Journal of Cardiology Cases xxx (2018) xxx–xxx

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

Restoration of vision by endovascular revascularization in Takayasu arteritis: A case series

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ARTICLE INFO

Article history:
Received 3 March 2018
Received in revised form 20 May 2018
Accepted 29 May 2018

Keywords:
Takayasu arteritis
Bald aortic arch
Vision
Angioplasty

ABSTRACT

Takayasu arteritis (TA) is a rare, inflammatory vasculitis affecting aorta and its branches. Stenotic occlusive lesions of multiple arch arteries can cause severe cerebral ischemia leading to impaired vision. We present three consecutive young patients of TA with severe diminution of vision on upright posture, where we attempted restoration of sight by improving cerebral blood flow by percutaneous endovascular revascularization. All three patients could be successfully revascularized with substantial improvement in vision. There was no complication. On follow up, one patient developed recurrence of visual symptoms due to restenosis, which was successfully treated by cutting balloon angioplasty. The objective of this case series is to highlight the role of endovascular techniques in reversing visual loss in such situations. <**Learning objective:** Takayasu arteritis (TA) is an inflammatory vasculitis that can affect arch arteries leading to near complete obliteration and often disabling symptoms such as loss of vision. This case series (3 cases) shows that stent-supported angioplasty is a minimally invasive technique which is safe and effective in restoring impaired vision caused by severe cerebral hypoperfusion in TA.>

Introduction

Takayasu arteritis (TA) is a rare, chronic inflammatory arteriopathy affecting the aorta and its major branches. Arch arteries are commonly involved in this disease. Severe stenosis and occlusion of arch arteries can lead to cerebral hypoperfusion which may present as syncope, stroke/transient ischemic attack, seizures, or visual loss [1,2]. Occlusion of carotid arteries, vertebral arteries, or the ophthalmic arteries can result in ocular ischemia [3,4]. In acute phase of TA, immunosuppressive treatment is used to reduce progression of disease [2]. This phase may progress into chronic obliterative phase with occlusion of multiple arch arteries giving rise to devastating neurological symptoms. At this stage revascularization becomes essential to improve perfusion of the severely ischemic cerebrovascular system. Bypass graft surgery has been used in such situations, but has several challenges due to requirement of thoracotomy, multifocal arterial involvement, inflamed fragile, or calcified tissues making surgery difficult [5]. Endovascular revascularization by angioplasty is a less invasive option [5–7]. However, its use in improving diminution of vision caused by TA has been rarely reported. We report three consecutive cases where we attempted to improve vision by improving perfusion by stent-supported angioplasty.

Case report

Three patients with TA who presented with severe visual impairment were studied. Detailed history, physical examination, diagnostic, and laboratory tests were obtained. The diagnosis of TA was made according to criteria of the American College of Rheumatology [8]. Patients with raised inflammatory markers were given oral glucocorticoids. Antiplatelet drugs were given to all these patients. After written and informed consent, femoral artery access was obtained, under local anesthesia, using standard catheterization techniques. All the cases were discussed at length with a neurologist and ophthalmologist to explore the feasibility of relief after endovascular intervention. Only when all consented were the cases taken up for intervention. All patients underwent thoracic and abdominal aortography to determine the extent of vascular disease. Selective angiography of the stenosed artery was performed to localize the site and extent of vessel involvement and distal circulation.

For the intervention, a 7-F, 90-cm-long, Shuttle Flexor sheath TM (Cook Inc, Bloomington, IN, USA) was advanced over a 5 F cerebral

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https://doi.org/10.1016/j.jccase.2018.05.013
1878-5409/

Please cite this article in press as: Tyagi S, et al. Restoration of vision by endovascular revascularization in Takayasu arteritis: A case series. J Cardiol Cases (2018), https://doi.org/10.1016/j.jccase.2018.05.013

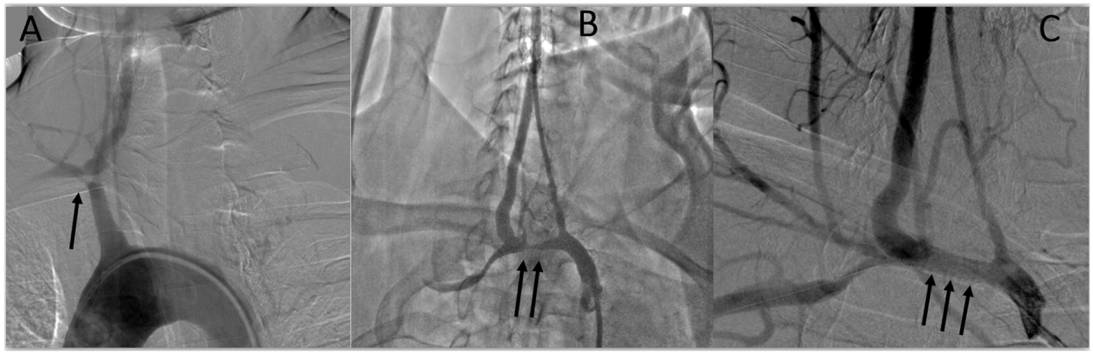

Fig 1. (A) Digital subtraction angiogram (DSA) of arch of aorta in left anterior oblique view shows total occlusion of left subclavian, left common carotid, and right common carotid artery, severe stenosis of pre vertebral segment of right vertebral artery. (B) Selective DSA of innominate artery shows severe stenosis of pre vertebral right subclavian artery. (C) Selective DSA after angioplasty and stent implantation in the pre-vertebral segment of right subclavian artery shows that there is marked improvement in lumen increasing perfusion through vertebral artery.

diagnostic catheter into the proximal portion of the stenosed artery. Intravenous heparin (5000 IU) was administered to maintain an activated clotting time of 200–250 s during the procedure. The stenotic lesion was crossed with a floppy-tipped 0.014″ coronary guidewire, taking care not to enter the intracranial segment of the artery. The lesions were dilated with a monorail balloon, inflated with diluted contrast until the waist on the balloon disappeared or rated burst pressure (RBP) of balloon was reached.

Case 1

A 23-year-old man presented with history of dizziness for the previous 2 months, which increased on standing. He had four episodes of syncope during this period. Along with this he gave history of sudden loss of vision in the right eye 4 months previously, which was diagnosed as retinal artery occlusion. Fundus examination at that time showed right eye disc edema, infranasal scarring, cherry red spot at macula, while left fundus showed no abnormal findings. There was no vision in the right eye while it was normal in the left eye. He gradually developed marked diminution of vision in the left eye for the past month with inability to see on standing. His upper limb pulsation and both carotids were not palpable. There was faint bruit in right suprasternal area. There was no renal bruit. There was no murmur.

His hemogram and blood biochemistry were normal. Erythrocyte sedimentation rate (ESR) was 29 mm in first hour, C-reactive protein (CRP) was 5.3 mg/L. Magnetic Resonance angiography of arch of aorta showed marked luminal narrowing of right common carotid artery and right subclavian artery. Left common carotid and left subclavian artery were not visualized. He had normal echocardiography. These findings suggested diagnosis of TA.

The patient was started on oral steroids, aspirin 150 mg, and clopidogrel 75 mg per day. He was taken up for aortography using standard technique as explained above. Aortogram showed total occlusion of left subclavian, left common carotid artery, severe, diffuse narrowing of right common carotid artery (Fig. 1A) and severe short segment stenosis of pre-vertebral right subclavian artery (Fig. 1B). Abdominal aorta and its branches were normal.

Angioplasty of pre-vertebral right subclavian artery stenosis was planned. Through right femoral artery approach, innominate artery was engaged as detailed above and dilatation of subclavian artery stenosis was carried out with 5×20 mm balloon. Post dilatation angiogram showed flow limiting dissection. A 6-mm diameter, 14-mm long balloon expandable stent (Express vascular SD monorail, Boston Scientific, Natick, MA, USA) was implanted. After stent implantation the pre-vertebral subclavian artery lumen diameter markedly improved with no residual stenosis. Flow through the right vertebral artery markedly improved (Fig. 1C).

The day after angioplasty, the patient reported good improvement in vision in right eye and his symptoms of dizziness on standing also disappeared. On follow up, the patient continued to have sustained improvement in symptoms. Restudy digital subtraction angiogram (DSA) done after 9 months showed continued relief of subclavian stenosis with mild intimal growth (Fig. 2). On follow up of two years, he continued to be asymptomatic with good vision with his left eye.

Case 2

A 27-year-old female presented to this hospital with complaint of blurred vision for the past 2 months, which used to further deteriorate on sitting up from a lying down position. She also had postural seizures (5–6/day for 1 month) usually precipitated by sitting up. Her symptoms were so severe that she was afraid to sit up. There was no history of headache, fever, vomiting, joint pain, or any focal neurological deficit.

On physical examination, bilateral carotid artery pulses and upper limb pulses were not palpable. On fundus examination Cup

Fig. 2. Digital subtraction angiogram on follow-up restudy shows continued patency of pre-vertebral segment of right subclavian artery perfusing the vertebral artery.

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