Endovascular Treatment of Multiple Aneurysms Complicating Cogan Syndrome

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To report the use of endografts to manage multiple aneurysms due to Cogan syndrome (CS). A 38-year-old woman with descending thoracic aorta and right common carotid artery aneurysms due to CS was treated with endovascular grafts. After 4 years, angio computed tomography scan demonstrated complete exclusion of the aneurysms with no signs of endoleak, whereas echo color Doppler showed patency of the carotid graft, no signs of restenosis, no progression of the disease in the landing zones, and complete aneurysm exclusion. Endovascular repair seems to have favorable long-term outcomes and should be considered a viable alternative to surgery in unfit for open surgery patients, even if they are young, and when the aneurysm size and location would pose a higher risk of perioperative and postoperative complications after an open surgical procedure.

Cogan syndrome (CS) is a rare, systemic, autoimmune disease which occurs predominantly in children and young adults of either sex. It was originally described as the combination of interstitial keratitis and audio vestibular disturbances, but it can also be associated with numerous systemic manifestations and, most characteristically, cardiovascular involvement.

Here we describe the case of a 38-year-old woman with descending thoracic aorta and right common carotid artery (CCA) aneurysms due to CS, both treated with endovascular endografts.

CASE REPORT

A 38-year-old woman with CS was admitted to our department with the diagnosis of aneurysms of the descending thoracic aorta and right CCA.

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The patient was hypertensive and also had dilated cardiomyopathy, with a 35% ejection fraction and mild pericardial effusion. Her medical history started in 1996, when she suffered the first episode of vertigo, nausea, and vomiting, diagnosed as Meniere's syndrome. A few months later, she experienced a new episode of vertigo associated with photophobia, tinnitus, and fever.

In 2000, the patient (28-year-old) suffered another episode of diplopia. Blood tests revealed elevated erythrosedimentation rate and white blood cell count. Anti nucleus antibodies and lupus anticoagulants were negative. The diagnosis of CS was made, and oral methylprednisolone and cyclophosphamide treatment was started. In 2003, 31-year-old, she developed left hemiparesis and brain computed tomography (CT) scan demonstrated an ischemic lesion in the right lobe. The echo color Doppler and the CT scan of the supra-aortic vessels revealed normal arteries without signs of stenosis or aneurysms. The patient showed good recovery after a period of physiotherapy. In 2009, 37-year-old, she was newly hospitalized and an angio CT scan was performed. A 6-cm diameter saccular thoracic aneurysm and a bilobed aneurysm of the right CCA were demonstrated. In this patient, the diagnosis of CS was based on the combination of inflammatory ocular disease and sensorineural hearing loss and the presence of multiple aneurysms combined with an improvement of the symptoms after the administration of corticosteroids.

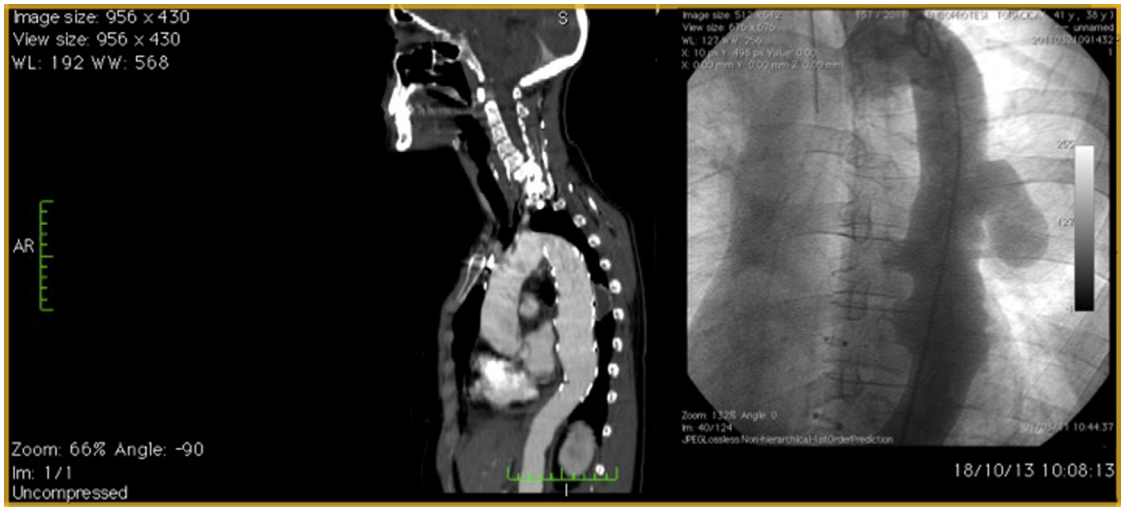

Fig. 1. Thoracic aorta aneurysm: preoperative angiography and postoperative angio CT scan (4-year follow-up).

After a careful evaluation of the risks and benefits, we opted for endovascular treatment, also in view of her poor general conditions.

The thoracic aneurysm was first treated with an endovascular procedure using a RELAY thoracic stent graft 34 × 34 × 162 mm (Bolton Medical, Sunrise, FL), whereas the carotid aneurysm was treated after 2 months (Fig. 1).

A 9 × 5-mm Viabahn Endoprothesis (W.L. Gore, Flagstaff, AZ) was placed using a left percutaneous femoral approach under local anesthesia to exclude the CCA aneurysm.

Follow-up consisted of clinical evaluation and echo color Doppler examination after 1 month from the procedure, and then every 3 months, and a CT scan every sixth month in the first year and then every year. After 4 years, all investigations demonstrated patency of the endografts, with complete exclusion of the aneurysms and no endoleak (Fig. 2).

DISCUSSION

CS can be differentiated between the typical form, characterized by acute interstitial keratitis, Menière-like vestibule and auditory dysfunction and atypical forms featuring other significant inflammatory eye lesions, non—Menière-like or remote vestibule—auditory symptoms. Atypical CS frequently mimics other rheumatologic syndromes

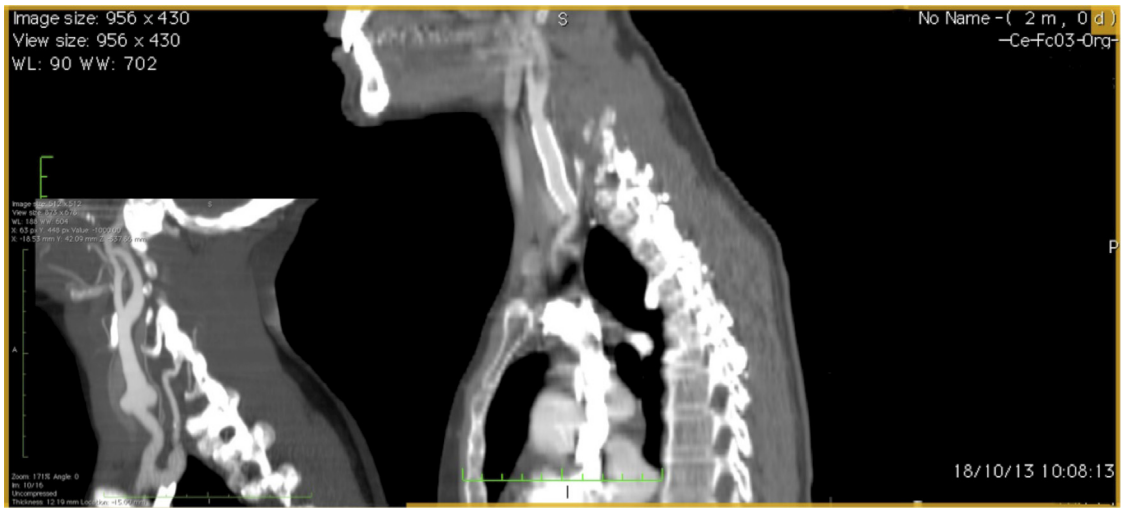

Fig. 2. Common carotid artery aneurysm: preoperative and postoperative CT scan (4-year follow-up).