CASE REPORT

True Saccular Aneurysm at the Iliac Bifurcation in an Adolescent: Case Report and Review of Lower Limb Vascular Embryology

Samuel C.J. Hawthorne *, Noel R. Atkinson, Korana Musicki
Department of Vascular Surgery, Royal Melbourne Hospital, Parkville, Victoria, Australia

Introduction: Isolated iliac artery aneurysms are an uncommon occurrence in the absence of concurrent aortic disease in the adult population and are a rare entity in children and adolescents. Paediatric patients may present with false aneurysms less frequently but true aneurysms are exceptional. In this report, the case of an iliac bifurcation true saccular aneurysm is described.

Report: An 18 year old woman without history of infection, trauma, connective tissue disorders, or vasculitis, was referred with an incidental left iliac bifurcation saccular aneurysm. She underwent open surgical resection of the aneurysm with primary re-anastomosis of the common to external iliac arteries and ligation of the internal iliac artery. Histopathological assessment did not show any inflammatory or other underlying disease process.

Discussion: A case is presented of an isolated iliac bifurcation true aneurysm in an adolescent and its successful treatment. It is plausible that incomplete involution of the embryologically dominant sciatic artery may have been the cause for this presentation and for other congenital iliac artery aneurysms. Literature review of other paediatric iliac aneurysms shows an array of postulated underlying causes and treatment strategies.

INTRODUCTION

Iliac aneurysmal disease is most commonly an extension of degenerative abdominal aortic aneurysmal disease, occurring in adult populations with vascular risk factors. Isolated iliac artery aneurysms account for <2% of abdominal aneurysms and approximately 0.03% of all aneurysms.¹ The most common presentation of an isolated iliac true aneurysm is degenerative in an elderly male. Paediatric presentations are even less common and are typically false aneurysms resulting from trauma or infection, although connective tissue, vasculitis, or congenital disorders may be present. This case report presents an adolescent patient with a saccular iliac artery true aneurysm without predisposing risk factors and proposes a cause through the vascular ontogeny of the lower limb. The literature on similar paediatric iliac aneurysms is also presented, which has not previously indicated this as a postulated mechanism.

CASE PRESENTATION

An 18 year old woman was referred with an incidental left iliac bifurcation aneurysm measuring 26 mm in diameter on pelvic ultrasound during investigation for oligomenorrhoea. She had no personal or family history of vasculitis or connective tissue disorders, and had no history of trauma, surgery, or infection in the pelvis. At initial presentation, she denied pain or systemic symptoms, had no syndromic features nor deformities or asymmetry between the limbs, and had all lower limb pulses present. She failed to attend review for a six month interval during which she had a levonorgestral intrauterine device implanted.

She re-presented urgently with left iliac fossa pain which settled over the next 48 hours. She remained afebrile and haemodynamically stable, with normal full blood count, biochemistry, and urinalysis. Pelvic ultrasound and computed tomography (CT) angiogram showed an unruptured and uninflamed 28 mm saccular aneurysm projecting inferomedially from the iliac bifurcation into the internal iliac artery with no thrombus, and preservation of external and internal iliac arteries measuring 7 mm in diameter (Fig. 1).

The iliac aneurysm saccular anatomy at four times the size of the native vessels, in addition to uncertain hormonal influences from the progesterone secreting device or a...
future pregnancy, weighed in favour of timely prophylactic repair. Under general anaesthesia, a Pfannenstiel incision was made, and the retroperitoneal plane was entered on the left. The left common, external, and internal iliac arteries were isolated to allow for full exposure of the aneurysm, which was found to involve predominantly the internal iliac artery origin (Fig. 2) and was clear of adjacent tissue. Following administration of 70 IU/kg of heparin, the vessels were clamped and the iliac aneurysm was resected. The common and external iliac arteries were primarily re-anastomosed with continuous 6/0 polypropylene. The internal iliac artery was suture ligated with 5/0 polypropylene when found to be short in length, small in calibre, and with excellent back bleeding. The wound was closed conventionally in layers.

The resected aneurysm tissue was analysed. Microscopy and culture detected no organisms. Histopathological assessment showed a smooth endothelial surface with focal intramural myxoid changes and fibrosis, with no evidence of active inflammatory processes or connective tissue disease.

After an uneventful recovery, the patient was discharged on day five post-operatively on aspirin 100 mg daily for 12 months. Left buttock claudication was not initially experienced because of limited physical activity during the COVID-19 pandemic lockdown; however, this was recognised at six month follow up and successfully managed with a dedicated exercise regimen. The left common and external iliac arteries remained patent with an antegrade triphasic waveform on duplex ultrasound, and the left internal iliac artery stump was also noted to have a retrograde triphasic waveform, indicating excellent quality collateral arterial supply (Fig. 3). The patient was discharged after five reviews over 12 months as there was neither graft nor residual aneurysm in situ for surveillance.

DISCUSSION

Iliac artery aneurysms are uncommon in a paediatric or young adult population. When present, they are frequently attributable to infection, trauma, arteritis, or connective tissue disorders such as Marfan's disease. Congenital idiopathic development can account for approximately half of this subset of aneurysms, and may better explain the aetiology of the aneurysm discussed in this report.

By the fourth week of foetal growth, the origin of the lower limb can be seen through the development of primitive limb buds from the lateral plate mesoderm. Through a fusion of the proximal portion of the umbilical and the fifth lumbar intersegmental arteries, the common iliac artery is formed. The umbilical artery first forms the presumptive internal iliac artery, which gives rise to the sciatic/ischiadic artery; this is the initial axial artery running down the centre of the developing lower limb and closely following its namesake sciatic nerve. The umbilical artery subsequently forms the external iliac artery, which gives rise to the femoral artery progressively along the superficial anterior aspect of the thigh. Following a series of anastomoses between the primitive femoral and sciatic arteries, the internal iliac fed sciatic artery gradually is remodelled and resorbed, eventually leaving the external iliac fed femoral artery as the dominant vessel to the lower limb. The involution of the sciatic artery goes on to leave the popliteal and peroneal/fibular arteries as the only residual axial supply to the lower limb.

Rarely, the sciatic artery may fail to regress and remain as the dominant supply to the lower limb post-natally. Anatomical variation with persisting sciatic arteries (PSAs) has been documented since the early twentieth century, with many more contemporary reports showing the breadth of presentations. While PSAs are reported in 0.03%—0.06% of the population, 48% of them become aneurysmal. Hypotheses of the mechanism suggest overstretching of less elastic material within the vessel wall. The histopathology from the present patient did not show wall abnormalities consistent with this hypothesis, and in the absence of a PSA it is unlikely to have been a result of stretching.
The precise manner in which the present patient developed her aneurysm is not clear. Given the proximity of the disease to the presumptive regression point of the sciatic artery, it is plausible to suggest an incomplete involution of the sciatic artery consistent with a Pillet type 3 or an Ahn-Min class IIA PSA led to the formation of this saccular aneurysm. An alternative diagnosis could be from infection, as there have been case studies of mycotic iliac aneurysm formation following umbilical artery cannulation; however, there was no history of intervention for the present patient.

Because of the rarity of aneurysmal disease in the pediatric and young adult setting, there are limited specific guidelines on their management. The European Society for Vascular Surgery recommends treatment of asymptomatic iliac artery aneurysms over a threshold of 35mm, but this is largely for more common degenerative aneurysms rather than a congenital one. The cumulative risk of aneurysm rupture over the lifespan of younger patients should encourage a lower threshold for intervention than would otherwise be considered. Pregnancy in particular may confer added risk in the setting of known iliac and aneurysmal disease; spontaneous rupture of the iliac arteries has been reported both with and without aneurysmal disease present.

A literature search of the PubMed database was consequently conducted to assess for other similar cases, using the search terms “iliac artery aneurysm” and “isolated iliac artery aneurysm” in conjunction with the terms “adolescent”, “teenage”, “young adult”, or “paediatric”. Papers were included if they were published in English, the patient age could be identified as younger than 20 years old, involved a true aneurysm, and involved isolated iliac aneurysms. The reference list from any paper meeting the inclusion criteria was manually reviewed for additional relevant literature.

A total of 23 publications were identified meeting the inclusion criteria from the past century, accounting for 25 patients with isolated iliac artery aneurysms (Table 1, Supplementary references 1-23). All were case reports, bar one retrospective case series which identified three cases over 32 years, of which only one was saccular. Of all the cases published, 12 were deemed to be of either idiopathic or congenital aetiology, with the most common cause identified overall as mycotic growth secondary to infection from either umbilical artery catheterisation as a neonate or bacteraemia/endocarditis.

Within a paediatric population, the recommendation is for resection with or without direct vessel repair where possible to allow for growth as the patient approaches maturity. Reconstruction with vein grafts can be limited by aneurysmal degeneration over time, while use of synthetic grafts can pose infection risk and patency issues, both ideally requiring lifelong surveillance to ensure problems are identified early. Despite this, five of the identified cases used Dacron conduit for their repair; however, there was...
Table 1. Compilation of all published cases of isolated iliac artery aneurysms in young patients. References included in supplementary data

| Case                        | Age – y/ gender | Location and size | Histopathology                                                                 | Aetiology                     | Treatment                                      |
|-----------------------------|-----------------|-------------------|--------------------------------------------------------------------------------|-------------------------------|-----------------------------------------------|
| MacLaren 1913               | 18F             | L IIA, unknown    |                                                                                | Rheumatism                    | Blind ligation                                |
| Fenn and Musgrove 1958      | 6M              | L CIA, saccular   | “size of a large hen’s egg”                                                   | Congenital weakness           | Aneurysmoplasty                               |
| Fays and Bretagne 1980      | 3M              | R CIA, 10 × 15 cm | saccular                                                                      | Microcalcification in fibrous tissue without elastic fibres | Excision and Dacron interposition graft       |
| Gronemeyer and deMello 1982 | 0.6M            | R CIA, bilobe     | saccular                                                                      | Inflammatory infiltrate through wall, with granulation tissue in adventitia, vasa vasoarum thickening | Nil (post mortem finding with iliocaval fistula) |
| Todd et al. 1984            | 0.5U            | L CIA, fusiform   | unknown size                                                                  | True aneurysm                 | Aneurysmoplasty                               |
| Villani, Leoni and Mora 1985| 7F              | L EIA, fusiform   | 3 cm                                                                           | Congenital                    | Excision and Dacron interposition graft       |
| Drucker et al. 1986         | 2M              | R C/EIA, fusiform | 5 × 6 cm                                                                      | Intimal absence, fibroblastic proliferation, diffuse calcifications       | Excision, IIA ligation, CIA-EIA anastomosis  |
| Moritz 1986                 | 2.5F            | L CIA and IIA,    | fusiform 6 × 3 cm                                                             | Thinned wall with irregular intimal fibrolyplasia, elastic membrane degeneration | Excision, aorto-EIA anastomosis, IIA ligation |
| Sarkar et al., 1991         | 4F              | R CIA, saccular   | 6 cm                                                                           | Idiopathic                    | Excision, cross over ilio-iliac bypass graft  |
| Lucas et al. 1994           | 9F              | L CIA, saccular   | 8 cm                                                                           | Mycotic, secondary to UAC     | Excision, CIA-EIA anastomosis                 |
| Taketani et al. 1997        | 3M              | L EIA, 6.7 cm     |                                                                                | Idiopathic                    | Excision, Dacron interposition graft          |
| Zimmermann et al. 2009      | 11F             | L CIA, occluded   | fusiform 4.2 cm                                                                | Degenerative vessel wall with calcifications, culture negative             | Excision, reversed fusiform vein and GSV spliced interposition graft |
| de Figueiredo Borges et al. 2010 | 0.16M         | R CIA, fusiform   | 4 cm                                                                           | Thinned arterial wall with disorganised collagen, limited collagen and lamellae | Menkes disease                                |
| Chithra et al. 2013         | 3M              | R CIA, fusiform   | 4.2 × 3.2 cm                                                                  | True aneurysm with calcification, no evidence of vasculitis, culture negative | Excision, reversed femoral vein interposition graft (L IIA ligation, R IIA preserved) |
| Davis et al. 2016           | 1. 0.2F         | R CIA, fusiform   | 0.5 cm; L IIA, fusiform 3.8 cm                                                | 1. Inflammation and fibrosis of arterial wall | 1. Plication of R CIA, bilateral IIA ligation |
|                             | 2. 2M           | L CIA, fusiform   | 0.4 cm; L CIA, fusiform 3.8 cm                                                | 2. Mural fibrosis with dystrophic calcification and chronic inflammation | 2. Excision, direct re-anastomosis             |
|                             | 3. 4F           | L CIA, fusiform   | 2.5 cm; R CIA, fusiform 3.8 cm                                                | 3. Medial thinning with intimal hyperplasia                                | 3. Excision, L IIA to R EIA anastomosis         |
| Lee et al. 2016             | 4F              | R ili a with agenesis of R EIA, saccular 5.2 cm | True aneurysm with intimal fibroplasia, culture negative | Idiopathic-congenital          | Direct excision                                |
inadequate follow up data published to assess their long term outcomes. In an adult, the option for reconstruction via an endovascular approach is available but does not have the long term data to support its use in a paediatric setting; four endovascular approaches have been published, of which three used coil embolisation, two in conjunction with covered stent exclusion. These methods may prove viable; however, their publications have only been made within the last two years and may require closer follow up to establish their safety profile and durability.

**Conclusion**

Although a rare clinical presentation, adolescent aneurysm disease necessitates treatment because of a cumulative risk of rupture over the course of the patient’s lifespan. In the absence of predisposing factors, a congenital cause through incomplete sciatic artery involution could be considered. Where intervention is indicated, the present authors would prefer direct resection and artery to artery reconstruction whenever possible. Although endovascular management may be an appropriate choice for an older patient, the durability has not been tested in the paediatric population. The need for lifelong surveillance if an endoluminal or open graft option is chosen should also be considered.

**Table 1—continued**

| Case                        | Age — y/ gender | Location and size | Histopathology | Aetiology | Treatment                                                                 |
|-----------------------------|-----------------|------------------|----------------|-----------|---------------------------------------------------------------------------|
| Hoshiko et al. 2017         | 12F             | R CIA, fusiform 9.3 cm | N/A, Endovascular treatment | Congenital idiopathic | Aneurysmotomy, haemostatic ligation of feeding vessel ostia, 
aorto-bifemoral PTFE bifurcated graft repair |
| Krysiak et al. 2019         | 0.1F            | R CIA, saccular 4.5 × 3.7 cm | N/A, Endovascular treatment | Congenital idiopathic | Unsuccessful thrombin injection, successful coil exclusion |
| Ng et al. 2019              | 0.15M           | R IIA, size not specified | Focal intimal fibroplasia with fibromyxoid matrix deposition | Menkes disease | Ligation† |
| Zaidan et al. 2019          | 9F              | R CIA, saccular 8.3 × 5.7 cm | True aneurysm with intimal fibroplasia and calcification, no evidence of vasculitis | Idiopathic | Excision, Dacron CIA to EIA interposition graft, ligation of IIA |
| Başpınar et al. 2020        | 2F              | L CIA-EIA, fusiform 2.6 × 3.6 cm | N/A, Endovascular treatment | * | Covered stent exclusion with IIA coil embolisation |
| Jevalikar et al. 2021       | 8M              | R CIA, fusiform, size not specified | N/A, Endovascular treatment | Mucormycosis | Covered stent exclusion with IIA coil embolisation |
| Sunil, Almanan, and Ismazizi 2021 | 12M             | R CIA, saccular, size not reported | N/A, Endovascular treatment | Mycotic, post-infective endocarditis three years prior | Covered stent exclusion across CIA-EIA |

CIA = common iliac artery; EIA = external iliac artery; IIA = internal iliac artery; N/A = not applicable; UAC = umbilical artery catheter; R = right; L = left; F = female; M = male; PTFE = polytetrafluoroethylene.

* Not discussed in paper.
† Graft material not specified.
‡ Surgery occurred in setting of rupture, sustaining fatal injury.

**FUNDING**

None.

**CONFLICT OF INTEREST**

None.

**APPENDIX A. SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at https://doi.org/10.1016/j.ejvsf.2021.10.016.

**REFERENCES**

1. Brunkwall J, Hauksson H, Bengtsson H, Bergqvist D, Takolander R, Bergentz S-E. Solitary aneurysms of the iliac arterial system: an estimate of their frequency of occurrence. *J Vasc Surg* 1989;10:381–4.
2. Sarkar R, Coran AG, Cilley RE, Lindenauer SM, Stanley JC. Arterial aneurysms in children: clinicopathologic classification. *J Vasc Surg* 1991;13:47–57.
3. Davis FM, Eliason JL, Ganesh SK, Blatt NB, Stanley JC, Coleman DM, et al. Pediatric nonaortic arterial aneurysms. *J Vasc Surg* 2016;63:466–76.
4. Sadler TW. *Langman’s medical embryology*. 13th ed. Philadelphia, Pennsylvania: Wolters Kluwer Health; 2015.
5. DeSesso JM. Vascular ontogeny within selected thoracoabdominal organs and the limbs. *Reprod Toxicol* 2017;70:3–20.
van Hooft IM, Zeebrugts CJ, van Sterkenburg SMM, de Vries WR, Reijnen MMPJ. The persistent sciatic artery. *Eur J Vasc Endovasc Surg* 2009;37:585—91.

Powell BC, Webb KM, Freeman BM, Carsten CG, Gandhi SS. Ligation of common iliac artery mycotic aneurysm in a neonate and review of the literature. *Ann Vasc Surg* 2019;59:312.e1—5.

Wanhainen A, Verzini F, Van Herzeel I, Allaire E, Bown M, Cohnert T, et al. Editor’s choice — European society for vascular surgery (ESVS) 2019 clinical practice guidelines on the management of abdominal aorta-iliac artery aneurysms. *Eur J Vasc Endovasc Surg* 2019;57:8—93.

Butorac D, Djaković I, Košec V, Kopjar M, Kuna K. Spontaneous rupture of internal iliac artery in pregnancy: case report. *Acta Clin Croat* 2018;57:157—60.

Peter SDS, Ostlie DJ. A review of vascular surgery in the pediatric population. *Pediatr Surg Int* 2007;23:1—10.