CASE REPORT

Congenital Abdominal Aortic Aneurysm in a Four Year Old Girl

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Introduction: Abdominal aortic aneurysm (AAA) in neonates, infants, and children is uncommon, usually occurring as a result of infections, connective tissue disorders, vasculitis, or iatrogenic trauma. A case of idiopathic congenital AAA, an extremely rare disease of unknown origin, is described.

Report: In March 2018, a 40 mm hypoechoic mass adjacent to the left kidney was detected incidentally by abdominal ultrasound for investigation of hypercalciuria in a four year old girl. Contrast enhanced computed tomography (CT) revealed an infrarenal fusiform AAA measuring 39 mm in maximum diameter, a 15 mm left renal artery aneurysm, a 14 mm right hypogastric artery aneurysm, and a 12 mm left hypogastric artery aneurysm. Cerebral magnetic resonance imaging revealed multiple intracranial aneurysms between 8 and 15 mm in diameter. Considering the size of the AAA and risk of rupture, surgical repair was planned. In May 2018, the congenital AAA was successfully repaired with a 10 mm Dacron aorto-aortic tube graft. Increases in the size of the left renal artery aneurysm and a left middle meningeal artery aneurysm were detected 12 and 14 months post-operatively, respectively. Coil embolisations were performed. An intracranial dural arteriovenous fistula (AVF) was discovered incidentally by cerebral angiography for treatment of the left middle meningeal artery aneurysm. Transarterial embolisation for AVF was also performed. At the 21 month post-operative follow up, the patient is doing well, and the untreated aneurysms have not grown.

Conclusion: Long term outcomes after surgical repair for congenital AAA are unclear. Moreover, growth of residual aneurysms was detected post-operatively, so follow up with frequent multimodality imaging for multiple aneurysms is necessary.

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Keywords: Aortic aneurysm, Aortic diseases

INTRODUCTION

Abdominal aortic aneurysm (AAA) in neonates, infants, and children is uncommon and usually caused by bacterial, tuberculous or fungal infection; congenital connective tissue disorders (Marfan syndrome, Ehlers–Danlos syndrome, or Loeys–Dietz syndrome), vasculitis (Takayasu arteritis, Kawasaki syndrome, or polyarteritis nodosa); or trauma from umbilical artery catheterisation.1 Idiopathic congenital AAA (CAAA) is an extremely rare disease of unknown origin.

A patient diagnosed with an idiopathic CAAA with multiple aneurysms is described.

CASE REPORT

In March 2018, a 40 mm hypoechoic mass adjacent to the left kidney was detected incidentally by abdominal ultrasound in a four year old girl with a two year history of cataract, hypercalciuria, and proteinuria. It was initially suspected to be a cystic lesion. Two months later, contrast enhanced computed tomography (CT) revealed an infrarenal fusiform AAA measuring 39 mm in maximum diameter, a 15 mm left renal artery aneurysm, a 14 mm right hypogastric artery aneurysm, and a 12 mm left hypogastric artery aneurysm (Fig. 1A). She had no family history of aneurysmal disease or connective tissue disorders. Her blood pressure was 112/63 mmHg. Clinical examination was unremarkable except for a palpable pulsatile abdominal mass. Cerebral magnetic resonance imaging (MRI) revealed multiple intracranial aneurysms.
between 8 and 15 mm in the intracavernous portion of the right internal carotid artery and bilateral middle meningeal arteries (Fig. 1B). The patient was referred for further management of the multiple aneurysms. Neurosurgeons considered that surgery or coil embolisation for the multiple cerebral aneurysms carried a high risk of intra-operative rupture. The maximum diameter of the AAA was more than four times the diameter of the normal abdominal aorta. Considering the size and risk of AAA rupture, surgical repair was planned. Regarding the other aneurysms, it was considered that revascularisation was extremely difficult because the normal diameter of the vessels was too small. Therefore, conservative management was planned and angiotensin II receptor blocker and beta blocker administration was initiated to prevent hypertension.

During the operation (May 2018), the AAA was exposed through a midline incision. The diameter of the infrarenal AAA was 41 mm, and it extended to the aortic bifurcation (Fig. 2A). After heparin infusion at 80 U/kg, the aorta and both common iliac arteries were cross clamped. The aortic aneurysm was opened, and a 10 mm Dacron aorto-aortic tube graft was anastomosed proximally to the infrarenal abdominal aorta and distally to the aortic bifurcation with a running 4-0 Prolene suture. Reconstruction of the inferior mesenteric artery was performed with a running 7-0 Prolene suture considering the risk of intestinal ischaemia following possible subsequent treatment of bilateral hypogastric artery aneurysms (Fig. 2B). A histological examination of the aneurysmal wall revealed thinning of the media with irregularity and fragmentation of the elastic lamina. There was no evidence of cystic degeneration of the media, active aortitis, or infection. These changes in the media were considered the cause of aneurysmal formation. Additionally, diffuse fibro-elastic thickening of the intima, chronic media dissection, slight chronic inflammatory infiltrate, adventitial fibrosis, and dilatation of the vasa vasorum, which were considered secondary changes, were observed (Fig. 3). Whole exome sequencing showed no genetic mutation of the known congenital connective tissue disorders associated with aneurysmal diseases. Contrast

Figure 1. Preoperative images of the abdominal aortic aneurysm and multiple aneurysms. A. Three-dimensional reconstruction of preoperative computed tomography angiography image showing an infrarenal abdominal aortic aneurysm with a maximum aortic diameter of 39 mm, a left renal artery aneurysm, and bilateral internal iliac artery aneurysms. B. Three-dimensional reconstruction cerebral magnetic resonance image showing multiple intracranial aneurysms.

Figure 2. Operative findings. A. Operative photograph showing an infrarenal abdominal aortic aneurysm with a maximum aortic diameter of 41 mm. B. Operative photograph of implanted 10-mm Dacron graft in the infrarenal aortic position.
enhanced CT on post-operative day seven confirmed complete exclusion of the AAA with patency of the graft and reconstructed inferior mesenteric artery (Fig. 4). Aspirin administration was initiated for post-operative graft thromboprophylaxis. At the 12 month post-operative follow up, abdominal MRI revealed an increase of the left renal artery aneurysm from 15 mm to 25 mm (Fig. 5A). Coil embolisation was therefore performed successfully for this aneurysm. Moreover, cerebral MRI revealed an increase of a left middle meningeal artery aneurysm from 15 mm to 19 mm (Fig. 5B) at the 14 month post-operative follow up, for which coil embolisation was also performed successfully. Simultaneously, a superior sagittal sinus dural arteriovenous fistula (AVF) was discovered incidentally on cerebral angiography (Fig. 5C), and transarterial embolisation with N-butyl-2-cyanoacrylate for AVF was also performed. Post-procedure MRI showed disappearance of blood flow into the enlarged aneurysms and decrease of abnormal blood flow into the AVF. At the 21 month post-operative follow up, the patient is doing well, and the untreated aneurysms have not increased in size, as seen on follow up imaging studies.

Figure 3. Pathological findings in the surgically resected abdominal aorta. A. The image shows irregularity and focal loss of the elastic lamina of the media, fibroelastic intimal thickening, and dilatation of the vasa vasorum (arrows). The upper portion of the media shows chronic dissection (asterisk) surrounded by thick elastic tissue (elastica van Gieson stain). B. The high-magnification image of the media (square region of A) shows fragmentation of elastic fibers (elastica van Gieson stain).

Figure 4. Contrast-enhanced computed tomography on post-operative day 7. Three-dimensional reconstruction of post-operative computed tomography angiography showing patency of the graft and reconstructed internal mesenteric artery.

Consent

Written consent for publication of this article was obtained from the parents of the patient.
**Figure 5.** Postoperative follow-up imaging. A: Magnetic resonance image at the 12-month follow-up showing an increase in size of the left renal artery aneurysm from 15 mm to 25 mm. B: Magnetic resonance image at the 14-month follow-up showing an increase in size of the left middle meningeal artery aneurysm from 15 mm to 19 mm. C: Cerebral angiography showing superior sagittal sinus dural arteriovenous fistula.

**Table 1.** Previously reported cases of congenital abdominal aortic aneurysms.

| Author        | Gender | Age at discovery | Location   | Other aneurysms | Symptoms related to aneurysms | Surgical treatment          | Outcome                                    |
|---------------|--------|------------------|------------|-----------------|-------------------------------|-----------------------------|-------------------------------------------|
| 1 Howorth Jr MB | Female | 1 day            | Infrarenal | None            | Large abdominal mass, vomiting, anorexia | Exploratory laparotomy     | Rupture and death during Operation        |
| 2 Darden WA    | Male   | 2.5 years        | Infrarenal | None            | None                          | Dacron aortic graft        | Died of Pneumonitis at 5 months after surgery |
| 3 Sterpetti AV | Male   | 19 years         | Infrarenal | None            | Middle epigastric pain, abdominal fullness, dysuria, abdominal pulsatile mass | Dacron aortic graft 18 mm | Healthy at 9 years after surgery          |
| 4 Odagiri S    | Male   | 1 year           | Infrarenal | Multiple left renal artery aneurysms, bilateral common iliac artery aneurysms | None                        | Dacron aortic graft 12 mm | Healthy at 10 months after surgery        |
| 5 Latter D     | Male   | 1 month          | Infrarenal | None            | Pulsatle abdominal mass       | Polytetrafluoroethylene tube graft 8 mm | Healthy at 10 months after surgery         |
| 6 Saad SA      | Male   | 6 weeks          | Infrarenal | Left common iliac artery aneurysm mass | Pulsatle abdominal mass | Aneurysmorrhaphy           | Healthy at 3 months after surgery         |
| 7 Myrmel T     | Male   | 30 years         | Infrarenal | None            | Pulsatle abdominal mass, acute abdominal pain | Albumin coated USCI graft sized 16×8 mm | Healthy at 1 year after surgery          |
| 8 Malee MP     | Female | 32 weeks’ gestation | Juxtarenal | Aneurysmal dilation of the bilateral iliac artery (details unknown) | Palpable abdominal mass, ileus compression from an aneurysm | None                        | Died of acute pulmonary hypertension and cardiac dysfunction at age 9 days |
| Author  | Gender | Age at discovery | Location | Other aneurysms | Symptoms related to aneurysms | Surgical treatment | Outcome |
|---------|--------|------------------|----------|----------------|-------------------------------|-------------------|---------|
| Kim ES  | Female | 9 days           | Juxtarenal| None           | None                          | None              | Died of heart failure secondary to renovascular hypertension at age 20 days |
| Mehall JR | Male   | 6 weeks          | Juxtarenal| Right common iliac artery aneurysm | None                | Bifurcated Gore-Tex graft 7-4 mm | Healthy at 1 month after surgery |
| Laing AJ | Male   | 12 months        | Infrarenal| None           | Pale, shocked, in an irresponsive State, vomiting, abdominal distention | Exploratory laparotomy | Rupture and death during operation |
| Dittrick K | Male | 12 years         | Infrarenal| None           | None                          | Collagen impregnated Dacron aortic graft 14 mm | Healthy at 2 years after surgery |
| Bell P | Female | 1 day            | Infrarenal| None           | Billous vomiting, large abdominal mass | Cryopreserved allograft 5 mm | Healthy at 14 months after surgery |
| Cheung SCW | Male | 6 months         | Juxtarenal| Bilateral common and external iliac artery aneurysms, right internal iliac artery aneurysm | None                | None              | Progression of thrombosis of the aneurysm and renal dysfunction at age 3 years |
| Buddingh KT | Male | 1 day            | Juxtarenal| Descending thoracic aortic aneurysm, left common iliac artery aneurysm | Bilous vomiting, anorexia, pulsatile abdominal mass | None              | Alive at 7 months, aneurysm has grown to a maximum diameter of 93 mm |
| Kim JI | None reported | 21 weeks’ gestation | Infrarenal| Bilateral common iliac artery aneurysms, left internal iliac artery aneurysm | None                | Dacron aortic graft 12 mm | Uneventful postoperative recovery |
| Malikov S | Male | 28 weeks’ gestation | Infrarenal| None           | Pulsatile abdominal mass | Repair with native iliac vessels | Healthy at 39 months after surgery |
| Cantinotti M | None reported | 22 weeks’ gestation | Unspecified | None reported | None reported | None reported | None reported |
| Tsunematsu R | Male | 25 weeks’ gestation | Unspecified | None           | Pulsatile abdominal mass | None              | Stable after 6 months follow up |
| McAteer J | Female | 32 weeks’ gestation | Thoracoabdominal | None | None | None | Died of rupture at age 4 weeks |
| Cho YP | Male | 23 months | Infrarenal| None | Irritability, vomiting, poor oral intake, diffuse tenderness, palpable pulsatile abdominal mass | Cryopreserved cadaveric artery 7 mm | Healthy at 10 months after surgery |
| Meyers RL | None reported | Neonate | Infrarenal| None | None | Decellularised, antigen reduced cryopreserved allograft | Healthy at 29 months after surgery |
| Author  | Gender | Age at discovery | Location   | Other aneurysms                                      | Symptoms related to aneurysms                                                                 | Surgical treatment            | Outcome                                                                 |
|---------|--------|------------------|------------|-----------------------------------------------------|---------------------------------------------------------------------------------------------|--------------------------------|------------------------------------------------------------------------|
| Ko Y    | Male   | 2 months         | Supraceliac| Two descending thoracic aortic aneurysms             | None reported                                                                              | Dacron aortic graft 10 mm    | Uneventful postoperative recovery                                      |
| Fettah ND | Female | 1 day            | Infrarenal | None                                                | Vomiting, abdominal distention, palpable pulsatile abdominal mass                           | Repair with polytetrafluorethylene patch | Died of sepsis and cardiopulmonary insufficiency at 4 weeks after surgery |
| Bivins HS | Male   | 19 weeks’ gestation | Infrarenal | Iliac artery aneurysms (details unknown)             | Large abdominal mass                                                                        | None                          | Died of renal failure at age 12 days                                   |
| Bansal A | Male   | 1 year           | Infrarenal | None                                                | Abdominal distension                                                                       | Dacron aortic graft 10 mm    | Uneventful postoperative recovery                                      |
| Kuboi T | Female | Neonate          | Infrarenal | None                                                | Lower back mass (subcutaneous vascular malformation)                                      | None reported                 | None reported                                                          |
| Sirisabya A | Female | 1 day            | Infrarenal | Left common iliac artery aneurysm, two small right renal artery aneurysms | Marked abdominal distension with a large pulsatile mass                                   | Gore-Tex vascular graft     | Thrombosis of the aortic graft and bilateral common iliac, internal iliac, and external iliac arteries at 13 months after surgery. Living a fairly normal life at 26 months after surgery |
| Higuchi K | Female | 4 years          | Infrarenal | Multiple intracranial aneurysms, bilateral hypogastric artery aneurysms, left renal artery aneurysm | Palpable pulsatile abdominal mass                                                           | Dacron aortic graft 10 mm    | Aneurysms of the left renal and left middle meningeal arteries were found to have increased and an intracranial arteriovenous fistula was detected. The aneurysms were embolised at 12 to 14 months after surgery. Healthy at 21 months after surgery |

Modified from Wang M, Tao Y. Diagnosis and treatment of congenital abdominal aortic aneurysm: a systematic review of reported cases. Orphanet J Rare Dis 2015;10:4
DISCUSSION

Idiopathic congenital AAA is an extremely rare disease of unknown origin; only 29 cases have been reported, including the present one (Table 1). The sex ratio among the published reports is 17 (males):nine (females), excluding those reports that fail to mention the sex of patients. The typical presentation of patients at hospital includes abdominal distention, vomiting, or abdominal pulsatile mass, and the diagnosis is confirmed by an abdominal ultrasound or CT. Seven cases (23%) were diagnosed prenatally by foetal ultrasound or MRI.1

In the current patient, AAA was probably part of a systemic disease rather than a local abnormality as multiple aneurysms were found. Whole exome sequencing showed no genetic mutation specific to the congenital connective tissue disorders associated with aneurysmal diseases. However, some genetic mutations may be involved in multiple aneurysms considering the patient’s age.

No universal management approach for congenital AAA exists; conservative management and surgical repair have been reported. Of these reported cases, 18 patients underwent revascularisation with vascular grafts, and Dacron graft or polytetrafluoroethylene graft were most frequently selected. Considering the risk of post-operative vascular graft infection and patient growth, revascularisation with cryopreserved allograft was performed in three patients. Revascularisation with native iliac vessels and aneurysmorrhaphy have also been reported. Conservative management was selected for patients in whom surgical repair carried a high mortality risk and was unlikely to improve life expectancy. Conservative management was undertaken in seven patients, four of whom died of heart or renal failure or aneurysm rupture. As conservative management for congenital AAA has a poor prognosis, surgical repair should be considered if the patient’s general condition is good and revascularisation is possible anatomically.

Long term follow up after surgery for congenital AAA in neonates, infants, and children has not been reported. Hence, the results of long term graft patency or need for reoperation are unclear. However, as an implanted Dacron graft does not grow, ischaemic symptoms of the lower limbs may arise as the patient grows. Although collateral circulation may prevent these symptoms, if they do arise, endovascular treatment such as balloon angioplasty or endografting, or surgical repair such as re-implantation with vascular graft or extra-anatomical bypass should be considered. Furthermore, as untreated aneurysms were seen to increase post-operatively in the present patient, frequent follow up with multimodality imaging for multiple aneurysms is necessary.

CONFLICTS OF INTEREST

None.

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