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CASE REPORT

Endovascular Aneurysm Repair (EVAR) of an Infrarenal Abdominal Aortic Aneurysm (AAA) in a Young Patient with Systemic Lupus Erythematosus (SLE)

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Introduction: While hypertension, pericardial, myocardial, and coronary artery disease are common cardiovascular manifestations of systemic lupus erythematosus (SLE), aortic aneurysms (AA) are rare but increasingly diagnosed, with the true incidence unknown.
Case report: A 40 year old female suffering from SLE with a 5.3 cm saccular eccentric infrarenal abdominal aortic aneurysm (AAA) was treated successfully with endovascular aneurysm repair (EVAR) using the Medtronic Endurant II bifurcated stent graft and followed up 2 years post-operatively. Pre-operatively, open and EVAR options were offered and the latter was chosen by the patient.
Discussion: Proposed mechanisms for AA formation in SLE including accelerated atherosclerosis brought about by chronic steroid use and SLE associated vasculitis and cystic medial degeneration (CMD) have been discussed in other case reports and series. To the authors' knowledge, the use of EVAR for AAA in SLE patients has not been reported in available literature. The need for earlier repair, screening, and detection as well as the long-term suitability, durability, and surveillance of EVAR remain unknown. The benefit of using the Ovation device in minimising late neck dilatation is also discussed.
Conclusion: EVAR was demonstrated to be a suitable form of repair in a young female patient with SLE and AAA, followed up 2 years post-surgery. The ideal repair and the natural history of these aneurysms remains to be studied.

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INTRODUCTION

While hypertension, pericardial, and coronary artery disease are common cardiovascular manifestations of systemic lupus erythematosus (SLE), aortic aneurysms (AA) are rare but increasingly diagnosed. Little is known about their pathophysiology and natural history.

Herein, a case is presented of a 40 year old female SLE patient whose AAA was successfully treated by EVAR.

CASE REPORT

A 40 year old female, diagnosed with SLE at age 24 years was referred with an asymptomatic pulsatile epigastric mass. Computed tomography aortogram (CTA) showed a 5.3 cm infrarenal right sided saccular aneurysm, 4 cm inferior to the origin of the left renal artery with an eccentric rind of mural thrombus and no suggestion of aortitis. Atherosclerotic calcification was noted within (Fig. 1).

The patient first presented with malar rash and lower limb swelling secondary to lupus nephritis, developing end stage renal failure at 27 years necessitating haemodialysis. Autoimmune panel was positive for anti-nuclear antibody, ENA, anti-Smith, anti-RNP, anti-Ro, anti-double stranded DNA, and anti-cardiolipin IgG. In addition, she suffered from chronic haemolytic anaemia, hypertension, dyslipidaemia, and coronary arterial disease. She had been on various anti-hypertensives, statins, immunosuppressant and disease modifying anti-rheumatic drugs including cyclophosphamide, prednisolone, azathioprine, and mycophenolate mofetil, with multiple admissions for drug related complications including thrombocytopaenia and osteoporosis from 24 years of age up to peri-operatively.

Serum C-reactive protein and erythrocyte sedimentation ratio were not elevated and blood cultures were negative although these may have been influenced by the use of immunosuppressants. Retrospectively, it was noted that a 4.3 cm infrarenal AAA was present 3 years previously during an ultrasonographic evaluation of her kidneys although she was not referred at that time.

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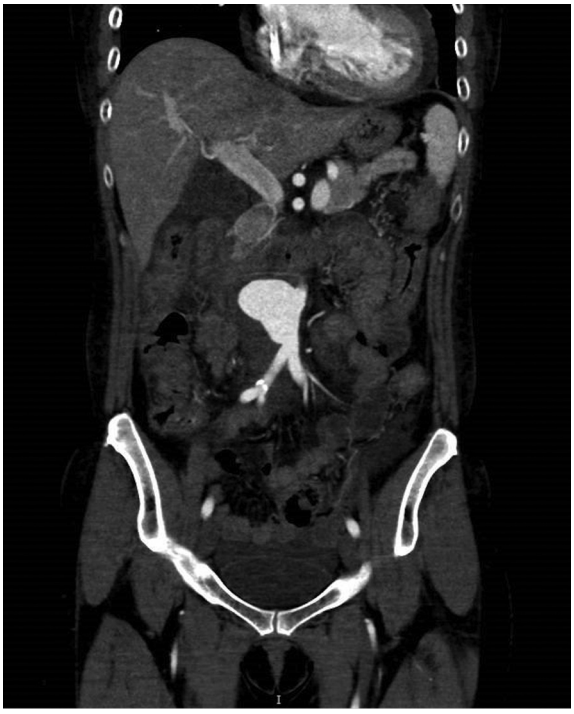

Figure 1. Coronal view of infrarenal saccular aneurysm.

A choice of open or EVAR was offered considering her age, and EVAR was chosen given the expected shorter length of stay, avoidance of laparotomy and its associated morbidities including increased pain, adhesions, incisional hernias, while bearing in mind risks of device failure, continued neck dilatation, and need for lifelong surveillance with EVAR.

The Medtronic Endurant II bifurcated stent graft (Medtronic International Ltd, Nasaco Tech Centre, Singapore) was inserted under local anaesthesia with mild sedation. The main body (ETBF 23-13c124 EE) was delivered via the right groin and the contralateral limb (ETLW 16-13c 82EE) through the left after ultrasound guided access. Operative time was 75 minutes and 160 mL of half strength contrast was used (Fig. 2A and B). Closure was achieved using the Abbott Perclose Proglide suture mediated closure system.

The patient was discharged well on the third post-operative day. CTA performed at 1 and 6 months post-EVAR (Fig. 3) and contrast enhanced ultrasonography at 2 years showed that the graft continues to exclude the sac satisfactorily.

DISCUSSION

AA is a rare but increasingly diagnosed cardiovascular manifestation of SLE.[1] This could be in part because of increased availability of non-invasive imaging and use of drugs which have led to prolonged survival of SLE patients.[2]

Little is known about the pathophysiology of these aneurysms; if they are purely associated with SLE or contributed to by other forms of vasculitis.[3] Nevertheless, it is clear that several differences exist between them and AA in the non-SLE population.

AA in SLE patients is seen to affect females predominantly at a younger age on long-term immunosuppressants but not in other conditions which require long-term steroids and immunosuppressants.[1,2] The reported locations have also been highly variable with enlargement of the entire aorta noted in rare cases.[4,5] Ohara et al. described his experience in only five SLE patients of 429 AA over 10 years.[1] Similar to the present patient, four received long-term corticosteroid treatment over a mean of 23 years. The average age at diagnosis was 20 years younger than in the non-SLE population.

The pathogenesis has been attributed to circulatory disturbances resulting from associated vasculitis and CMD as

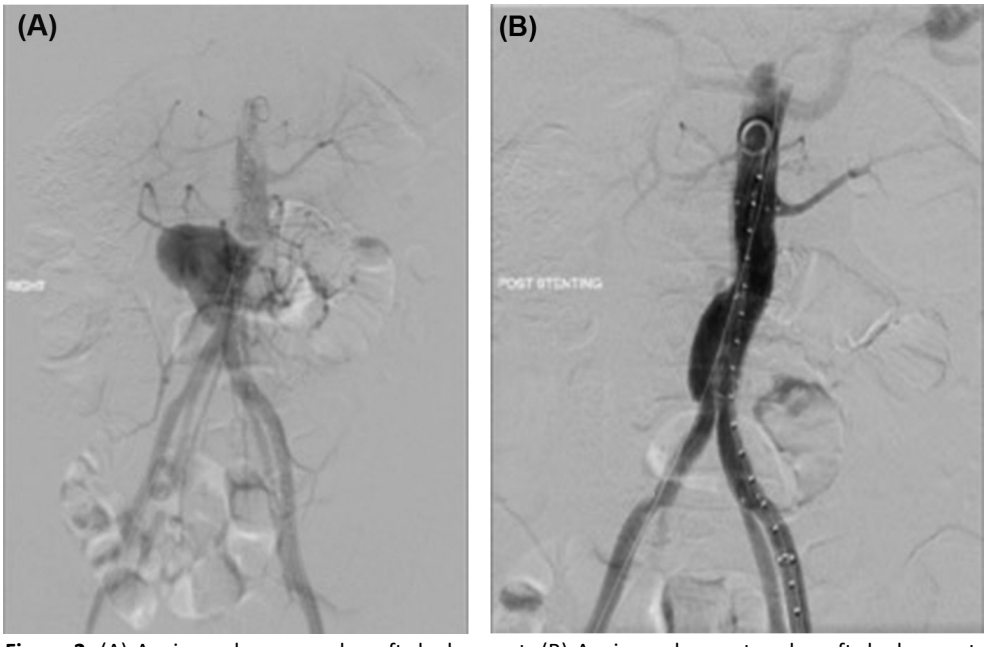

Figure 2. (A) Angiography, pre-endograft deployment. (B) Angiography, post-endograft deployment.

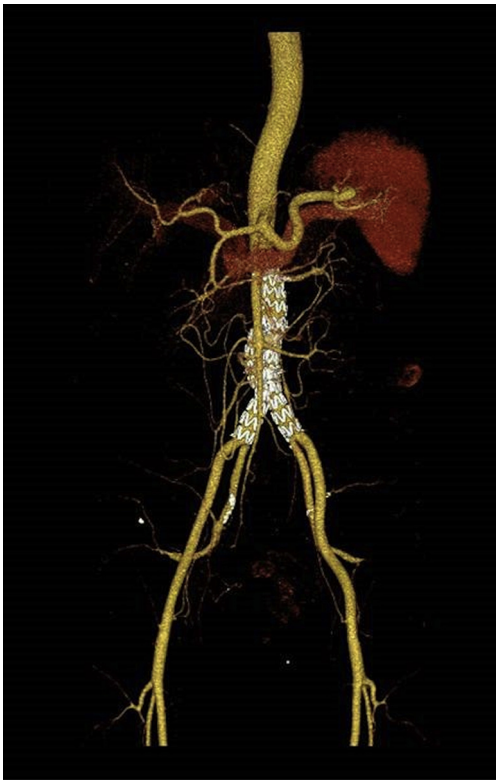

Figure 3. Virtual reality rendition of aorta post-endograft deployment.

well as accelerated atherosclerosis.[2] Atherosclerosis is considered to be caused by long-term steroid treatment contributed by dyslipidaemia, hypertension, and nephrotic syndrome.[2] The present patient had been on atorvastatin over the previous 10 years. At the time of diagnosis, her lipid panel was well controlled (total cholesterol 3.75, HDL 1.38, TG 1.07, LDL 1.88 mmol/L). In addition, she had been on immunosuppressants and steroids for over 15 years as well as peri-operatively. Considering that AA formation and rupture results from an early failure of elastin and a late failure of collagen, it is possible that steroid usage has a bearing on collagen failure. This is notable in wound failure, notwithstanding other effects including down regulated lysosomal activity.

Kurata et al. published a meta-analysis to clarify characteristics that may contribute to aneurysm formation in SLE.[2] Relevant studies from 1969 to 2008 were analysed and only 35 cases were identified. Factors correlating with thoracic or abdominal aneurysms differed and two principal patterns were proposed, one being the fatal non-atherosclerotic thoracic aneurysm which was associated with CMD and vasculitis and the other, the atherosclerotic abdominal aneurysm which was associated with long-term steroid treatment and a relatively favourable prognosis. Kurata proposed that CMD in SLE occurred at a younger age and was closely associated with vasculitis rather than atherosclerosis, affecting thoracic over abdominal aorta. Early recognition and careful evaluation of patients with vasculitis and CMD is crucial in thoracic lesions. Abdominal lesions, on the other hand, tend to be related to the duration of steroid therapy. Prolonged steroid use promotes atherosclerosis and may facilitate aneurysm formation at the abdominal aorta, which is a major site of atherosclerosis. Likewise, Washiyama et al. proposed that in patients with SLE, the aneurysms may be classified into those caused by inflammation and those resulting from atherosclerosis caused by prolonged steroid therapy.[3]

Some reports have described detailed histopathological findings of these aneurysms. Stehben et al. gave a pathological description of a non-dissecting AA with SLE.[6] The findings of thrombi with underlying atheromatous debris containing lipophages and medial elastic fibres deep to the debris, did not differ from findings in atherosclerotic AAA without SLE. The absence of necrotic micro-vasculitis accompanied by fibrinoid necrosis otherwise reflected the effect of long-term steroid therapy which resulted in weakening of the medial elastic lamina. Meanwhile other reports have shown elements of vasculitis or CMD in these aneuryms.[7,8]

It is possible that the disease pattern or pathogenesis is a balance between vasculitis or SLE associated auto-immune conditions and the accelerated atherosclerosis. With optimal medial therapy, accelerated atherosclerosis may have a bigger role than vasculitis in the pathogenesis, while the converse is true in suboptimal medical therapy. Perhaps, the pathogenesis is multifactorial, thereby leading to the spectrum of aortic diseases reported.

Hypertension, the most common cardiovascular manifestation of SLE is also often thought to be a further risk factor for acceleration of atherosclerosis and dissection or rupture.[2] Wang et al. stated that “The patient who had SLE associated with vasculitis was often normotensive, while those without vasculitis had a history of hypertension.”[2] Likewise, in the meta-analysis, a history of hypertension had a negative correlation with rupture and vasculitis. Overall this suggests that the present patient having a history of hypertension, although well controlled, had a SLE associated abdominal aneurysm that conformed more to the prolonged steroid, accelerated atherosclerosis disease pattern.

To the authors’ knowledge, the use of EVAR for AAA in SLE has not been reported. Oliveira et al. reported a ruptured descending thoracic AA in a 25 year old female with SLE who underwent 19 years of steroid therapy.[9] She was treated with two endovascular stent grafts and discharged well 13 days after the procedure. Three months later, she returned with haemorrhagic shock from an aortic-esophageal fistula. She underwent open emergency surgery, and died post-operatively. Two other cases of successful stent grafting of descending thoracic AA were reported but little is known about long-term durability of repair.[10]

In addition, it is recognised that the Ovation abdominal stent graft platform may be superior in this case in terms of reducing late neck dilation particularly in a young patient as the graft’s customisable ring shaped channels can be injected with a polyethylene glycol based polymer, thus expanding the endograft against the aorta creating a circumferential proximal seal conforming to vessel wall irregularities at the mid-point of the sealing ring. This exerts

no chronic outward force and insulates the neck from blood pressure. Patients treated as such had no neck dilatation and late Type 1 endoleaks at 3 years.[11] The Ovation device was not available at the time of this patient's management.

CONCLUSION

In conclusion, EVAR was demonstrated to be a suitable technique with good results in a young female patient with SLE and AAA, followed up 2 years post-surgery. It is difficult to know the natural history, ideal repair, and long-term follow-up required given the disease rarity even with further studies. However, given the morphology, expansion rate, as well as association with underlying autoimmune or inflammatory process, the present authors elect to repair and feel that treatment before the typical diameter of 5cm is necessary to avoid catastrophic rupture. Long-term complications including re-intervention rates are unknown and surveillance remains highly relevant.

CONFLICT OF INTEREST

None.

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REFERENCES

1 Ohara N, Miyata T, Kurata A, Oshiro H, Sato O, Shigematsu H. Ten years- experience of aortic aneurysm associated with systemic lupus erythematosus. *Eur J Vasc Endovasc Surg* 1999;**3**:288—93. https://doi.org/10.1053/ejvs.1999.0982.

2 Kurata Atsushi. Aortic aneurysms in systemic lupus erythematosus: a meta-analysis of 35 cases in the literature and two different pathogeneses. *Cardiovasc Pathol* 2010;**20**: e1—7.

3 Washiyama N, Kazui T, Takinami M. Surgical treatment of recurrent abdominal aortic aneurysm in a patient with systemic lupus erythematosus. *J Vasc Surg* 2000;**32**:9—12.

4 Walts AE, Dubois EL. Acute dissecting aneurysm of the aorta as the fatal event in systemic lupus erythematosus. *Am Heart J* 1977;**93**:378—81.

5 Yoshimoto K, Saima S, Nakamura Y, Ishikawa H, Kinoshita M, Yokohari R, et al. A case of acute dissecting aneurysm of the aorta in systemic lupus erythematosus. *Nippon Jinzo Gakkai Shi* 1989;**31**:1211—6.

6 Stehbens WE, Delahunt B, Shirer WC, Naik DK. Aortic aneurysm in systemic lupus erythematosus. *Histopathology* 1993;**22**:275—7.

7 Ruddy JM, Jones JA, Spinale FG, Ikonomidis JS. Regional heterogeneity within the aorta: relevance to aneurysm disease. *J Thorac Cardiovasc Surg* 2008;**136**:1123—30.

8 Corominas H, Tsokos M, Quezado M, Tsokos GC. Aneurysm of the ascending aorta in systemic lupus erythematosus: case report and review of the literature. *Eur J Rheumatol* 2017 Jun;**4**:133—5.

9 De Conti Daniel Oliveira, Dias Ricardo Ribeiro, Fiorelli Alfredo Inácio, Stolf Noedir AG. Ruptured thoracic aortic aneurysm in patient with systemic lupus erythematosus. *Rev Bras Cir Cardiovasc* 2011;**26**:128—30.

10 Kunihara T, Sasaki S, Nishibe T, Akimaro Kudo F, Shiiya N, Murashita T, et al. Successful endovascular stent-grafting for thoracic aortic aneurysms in systemic lupus erythematosus. Report of 2 cases and review of the literature. *J Cardiovasc Surg (Torino)* 2002;**43**:235—40.

11 Core Lab Data. Ovation global pivotal trial. Data as of July 25, 2014.