominal aneurysm.[3] The authors proposed several points including preoperative screening for aneurysm at remote sites and suggested conservative management due to increased risk of morbidity/mortality.[3] Key differences included stent type: we utilized a flexible pipeline flow-diverting stent and achieved near-complete resolution, whereas the prior study utilized a more rigid polytetrafluoroethylene-covered stent which could not navigate the tortuosity and thus failed.[3] One further case report indicated successful stent placement with no complications.[2] In the second study, utilizing the smallest soft-tipped catheter with minimal intravascular manipulation was paramount to the success of the procedure due to fragility of the vessels, along with utilizing a flexible pipeline flow-diverting stent.[2] In our procedure, a soft-tipped, small catheter was used in this fashion, which likely contributed to the success of the procedure.

CONCLUSION

Our case illustrates that pipeline stent implantation may be a viable corrective surgical option for patients with connective tissue disorders (specifically Ehlers–Danlos type IV) who present with pseudoaneurysm formation. Regardless of bleeding risk, dual-antiplatelet therapy is necessary for 3–6 months postprocedure to lower the risk of in-stent thrombosis.

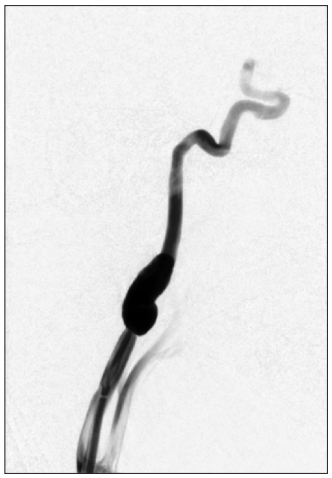

Figure 2: Immediate poststent DSA demonstrating contrast filling and runoff through the stent with no intraluminal thrombus or irregularity

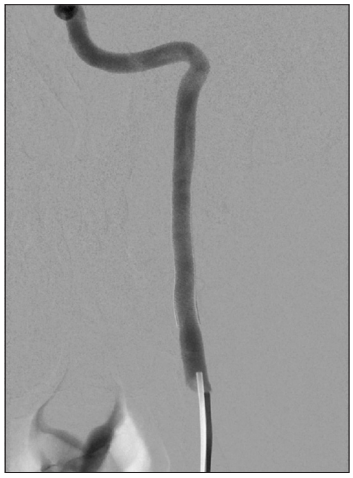

Figure 3: Seven-month poststent DSA demonstrating near-complete resolution of left ICA pseudoaneurysm with no intraluminal defects

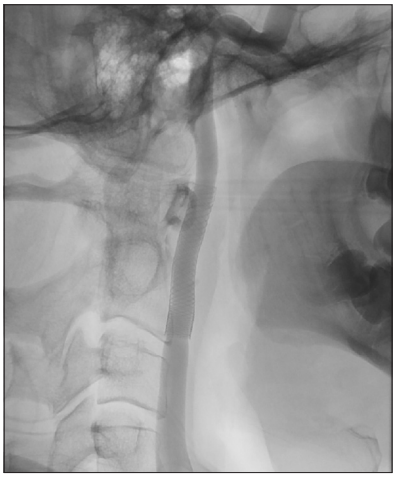

Figure 4: Seven-month poststent DSA demonstrating area of minimal contrast stasis within pseudoaneurysm in late arterial phase

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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