Staged Hybrid Treatment of a Spontaneous Non-Infectious Innominate Artery Pseudoaneurysm with an On-Table Modified Endograft

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Abstract
Innominate Artery pseudoaneurysm is an uncommon disease. Often secondary to trauma or infection, open chest surgery has been the main surgical approach. Exclusive endovascular treatment has been used in few reports and with limited applications. Hybrid repair has also been reported, with the use of cervical debranching and endografts.

We present a unique case of a spontaneous rapidly enlarging Innominate Artery pseudoaneurysm due to Erdheim-Gsell disease repaired with a staged hybrid repair involving cervical debranching, an on-table modification and deployment of a thoracic endograft and occlusion of the pseudoaneurysm origin, followed by an Aorto-carotid bypass and aortic valve replacement in a second time.

Keywords: TEVAR; Supraaortic vessels; Embolization

Introduction
Innominate Artery (IA) pseudoaneurysm (PSA) is a very uncommon disease. Most documented aneurysmal lesions affecting the IA are true atherosclerotic aneurysms, which account for 3% of supraaortic vessels aneurysms [1]. Pseudoaneurysms of the IA are often secondary to trauma or infectious in origin [2-5]. They present a very challenging scenario with high risk of exsanguination if rupture occurs and risk of distal embolization with potential brain infarction that demands urgent treatment [6]. Different therapeutic approaches have been described, with open chest surgery being the most common access to repair the supraaortic trunks [3].

Exclusive endovascular approaches have been reported [7,8], however they are not universally applicable due to the lack of suitable stent grafts to cover a large diameter and rather short artery (around 15-16 mm mean diameter) [9]. Also depending on the proximity of the PSA regarding the IA origin, seal might be compromised [10].

We present a rare case of a spontaneous innominate trunk pseudoaneurysm secondary to Erdheim-Gsell disease that was treated with a staged hybrid repair involving an on-table modification of a thoracic endograft.

Case Report
A 71-year-old man with coronary artery disease, hypertension and moderate chronic heart failure (Class II of New York Heart Association) presented with four days history of a rapidly enlarging painless pulsatile mass on the right side of the neck base. Patient denied any fever or prior trauma and complained of persistent headaches, malaise and 9 kg weight loss in the past 10 months.

Patient was hemodynamically stable but complaining of a rapid mass growth.

On examination, the patient had a large, painless pulsatile mass on the right side of the neck base. There was no erythema or any other sign of infection. He presented with mild anemia (Hb 10,6 g/dL), a normal white cell blood count (9700 x 10^3/uL) and a slightly elevated erythrocyte sedimentation rate (27 mm/hr).

A neck/chest Computed Tomography Angiography (CTA) showed a 10 cm Innominate Artery PSA, originating 6 mm form its origin
The mass was in direct contact with the sternum and bulging towards the right side of the neck. He also had a severe stenosis on the left Subclavian Artery (Figure 1).

An open chest procedure was deemed too high risk given the proximity of the PSA to the sternum, so decision was made to proceed with a hybrid repair.

An intraoperative transesophageal echocardiogram was performed ruling out vegetations. It revealed, however, a severe aortic regurgitation. Decision was made to defer treatment at that time.

An 8 mm Dacron left Subclavian to Carotid Artery and a Carotid-carotid bypass with a retropharyngeal tunnel were performed. The left Carotid Artery was ligated proximal to the bypass. Two (8 and 10 mm) Begaft stents (Bentley, InnoMed, Hechingen, Germany) were deployed in the left Subclavian Artery (LSCA) to treat the stenosis.

We decided the off-label use of a thoracic endograft. A 36x150 Valiant endograft (Medtronic, Santa Rosa, Calif, USA) was deployed on the operating table. The endograft was subsequently re-sheathed, vigorously flushed with saline and brought up through a left femoral artery cutdown. The endograft was positioned and deployed in the Ascending Aorta with aid of deep hypotension, covering the bovine trunk and landing right proximal the LSCA origin, so the entire brain was being perfused through the stented LSCA and the cervical debranching. While deploying the endograft it moved one cm proximally, landing 4 cms distal to the aortic valve but compromising distal seal, thus a second endograft was customized and deployed right proximal de LSCA origin to obtain an adequate seal.

Once completed, we obtained retrograde right Carotid Artery access proximal to the bypass and under direct vision advanced an 8 Fr sheath. Multiple Concerto coils (Medtronic, Santa Rosa, Calf, USA) and a 20 mm Amplatzer vascular plug II (Abbott, Abbott Park, IL, USA) were deployed and packed proximally, occluding the PSA origin. The Innominate Artery bifurcation was preserved in order to secure flow to the right Vertebral and Brachial arteries (Figure 2).

Patient recovery was uneventful, and the mass was noted to be pulseless. Cultures were negative. Due to the suspicion of Giant Cell Arteritis, the patient underwent bilateral Temporal artery biopsy that came back negative.

Given his good overall condition but symptomatic severe aortic regurgitation a standard open aortic valve replacement with a biological graft was performed 3 weeks after initial surgery. During sternotomy PSA cavity was visible with multiple coils and no signs of bleeding. Given the risk of leaving brain perfusion only to the stented left subclavian artery, a 10 mm Dacron ascending Aorta to right Carotid bypass was performed. Aortic wall tissue cultures and biopsy were obtained.

Patient recovery was uneventful, except for a new onset atrial fibrillation, and was discharged two weeks after the last surgery on Coumadin. Aortic biopsy showed evidence of Cystic Medial
Necrosis (CMN) (Figure 3). Considering age, absence of familiar history and no clinical features of connective tissue disorders the patient was considered an idiopathic form of CMN.

Patient resumed his daily activities shortly after discharge and at one-year follow-up he presents with no complaints. CTA showed the thoracic endograft in position and neck debranching with patency of all the bypasses (Figure 4).

Discussion

Innominate Artery PSA is a very uncommon emergency. More frequently related to infection or trauma, we present an extremely rare presentation secondary to Cystic Medial Necrosis (CMN) not yet described to our knowledge.

Also known as Erdheim-Gsell, CMN was first described by Erdheim and Gsell in 1929 [11]. It is characterized by the formation of cystic cavities with mucoid material and fragmentation of elastic medial filaments [12]. It has been associated with aortic aneurysms and dissection in patients with Marfan syndrome [13-15] and it has also been described as an idiopathic form most frequently present in elderly population [16]. The Ascending Aorta is the most common site affected by CMN, with reports showing a 20-65% incidence among patients operated for type A Aortic Dissection [15-17]. It has been hypothesized that the higher rate of CMN in the Ascending Aorta might be related to the higher concentration of elastic fibers in the media compared to the Descending or Abdominal Aorta [16]. It has also been reported in many different locations as the Supraaortic Vessels (Carotid and Subclavian arteries) [13,18], Abdominal Aorta [19], and even iliac Artery [14]. Given the pathologic disruption of the elastic fibers of tunica media, this disease confers high risk for aneurysm formation, dissection and spontaneous rupture [20]. Patients present long-life risk for complications; therefore, close long-term surveillance is warranted [15].

Our case presented several difficulties. Open surgery was ruled out by the cardiac surgery team due to the high risk of exsanguination during sternotomy. A pure endovascular approach, covering the Innominate Artery would have been unsuccessful given the short neck to obtain an adequate seal in the proximal Brachiocephalic Artery (around 6 mm). The presence of a common origin of the IA with the left Carotid Artery (bovine trunk) also precluded the use of total endovascular fenestrated or branched devices in this case. The use of chimneys might have been considered; however, this approach could still leave flow within the gutters, thus maintaining risk of PSA rupture.

Despite the fact there is scarce experience of innominate PSA repaired with hybrid approaches (and predominantly for infectious and traumatic false aneurysms) [4,6,10,21], we consider hybrid management with the off-label use of endografts and cervical debranching the best possible treatment in a patient with an emergency and no open surgical options.

Our approach involved extraanatomical supraaortic vessel debranching, with an on-table modification of a thoracic endograft and its deployment in the Ascending Aorta, covering the bovine arch and occluding the IA and PSA origin with coils and an Amplatz plug. In a second stage we secured brain perfusion with an Ascending Aorta to Carotid bypass along with an aortic valve replacement.

Our patient made a full recovery and, despite the need for lifetime surveillance, has an excellent long-term prognosis.

Declarations of Interest

None
References


