Congenital Abdominal Aortic Aneurysm: A Case Report and Literature Review

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Congenital abdominal aortic aneurysm is a rare disease with unknown etiology, and the common symptoms are abdominal pulsatile mass and pain caused by aneurysm rupture. The disease has a high mortality rate and fewer reports of surgical treatment. Here, we present a case of an idiopathic congenital abdominal aortic aneurysm. A 4-year-old boy had an abdominal pulsatile mass, and computed tomography angiography revealed an isolated infrarenal abdominal aortic aneurysm. To prevent rupture of the aneurysm, we repaired the aneurysm with artificial graft transplantation. No genetic mutation of the known congenital aneurysmal diseases was found in the whole-exome sequencing of the patient and his parents. There was no graft obstruction, and the patient grew well 40 months after surgery. Open surgery is the best treatment for idiopathic congenital abdominal aortic aneurysms. Surgical details such as timing and graft selection need to be further explored.

Keywords: aortic aneurysm, aortic diseases, congenital, children, surgical treatment

INTRODUCTION

Abdominal aortic aneurysms in children are very rare. The common causes are congenital connective tissue disorders, vasculitis, traumatic umbilical artery intubation, and infection, while congenital abdominal aortic aneurysms (cAAAs) is rarely reported and has an unknown etiology and high mortality. This article reports the case of a 4-year-old child with isolated cAAA and the results of a 40-month follow-up of open surgery.

CASE DESCRIPTION

In June 2018, a 4-year-old boy was hospitalized because of the discovery of a left abdominal pulsatile mass for 2 months. Computed tomography angiography (CTA) revealed an isolated infrarenal AAA with a maximum diameter of 67 mm (Figure 1). The patient was 112 cm tall and weighs 23 kg and he had no family history of aneurysmal disease, connective tissue disorders, a history of trauma, umbilical cannulation, and infection. Laboratory tests revealed no abnormalities, and blood pressure was normal.

DIAGNOSTIC ASSESSMENT, THERAPEUTIC INTERVENTION, FOLLOW-UP, AND OUTCOME

In July 2018, we repaired the AAA using open surgery to prevent aneurysm rupture. When we blocked the blood flow at both ends of the aneurysm and opened the aneurysm through a midline
incision, we found that the intima of the aneurysm was smooth without thrombus. We then used a 10-mm Dacron aorto-aortic tube graft to replace the AAA; the graft was oversized by 6 cm and formed a “C” shape to allow aortic growth. The aneurysmal sac was wrapped around the graft to avoid aortoduodenal fistula. The patient recovered well after the operation. Whole-exome sequencing of the boy and his parents revealed no genetic mutations of the known congenital aneurysmal diseases. The patients had frequent follow-ups outside the hospital, and at 40 months post-operative follow up without any antiplatelet drugs, the patient was 140 cm tall and weighs 43 kg, and the CTA revealed that the graft blood flow was unobstructed (Figure 2).

DISCUSSION

AAA is more common in the elderly with arteriosclerosis, and is rare in children and infants, and is commonly caused by congenital connective tissue disorders, vasculitis, umbilical cannulation, and infection, while cAAA is extremely rare and has an unknown etiology. One relevant hypothesis is that cAAA results from a developmental defect during embryogenesis that creates a focal narrowing of the abdominal aorta, which leads to poststenotic turbulent blood flow and subsequent aneurysm formation (1). There are no epidemiological data related to cAAA, and as of December 2021, only 31 cases have been reported (Table 1) (2–8). There are fifteen patients diagnosed before 1 year of age, eight patients were diagnosed after 1 year of age, and the remaining eight patients were diagnosed at 19–30 weeks of gestation. The male-to-female ratio was 18:10, 20 of 31 patients had infrarenal AAA, and 19 of 30 patients had other aneurysms. The reason for admission of patients is usually abdominal pulsatile mass or rupture of aneurysm; suspected diseases should receive vital imaging examination, such as ultrasound, which can provide a clear diagnosis.

The histopathological changes in the intima of cAAA include calcifications, thromboses, and ulcerations and ruptures of the layers (2, 9). Molecular genetic defects considered to be associated with AAA, Marfan’s syndrome, and Loeys-Dietz syndrome are caused by mutations in the genes encoding TGF-β2 or TGF-β receptor (TGFBR) I or II. Mutations in the fibrillin-1 (FBN1) gene have also been found to be associated with the occurrence of coronary aneurysms (10–13). Unfortunately, no similar genetic or molecular changes have been found in cAAA, including this case.

The mortality caused by cAAA rupture and renal failure was 30.76% (2). There is no universal approach to the management of cAAA. Although steroids, cyclophosphamide, antihypertensive drugs, non-steroidal anti-inflammatory drugs, and statins have certain curative effects, the reported mortality of conservative treatment is still as high as 57.14% (4/7). It is still unclear how to judge the diameter of aneurysms in the intervention, and the uncertainty of children’s activity cannot refer to the surgical standards of adults. Surgical repair after diagnosis should be considered. Endovascular aneurysm repair (EVAR) is not feasible in infants or children because of the lack of an appropriate endograft and the impact on patients’ growth and development. Artificial grafts and allografts were most frequently selected for revascularization, and in the past, 13 cases were reported using Dacron graft or polytetrafluoroethylene (PTFE) graft, 4 cases of allografts, and 1 case of native vessels. Although allografts have the advantages of high long-term patency and low risk of
| Author           | Gender | Age at discovery | Location      | Other aneurysms                                      | Other aneurysms                                      | Surgical treatment                      | Outcome                                      |
|------------------|--------|------------------|---------------|-----------------------------------------------------|-----------------------------------------------------|------------------------------------------|----------------------------------------------|
| Howorth Jr. MB   | Female | 1 day            | Infrarenal    | None                                                | Large abdominal mass, vomiting, anorexia             | Exploratory laparotomy                   | Rupture and death during operation          |
| Darden WA        | Male   | 2.5 years        | Infrarenal    | None                                                | None                                                | Dacron aortic graft                      | Died of pneumonitis at 5 months after surgery |
| Sterpetti AV     | Male   | 19 years         | Infrarenal    | None                                                | Middle epigastric pain, abdominal fullness, dysuria, abdominal pulsatile mass | Dacron aortic graft 18 mm               | Died of pneumonitis at 5 months after surgery |
| 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          |
| Latter D         | Male   | 1 month          | Infrarenal    | None                                                | Pulsatile abdominal mass                             | Polytetrafluoroethylene tube graft 8 mm | Healthy at 10 months after surgery          |
| Saad SA          | Male   | 6 weeks          | Infrarenal    | Left common iliac artery aneurysm mass              | Pulsatile abdominal mass                             | Aneurysmorrhaphy                         | Healthy at 3 months after surgery           |
| Myrmel T         | Male   | 30 years         | Infrarenal    | None                                                | Pulsatile abdominal mass, acute abdominal pain       | Albumin coated USCI graft sized 16 × 8 mm | Healthy at 1 year after surgery            |
| 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 |
| 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 GoreTef graft 7–4 mm          | Healthy at 1 month after surgery            |
| Laing AJ         | Male   | 12 months        | Infrarenal    | None                                                | Pale, shocked, in an unresponsive 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                                                |                                        | 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 | Billous vomiting, anorexia, pulsatile abdominal mass | None                                     | Alive at 7 months, aneurysm has grown to a maximum diameter of 93 mm |
| Kim JI           | None   | 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 | Juxtarenal | None                                                | Pulsatile abdominal mass                             | Repair with native iliac vessels         | Healthy at 39 months after surgery          |

(Continued)
TABLE 1 | Continued

| Author | Gender | Age at discovery | Location | Other aneurysms | Other aneurysms | Surgical treatment | Outcome |
|---------|--------|-----------------|----------|-----------------|-----------------|--------------------|---------|
| 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 | Intimability, 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 |
| 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 |
| Sirisabuya 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 |
| Kuboi T | Female | Neonate | Infrarenal | None | Lower back mass (subcutaneous vascular malformation) | None reported | None reported |
| 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 | Healthy at 21 months after surgery |
| Tanga CF | Male | 11 years | Infrarenal | Bilateral common iliac artery aneurysms, bilateral internal iliac artery aneurysm | Abdominal pain, shock | Dacron aortic graft 12 mm | Healthy at 10 months after surgery |
| LeNguyen A | Female | 36 weeks’ gestation | Infrarenal | Bilateral common iliac artery aneurysms, bilateral internal iliac artery aneurysm | None | Cryopreserved cadaveric artery 5 mm | Healthy at 12 months after surgery |

Modified from Wang and Tao (2).

postoperative graft infection, there are difficulties with the long-term use of immune-suppressants and allograft sources. Malikov reported a successful case of revascularization with native iliac vessels (14).

The common complications of artificial vascular grafts are graft stenosis and obstruction. The diameter of the artificial graft should be considered to match with the artery and ensure blood supply to the lower limbs. At present, the reported diameter...
is mostly between 8 and 12 mm, which is easier for older children to choose. There is a high risk of synthetic vascular graft occlusion with a diameter of <6 mm for a neonatal patient; it may, therefore, be more appropriate to delay surgery for smaller aneurysms to produce better results and prevent the need for follow-up surgery (15). It is necessary to reserve appropriate length for artificial grafts to adapt to the patient’s growth; however, most reports do not indicate the specific appropriate length. Dueppers et al. (16) reserved 4-cm graft to meet the growth needs of children. We considered the patient’s age, preoperative aortic diameter and adult physique estimated from his parents’ physique, and decided to use 10-mm Dacron graft to reconstruct the diseased artery with a 6-cm long graft reserved to form a “C” shape; our length selection principle is to keep a certain length on the premise of avoiding angulation.

The average follow-up time of the 16 patients who underwent surgical repair with follow-up records was 19 months. The longest follow-up time with artificial grafts under the age of 18 years was 26 months. Of these 16 patients, two died of infection during follow-up, an anastomotic stenosis of allografts occurred 7 days after surgery, and one PTFE graft was completely occluded 13 months after the operation. The graft patency rate was 93.3% (14/15). There is no literature recommending the routine use of anticoagulant or antiplatelet drugs for AAA patients with reconstructed branches. Le Nguyen et al. (8) continued to use low-molecular-weight heparin for patients with anastomotic stenosis, resulting in graft patency at 1-year follow-up. During the 40-month follow-up in our case, the artificial graft was twisted, but there was no obvious angulation and it remained unobstructed. The patient’s physical development was not affected. We will continue to follow up the patient to observe graft patency.

CAAA is rare and has unknown etiology, and for patients with confirmed aneurysms, it is suggested to improve the systemic examination and long-term follow-up to exclude other lesions. Due to the high mortality rate, the long-term results of open repair in children are still unclear. The patency of grafts also requires long-term follow-up observations and necessary drug adjuvant treatment. In cases of complications, timely and effective interventions are necessary.

DATA AVAILABILITY STATEMENT

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding authors.

ETHICS STATEMENT

The studies involving human participants were reviewed and approved by Medical Ethical Committee of the First Affiliated Hospital of Zhengzhou University. Written informed consent to participate in this study was provided by the participants’ legal guardian/next of kin. Written informed consent was obtained from the minor(s)’ legal guardian/next of kin for the publication of any potentially identifiable images or data included in this article.

AUTHOR CONTRIBUTIONS

ZZ was wrote the manuscript and was assistant in surgery. KM and YY were assistant in surgery and participate in editing the articles. ZH and ZL designed the operation and revising the manuscript. All authors contributed to the article and approved the submitted version.

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