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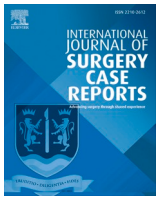

Case report

Ruptured abdominal aortic aneurysm in a young male patient, a rare case report

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ABSTRACT

Introduction and importance: Abdominal aortic aneurysm (AAA) is commonly a disease of the elderly population with an atherosclerotic aorta. We present a rare case scenario of a large ruptured AAA in a young patient.
Case presentation: A 32-year-old man presented to the Emergency Department with abdominal pain. On examination he had hypotension with a severely tender abdomen. Imaging revealed a ruptured 10 cm abdominal aortic aneurysm (AAA). He underwent an emergency open aneurysm repair and was discharged well on post-operative day 12. Apart from smoking, he had no known significant risk factors contributing to an AAA of such size. Clinical features and family history suggested a possible underlying connective tissue disorder.
Clinical discussion: A painful abdomen and hypotension in a young patient should prompt investigations to rule out a rare but life-threatening diagnosis of a ruptured AAA.
Conclusion: A possible underlying connective tissue disorder should be investigated for in any young patient presenting with an AAA.

1. Introduction

An abdominal aortic aneurysm (AAA) is defined as an aorta having a diameter of at least one and a-half times the normal diameter of the aorta at the level of the renal arteries [1]. Common risk factors for AAA include advancing age, smoking, hypercholesterolemia, hypertension and the male gender. AAAs are more common in patients with atherosclerosis, but other pathologies have been associated as well including connective tissue diseases such as Ehlers-Danlos syndrome and infective diseases such as syphilis or HIV [2,3].

This case report has been reported in accordance with the SCARE criteria [4].

2. Presentation of case

A 32-year-old Vietnamese man presented to the emergency department with sudden onset severe abdominal pain associated with syncope and hypotension (systolic blood pressure 50+). He was a smoker of 2-pack year history, an occasional drinker and worked as a technician in a factory. There was no change in bowel habit, no recent loss of appetite or loss of weight and he had never experienced abdominal pain like this before.

Examination revealed a severely tender and tense abdomen. A bedside ultrasound demonstrated free fluid, with a large 8x9cm AAA. A CT aortogram further confirmed tortuous and aneurysmal dilatation of the abdominal aorta measuring up to 10.8 × 10.8 cm in axial dimension and 24.1 cm in cranio-caudal length extending into the left common iliac artery, with the proximal aspect involving the origin of the bilateral renal arteries. A large retroperitoneal hematoma was also seen (Fig. 1a, b,c).

The patient underwent an emergency open abdominal aortic aneurysm repair (Fig. 2). The surgical procedure and overall management of patient was performed under the leadership of VVS, a Senior Consultant Vascular Surgeon with decades of experience in open and endovascular repair of Aortic emergencies. Access was difficult and due to the haemodynamic instability and the need for an expedited procedure, an aortic cross clamp was applied beneath the right renal artery but just proximal to left renal artery. This gave sufficient room for a bifurcated aortic graft to be anastomosed proximally without need for renal artery implantation and distally to the bilateral common iliac arteries.

An on table angiogram of the left lower limb was also performed as it appeared pale intraoperatively. This demonstrated an abrupt cut-off at the level of the left popliteal artery. A transverse arteriotomy was made over the left common femoral artery and a Fogarty balloon catheter was

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used to trawl out multiple thrombi. A final angiogram demonstrated in-line flow to the foot via the anterior tibial artery. Foot pulses were palpable and strong at the end of the procedure.

The patient was transferred to the intensive care unit (ICU), and was extubated on post-operative day (POD) 1. He was given intensive chest physiotherapy and incentive spirometry and started on nasogastric feeding on POD2, escalating to oral diet by POD 6. On the 4th day of his ICU stay he developed a fever, likely secondary to basal atelectasis. He was empirically started on a course of piperacillin-tazobactam, which was stopped 4 days later in view of a negative septic work up and down-trending inflammatory markers. His pro-calcitonin level was within normal limits. The surgical sites were healing well with no hematoma or signs of infection. During his ICU stay, the patient developed an initial acute kidney injury which was managed with proper hydration with normalising of his renal function. A transient liver enzyme rise was also noted which resolved gradually. His coagulation profile was preserved. By POD 12, he was clinically stable, independently mobile and fit for discharge from hospital. The patient was grateful to the surgical and intensive care unit for saving his life and taking good care of him.

Histopathological examination revealed degenerative changes of the aortic wall that was consistent with aortic rupture. However, no granulomas, giant cells, obliterative phlebitis or storiform fibrosis nor malignancy were identified. The appearance was also not suggestive of tuberculosis, vasculitic changes or IgG4 sclerosing disease. There was no evidence of any infective process in the aortic wall on microbiological investigation.

The etiology of the AAA was of interest due to its large size and his young age at presentation and he had multiple investigations while in hospital. However, soon after his discharge, he returned to Vietnam to be with his family and was lost to follow up before genetic testing could be arranged.

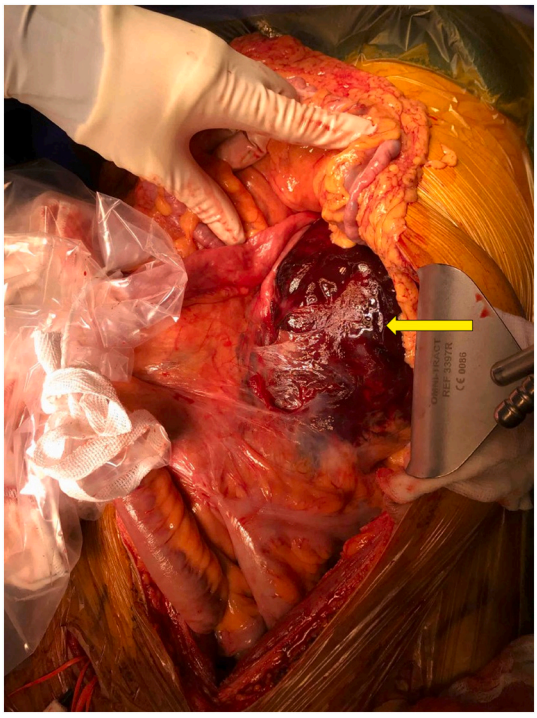

Fig. 2. Intraoperative picture demonstrating the aneurysmal dilatation of abdominal aortic aorta (yellow arrow). Bowel has been eviscerated into bowel bag to the patient's right and the retractor is on the left. Surgeon's hand is holding the transverse colon cranially. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

3. Discussion

Common causes of an acute abdomen with hypovolemic shock in a young patient usually include a bleeding intestinal ulcer or a ruptured ectopic pregnancy. A ruptured abdominal aortic aneurysm is a very rare contributing factor. However, anticipation with prompt diagnosis and treatment of such a grievous condition cannot be overemphasized.

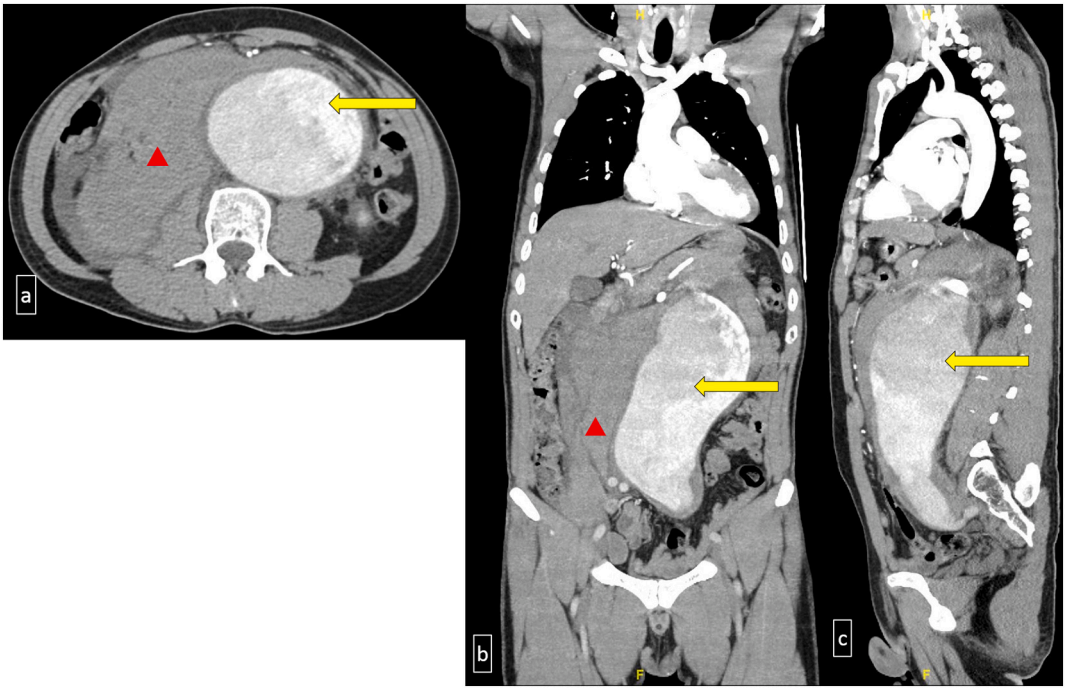

Fig. 1. CT aortogram (a. Axial section b. Coronal Section c. Parasagital section) demonstrating the large ruptured abdominal aortic aneurysm (10.8 × 10.8 × 24.1 cm) (yellow arrow) and the retroperitoneal haematoma (red arrowhead). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Aortic aneurysms have a strong association with atherosclerosis and arterial remodeling resulting in thinning of the tunica media and decrease in elastin and collagen [5]. Risk factors such as hypercholesterolemia, hypertension and smoking are commonly attributed to atherosclerosis. In addition, several connective tissue disorders have been implicated in aneurysms of the young population. Genetic association is noted mainly in proximal or thoracic aortic aneurysms of the young population and can be syndromic or sporadic [6–8]. Syndromic aortopathies are usually autosomal dominant disorders mainly due to defect in the extracellular matrix such as fibrillin-1 of elastin in Marfan syndrome (MFS) and type 3 collagen in vascular type (type IV) of Ehlers-Danlos syndrome (EDS), cytokine pathway overexpression of transforming growth factor beta (TGFB) pathway in Loeys-Dietz syndrome (LDS) or ciliopathy like autosomal dominant polycystic kidney disease. Whereas, non-syndromic aneurysms can be due to defects in neural crest migration (bicuspid aortic valves), smooth muscle contraction proteins (familial disorders) or those involving TGF-beta pathway resulting in Loeys-Dietz variants.

The probability of a connective tissue disorder with genetic predisposition is high in our patient. Early onset aortic aneurysms (<51 years) tend to be associated with smoking, other more commonly identifiable causes of aneurysms, be more symptomatic, more commonly proximal aorta (juxta renal, supra renal and thoraco- abdominal) and larger sized aneurysms (>6.9 cm at presentation) [9]. This is similar to the scenario of our patient who was 32 years old and a smoker, presenting acutely with abdominal pain and with a 10 cm juxta-renal aortic aneurysm.

The patient and his family originated from Nghe An in Central Vietnam. He was married with a healthy 5-year-old son. However, his mother had significant bilateral hallux valgus. He had 2 younger siblings, the youngest being a 25-year-old male with a history of pectus carinatum, which developed in his teenage years. Otherwise, there was no known cardiac, lung or rheumatological disease in the family. The patient was notably taller than his peers and stood at 179 cm tall. His body mass index was 24.2 kg/m^2. He had an average head width of 19 cm and head length of 22 cm, a normal arched palate and a triangular uvula centre. He did have a mild form of pectus excavatum. There was no obvious scoliosis or truncal striae. His fingers and toes were unusually long and slender without any deformities or positive thumb sign. He also had bilateral pes planus. (Fig. 3). Despite the lack of histological evidence, these clinical features point towards a connective tissue disorder.

There might be phenotypic overlap among patients with MFS, vascular type (type IV) EDS and LDS. However, a few distinctive features might help to differentiate them [10]. Our patient likely fits into LDS. Loeys-Dietz is a relatively new connective tissue disorder and is classically characterised by arterial tortuosity and aneurysm formation, hypertelorism and bifid uvula or cleft palate [11]. MFS mainly presents with lens dislocation, myopia and mitral valve prolapse whereas the vascular type of EDS presents with fragile skin and lax joints. Our patient displayed mild cranio-facial features, a tortuous aneurysmal aorta, arachnodactyly and pectus likely fitting into LDS type IV. He also developed keloid at the midline laparotomy wound which further supports a possible TGFB pathway abnormality as noted in LDS [12]. Our patient had a juxta-renal aortic aneurysm supporting the proximal aortic aneurysms seen in connective tissue disorders. Type IV EDS presents with loss of tensile strength, vascular fragility and brittle tissue and is usually associated with poor operative outcome as compared to LDS [11]. Our patient also survived the acute contained rupture of his large juxta-renal aortic aneurysm favouring LDS over EDS.

Our patient presented with very high C-reactive protein (241.6 mg/L) that remained elevated on discharge (71.6 mg/L). His erythrocyte sedimentation rate was also elevated at 35 mm/h. The elevated CRP reflects the severe acute systemic inflammatory state relating to the aortic wall degeneration [[13,14]] with associated poor prognosis [15]. AAAs are commonly associated with atherosclerosis and inflammation while thoracic aortic aneurysms are mainly hereditary without significant atheroma [8]. About 20% of thoracic aortic aneurysms or dissections display an autosomal dominant pattern of inheritance while abdominal aortic aneurysms usually do not [16]. However, our patient had no evidence of atheroma on imaging and had normal serum cholesterol. Also, normal homocysteine level precluded the chances of precocious atherosclerosis and aneurysmal dilatation of aorta.

Young-onset aortic aneurysms are also noted in rheumatological conditions such as systemic lupus erythematosus [17]. Our patient lacked the tell-tale clinical features of rheumatological disorders such as dry eyes, xerostomia, abnormal hair loss or rashes. The histopathological examination of the aortic wall was also unremarkable for any vasculitis.

We explained to the patient multiple times about the need for genetic testing for prognostication and the possible need to screen family members. We also offered to provide free genetic testing. However, the patient was lost to follow up before this could be arranged. Echocardiography was also not performed that would help to characterize any concomitant aortic root or cardiac involvement.

4. Conclusion

We present a rare case of a ruptured AAA in a young man with likely connective tissue disorder. He was successfully treated and discharged with good post-operative outcomes. It is imperative to consider the rare

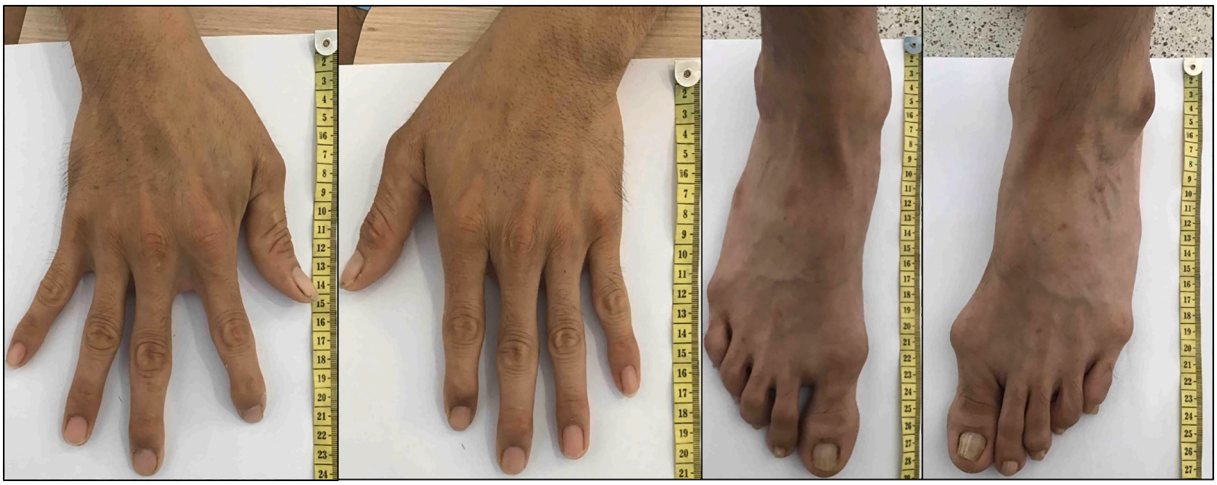

Fig. 3. Photos of the right and left hands and feet of the patient demonstrating arachnodactyly and pes planus.

but life-threatening diagnosis of AAA even in a young patient presenting with abdominal pain and hypotension. It is also essential to evaluate the underlying causal factors of an AAA in such patients for directed treatment and surveillance.

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Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy if the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration (for case reports detailing a new surgical technique or new equipment/technology)

Not applicable.

Guarantor

Dr. Kush Raj Lohani, Dr. Geraldine Wong Yan Xin, Dr. Jiaqian Cui, Dr. Vikram Vijayan Sannasi

Provenance and peer review

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CRediT authorship contribution statement

1. Dr. Kush Raj Lohani: Writing – original manuscript draft, review and editing.
2. Dr. Geraldine Wong Yan Xin: Writing – original manuscript draft, review and editing.
3. Dr. Jiaqian Cui: Writing – original manuscript draft, review and editing.
4. Dr. Vikram Vijayan Sannasi: Writing – Final review and editing.

Declaration of competing interest

None.

References

[1] A.T. Hirsch, Z.J. Haskal, N.R. Hertzer, C.W. Bakal, M.A. Creager, J.L. Halperin, et al., ACC/AHA 2005 practice guidelines for the management of patients with peripheral arterial disease (lower extremity, renal, mesenteric, and abdominal aortic): a collaborative report from the american Association for Vascular Surgery/Society for Vascular Surgery, Society for Cardiovascular Angiography and Interventions, Society for Vascular Medicine and Biology, Society of Interventional Radiology, and the ACC/AHA task force on practice guidelines (Writing committee to develop guidelines for the Management of Patients with Peripheral Arterial Disease): endorsed by the american Association of Cardiovascular and Pulmonary Rehabilitation; National Heart, Lung, and Blood Institute; Society for Vascular Nursing; TransAtlantic inter-society consensus; and vascular disease foundation, Circulation 113 (11) (2006 Mar 21) e463–e654.

[2] P.M. Shaw, J. Loree, R.C. Gibbons, Abdominal aortic aneurysm [cited 2021 Sep 6]. Available from:, in: StatPearls [Internet], StatPearls Publishing, Treasure Island (FL), 2021 http://www.ncbi.nlm.nih.gov/books/NBK470237/.

[3] K. Singh, K.H. Bønaa, B.K. Jacobsen, L. Bjørk, S. Solberg, Prevalence of and risk factors for abdominal aortic aneurysms in a population-based study: the Tromsø study, Am. J. Epidemiol. 154 (3) (2001 Aug 1) 236–244.

[4] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230. Dec.

[5] J. Golledge, G.-P. Shi, P. Norman, Pathogenesis of aortic aneurysms [cited 2021 Sep 6]. Available from:, in: R. Fitridge, M. Thompson (Eds.), Mechanisms of Vascular Disease: A Reference Book for Vascular Specialists [Internet], University of Adelaide Press, Adelaide (AU), 2011 http://www.ncbi.nlm.nih.gov/books/NBK534278/.

[6] The Genetics of Aortic Aneurysms [Internet], American College of Cardiology, Available from: https://www.acc.org/latest-in-cardiology/articles/2018/05/02/12/52/http%3a%2f%2fwww.acc.org%2flatest-in-cardiology%2farticles%2f2018%2f05%2f02%2f12%2f52%2fthe-genetics-of-aortic-aneurysms, 2021.

[7] I. Luyckx, B.L. Loeys, Curriculum topic: disease of the aorta and trauma to the aorta and heart. The genetic architecture of non-syndromic thoracic aortic aneurysm, Heart Br. Card. Soc. 101 (20) (2015 Oct) 1678–1684.

[8] M.E. Lindsay, H.C. Dietz, Lessons on the pathogenesis of aneurysm from heritable conditions, Nature 473 (7347) (2011 May 19) 308–316.

[9] S.C. Muluk, J.P. Gertler, D.C. Brewster, R.P. Cambria, G.M. LaMuraglia, A. C. Moncure, et al., Presentation and patterns of aortic aneurysms in young patients, J. Vasc. Surg. 20 (6) (1994 Dec) 880–886, discussion 887–888.

[10] M. Murphy-Ryan, A. Psychogios, N.M. Lindor, Hereditary disorders of connective tissue: a guide to the emerging differential diagnosis, Genet. Med. Off. J. Am. Coll. Med. Genet. 12 (6) (2010 Jun) 344–354.

[11] B.L. Loeys, U. Schwarze, T. Holm, B.L. Callewaert, G.H. Thomas, H. Pannu, et al., Aneurysm syndromes caused by mutations in the TGF-beta receptor, N. Engl. J. Med. 355 (8) (2006 Aug 24) 788–798.

[12] J. Jagadeesan, A. Bayat, Transforming growth factor beta (TGFbeta) and keloid disease, Int. J. Surg. Lond. Engl. 5 (4) (2007 Aug) 278–285.

[13] J. De Haro, F. Acin, S. Bleda, C. Varela, F.J. Medina, L. Esparza, Prediction of asymptomatic abdominal aortic aneurysm expansion by means of rate of variation of C-reactive protein plasma levels, J. Vasc. Surg. 56 (1) (2012 Jul) 45–52.

[14] E.N. Kim, J. Yu, J.S. Lim, H. Jeong, C.J. Kim, J.-S. Choi, et al., CRP immunodeposition and proteomic analysis in abdominal aortic aneurysm, PLoS ONE 16 (8) (2021 Aug 24), e0245361.

[15] M. Schillinger, H. Domanovits, K. Bayegan, T. Hölzenbein, M. Grabenwöger, J. Thoenissen, et al., C-reactive protein and mortality in patients with acute aortic disease, Intensive Care Med. 28 (6) (2002 Jun) 740–745.

[16] A. Pinard, G.T. Jones, D.M. Milewicz, Genetics of thoracic and abdominal aortic diseases, Circ. Res. 124 (4) (2019 Feb 15) 588–606.

[17] D. Noorvash, K. King, M. Gebrael, Two cases of catastrophic AAA rupture in young women with systemic lupus erythematosus, Case Rep. Emerg. Med. 2018 (2018), 1049568.