Endovascular Treatment of Isolated Chronic Abdominal Aortic Dissection

Tratamento Endovascular da Disseção Crónica Isolada da Aorta Abdominal

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ABSTRACT

Isolated acute abdominal aortic dissection is a relatively rare event. Its natural history is not fully understood and its optimal treatment is not established. Open surgery represents the most described treatment but endovascular intervention has had increasing application. Isolated chronic abdominal aortic dissection is even less described in the literature. We describe three patients with isolated chronic abdominal aortic dissection who underwent endovascular treatment in our institution. Mean age at presentation was 82 years. Indication for surgical intervention was aneurismal degeneration. Mean aortic diameter at presentation was 46.7 mm. There was no perioperative mortality or reinterventions. Mean follow-up was 5.3 years (2-12 years). Late reintervention was needed in one patient, eight years after initial surgery, due to type 1 endoleak. According to our experience, endovascular intervention represents an effective and durable treatment option in isolated chronic abdominal aortic dissection. However, long-term follow-up is mandatory. Furthermore, larger studies are still needed to understand this disease and its adequate treatment.

Keywords: Aneurysm, Dissecting; Aortic Aneurysm, Abdominal; Blood Vessel Prosthesis Implantation; Endovascular Procedures.

RESUMO

A disseção isolada da aorta abdominal é um evento raro. A sua história natural e tratamento ideal não estão totalmente esclarecidos. A intervenção endovascular tem ganho uma preponderância crescente. A disseção crónica isolada da aorta abdominal está ainda menos descrita na literatura. Descrevemos três casos de doentes com disseção crónica isolada da aorta abdominal submetidos a tratamento endovascular na nossa instituição. A média de idades dos doentes era de 82 anos. A indicação terapêutica foi degeneração aneurismática. O diâmetro aórtico médio à apresentação era de 46,7 mm. Não houve mortalidade ou reintervenção peri-operatória. O seguimento pós-operatório médio foi de 5,3 anos (2-12 anos). Realizou-se reintervenção tardia num doente, oito anos após a cirurgia inicial, por endofuga tipo I. Segundo a nossa curta experiência o tratamento endovascular representa uma opção terapêutica eficaz na disseção crónica isolada da aorta abdominal. O *follow-up* alargado é mandatório. São necessários estudos com maiores amostras e *follow-up* para melhor compreender esta patologia.

Palavras-chave: Aneurisma da Aorta Abdominal; Aneurisma Dissecante; Implante de Prótese Vascular; Procedimentos Endovasculares.

INTRODUCTION

The thoracic aorta is the most commonly involved aortic section when acute aortic dissection (AD) occurs. It may also extend to the abdominal aorta.[1] However, acute isolated abdominal aortic dissection (IAAD) is a relatively rare event, representing only 1% to 4% of the AD cases, with only 18 cases reported in the International Registry of Acute Aortic Dissection (IRAD) between 1996 and 2003.[2,3] It can occur spontaneously or in association with trauma and surgical procedures (specially endovascular).[4-6] The association with atherosclerosis or an iatrogenic etiology is stronger for acute IAAD than for classical Stanford type B AD. Acute IAAD most commonly presents with abdominal pain, visceral ischemia (including acute renal failure) and limb ischemia. Mean reported age of presentation is 67.7 ± 13.3 years[2], similar to that of classical type B aortic dissection.

Medically treated chronic dissection tends to progress with aneurysmal degeneration. Due to wall weakening induced by chronic dissection, with higher rupture risk, many surgeons advise earlier intervention in dissecting aneurysms, usually when they reach a diameter of 3 cm.[1,6,7]

In result of the scarcity of reported cases, management of IAAD is still a matter of controversy. We describe three cases of chronic IAAD who underwent endovascular treatment. These cases occurred during a time frame of 10 years and no other cases of IAAD were described in our institution during this period.

CASE REPORT

Case 1

An asymptomatic abdominal aortic aneurysm was found on duplex ultrasound in a 78 year-old male with hypertension. CT angiography (CTA) revealed an 85mm fusiform infra-renal AAA with an associated isolated abdominal aortic dissection (CTA images not available). The thoracic aorta had a normal diameter and lumen.

The patient was considered fit for open surgery but due to his preference endovascular treatment was chosen. A bifurcated abdominal aortic Talent endograft (Medtronic Vascular, USA) was deployed through a bilateral femoral cutdown. No early, medical or surgical, postoperative complications were observed.

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During follow-up a consistent reduction of aortic diameter was observed with a maximum aortic diameter at year 7 of 73 mm with no endoleaks. During the 8th year post-EVAR the patient repeated a CTA which revealed an endoleak (suggestive of type III EL) and growth of aneurysm diameter (79,7 mm). EVAR was chosen to treat this late complication, intra-operative angiography revealed a type Ia endoleak which was treated with an aorto-uni-iliac stent graft (to ensure that a possible associated type III EL would be treated as well) and cross-over femoro-femoral bypass. Two early surgical reinterventions were performed to drain groin hematomas. At the 12th year of follow-up (four years after reintervention), the patient was asymptomatic and CTA showed aneurysm diameter stabilization at 80 mm with no endoleaks.

Case 2

An 83 year-old male presented with an asymptomatic iliac dissecting aneurysm discovered on a CT during the follow-up of a urinary bladder adenocarcinoma. Patient was an ex-smoker, with stage 3 CKD (glomerular filtration rate of 30-59 mL/min). CTA showed an IAAD beginning distal to the inferior mesenteric artery's (IMA) origin and extending to the mid third of the right external iliac artery (EIA) with a right iliac artery aneurysm with a maximum diameter of 30 mm and a proximal neck length (to the aortic bifurcation) of 22 mm (Fig. 1). Distal aortic diameter was 25 mm and both the left CIA and external iliac arteries measured 15 mm in diameter.

Endovascular treatment was chosen. Through a bilateral open femoral access, an Excluder endograft (W.L. Gore & Associates, Flagstaff, Arizona, USA) was implanted and extended to the EIA on the right side. No early, medical or surgical, postoperative complications were observed.

During follow-up (33 months), the patient presented a tolerated right gluteal claudication. Follow-up CTA's revealed a reduction of iliac aneurysm's diameter to 25 mm with no endoleaks.

Case 3

An 85 year-old female, without known cardiovascular risk factors, was diagnosed with a saccular aneurysm of the terminal abdominal aorta on DUS. CTA confirmed a 25 mm saccular aneurysm proximal to the aortic bifurcation in relation to an IAAD originating at the IMA level and with a reentry point 15mm proximal to the aortic bifurcation (Fig. 2). She had a proximal, infra-renal, neck measuring 40mm in length without angulation. The maximum aortic diameter was 16 mm with 15 mm at the bifurcation level; right and left CIA's measured 9 mm and 8 mm respectively.

Due to patient's age and suitable anatomy it was decided to proceed with endovascular treatment. Surgical femoral access was obtained to deliver an Endurant II endograft (Medtronic Vascular, USA) iliac limb 20 x 80 mm trimmed down to 70 mm length (Fig. 3). No early, medical or surgical, postoperative complications were observed.

At 24 months of follow-up the patient was asymptomatic and aneurysm sac shrinkage (20 mm of maximum diameter) without endoleaks was confirmed on CTA.

DISCUSSION

Treatment strategies for acute IAAD include open surgery, endovascular intervention or medical treatment. The results from a meta-analysis published by Jonker et al[8] suggest that an early aggressive (open or endovascular surgery) may be associated with improved survival. However this is a retrospective study with only 19 patients treated with endografts, therefore, in our institution, we still manage the AIAAD with medical treatment and treat these patients in case of ischemic complications. In the chronic phase we perform surgery if they develop an aneurysm (> 3 cm or saccular morphology).

Despite the good historical results described for open surgery,[1,2,6] especially in good risk patients, the evolution of endovascular therapies has allowed the effective treatment of high operative risk patients with good long-term results[9]. In fact, the excellent results obtained by some centers supports endovascular techniques as the first-line treatment in all patients given the presence of adequate anatomy.[9,10]

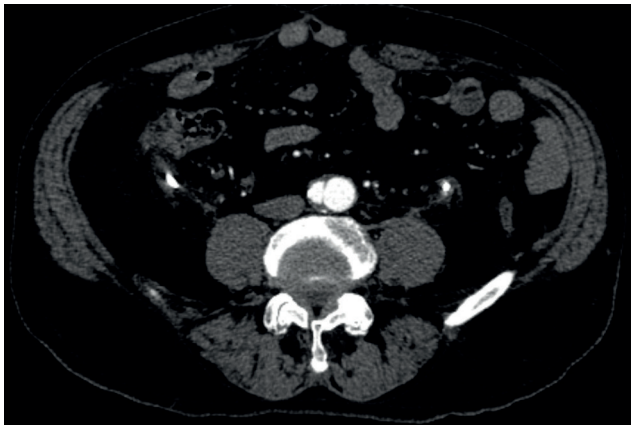

Figure 1 - Chronic IAAD in the distal infra-renal aorta extending to the mid third of the right external iliac artery (EIA) with a right iliac artery aneurysm in an 83 year old male

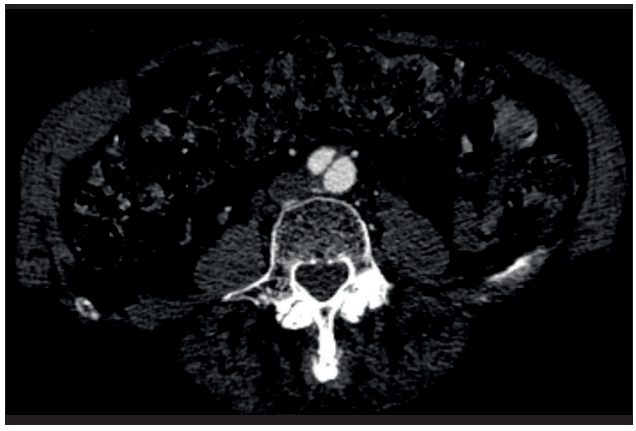

Figure 2 - A 25 mm saccular aneurysm proximal to the aortic bifurcation in relation to a chronic IAAD in an 85 year old female

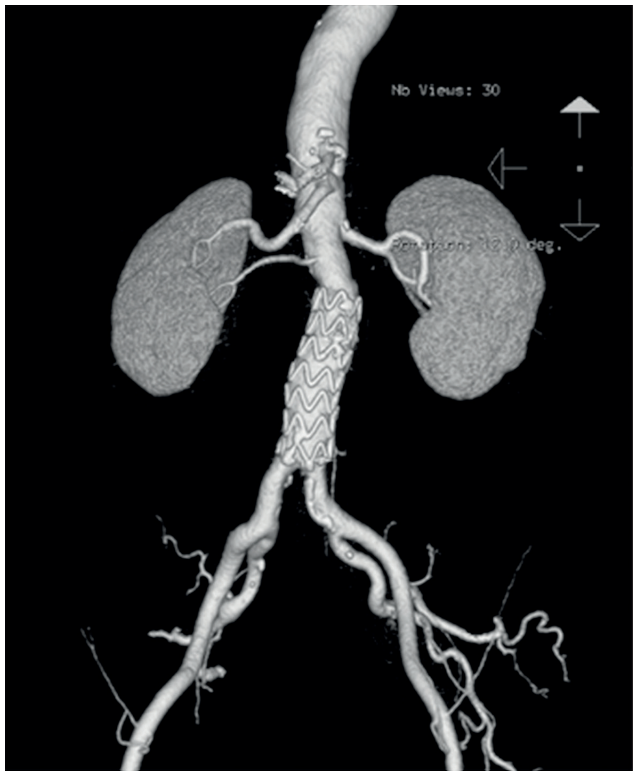

Figure 3 - Saccular dissecting aortic aneurysm treated with an Endurant II (Medtronic Vascular, USA) iliac limb 20 x 80 mm trimmed down to 70 mm length

We emphasize the fact that all 3 cases described above were old patients at presentation (the younger patient was 78 years-old), older than the mean age reported in most studies both for chronic IAAD and classical type B aortic dissection. This fact was also accounted for the choice of an endovascular approach. However, in our institution, age itself isn't a criterion for treatment's selection; in fact, comorbidities, anatomic criteria and the patient's informed decision play a preponderant role. Despite old age, no early deaths were observed and the sole reintervention occurred eight years after the initial procedure due to a late endoleak. This reflects that endovascular treatment is effective in appropriately selected patients but also underlines the importance of maintaining long-term clinical and imaging follow-up. In our institution patients treated for chronic IAAD enter the same follow-up protocol as those treated with endografts for AAA (CTA at 3, 9 months after surgery and then yearly).

In chronic IAAD, due to aortic wall weakening, adequate neck (defined as the normal infra-renal aorta above the proximal intimal tear/aneurysm sac) are imperative to allow good seal and prevent graft migration (> 15 mm in length, diameter < 28 mm and angulation < 60).

Case 3 also shows that even in cases with inadequate anatomy for conventional abdominal aortic endografts, presenting with small aortic and iliac diameters, modified endovascular techniques have the flexibility to provide safe and effective treatment given an adequate sealing zone.

The retrospective nature of this case-series is one of the limitations of this report. The small sample also precludes any definitive conclusions. The long time lapse between the treated cases can also introduce a bias in the results due to the evolution of better endovascular devices and of the surgeons' endovascular skills.

CONCLUSION

Our results show that endovascular treatment for chronic IAAD is feasible, effective and durable. However, long-term follow-up is mandatory as late endoleaks may occur. Due to its rarity, chronic IAAD is still a relatively understudied pathology and its optimal treatment is not well established. Therefore, we consider that further multicentre studies with long-term follow-ups are necessary; however, these are unlikely to be expected due to the rarity of the pathology.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patient's data publication.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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Elastose Perfurante Serpiginosa e Doença de Wilson: Uma Consequência Rara, mas Previsível da Terapêutica a Longo Prazo com D-Penicilamina

Elastosis Perforans Serpiginosa and Wilson Disease: A Rare but Predictable Consequence of Long-term Therapy with D-Penicillamine

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RESUMO

A elastose perfurante serpiginosa é uma dermatose perfurante rara, encontrada sobretudo em adolescentes e adultos jovens, caraterizada pela eliminação transepidérmica de fibras elásticas anómalas. O único fármaco conhecido capaz de induzir elastose perfurante serpiginosa é a D-penicilamina. Descrevemos o caso de uma doente de 52 anos com pápulas queratósicas confluentes com disposição anular e crescimento centrífugo, localizadas na região cervical anterior. A doente estava cronicamente medicada com D-penicilamina, por doença de Wilson. A biópsia lesional revelou eliminação transepidérmica de fibras elásticas com aumento da eosinofilia, espessadas, ramificadas e de aspeto em dentes de serra. Os achados clinicopatológicos foram compatíveis com elastose perfurante serpiginosa secundária à D-penicilamina. Estima-se que a elastose perfurante serpiginosa ocorra em 1% dos doentes medicados com D-penicilamina. Bloqueando direta ou indiretamente as ligações cruzadas de desmosina da elastina, a D-penicilamina leva à síntese de fibras elásticas anómalas dérmicas, mas também extracutâneas. A elastose perfurante serpiginosa pode ser a primeira manifestação de um processo degenerativo multissistémico do tecido conjuntivo elástico.

Palavras-chave: Degeneração Hepatolenticular; Doenças da Pele/induzida quimicamente; Penicilamina; Quelantes/uso terapêutico.

ABSTRACT

Elastosis perfurans serpiginosa (EPS) is a rare perforating dermatosis found primarily in adolescents and young adults, characterized by transepidermal elimination of abnormal elastic fibers. The only drug known capable of inducing EPS is D-penicillamine. We report the case of a 52 year-old woman with keratotic papules arranged in an annular pattern with central clearing and centrifugal growth, located in the anterior cervical region. The patient was chronically treated with D-penicillamine for Wilson disease. Lesion biopsy showed transepidermal elimination of thickened, eosinophilic, branched, sawtooth-like elastic fibers. The clinical and pathological findings were consistent with EPS secondary to D-penicillamine. It is estimated that EPS occurs in 1% of patients treated with D-penicillamine. By blocking directly or indirectly the desmosine cross-links between elastin molecules, D-penicillamine leads to the synthesis of abnormal dermal and extracutaneous elastic fibers. EPS may be the first manifestation of a multisystemic degenerative process of elastic connective tissue.

Keywords: Chelating Agents/therapeutic use; Hepatolenticular Degeneration; Penicillamine; Skin Diseases/chemically induced.

INTRODUÇÃO

A elastose perfurante serpiginosa (EPS) é uma entidade rara que pertence ao grupo das "dermatoses perfurantes primárias" onde se incluem a colagenose perfurante reativa, as foliculites perfurantes e a doença de Kyrle.[1]

O conceito "dermatose perfurante adquirida" é usado por alguns autores, em sentido lato, para incluir as formas primárias que surgem no contexto de diabetes *mellitus,* doença renal crónica, ou ambas.[2]

As dermatoses perfurantes são assim designadas por partilharem o achado histológico de perfuração epidérmica com eliminação transepidérmica de tecido conjuntivo dérmico, e são em geral divididas de acordo com o tipo material expelido e o local da perfuração. Na colagenose perfurante reativa são eliminadas sobretudo fibras de colagénio, na EPS fibras elásticas, enquanto nas foliculites perfurantes o processo de eliminação ocorre através do epitélio do folículo piloso.[1-3]

A EPS é encontrada predominantemente em adolescentes e adultos jovens, sendo até três vezes mais frequente no sexo masculino.[1]

Esta entidade tem sido tradicionalmente classificada, por ordem decrescente de frequência, como idiopática – a maioria dos casos, reativa – cerca de 25 - 40%, ou excecionalmente fármaco-induzida.[3-5]

Entre os casos idiopáticos tem sido descrita associação familiar, com formas hereditárias autossómicas dominantes ou recessivas.[5] A forma reativa pode ocorrer associada a síndrome de Down, síndrome de Marfan, síndrome de

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