Clinical Case Report

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Marfan syndrome combined with huge abdominal aortic aneurysm size of 20 × 11 cm

A case report of surgical approach

Tian-Hua Zhang, PhD[a], De-Cai Chi, MD[a], Wei-Liang Jiang, PhD[a], Shuai Qiang, MD[b,*]

Abstract

Rationale: Abdominal aortic aneurysm is one of the most common aneurisms. Patients presenting with secondary back pain should be given prompt medical attention. Herein, a rare case of a giant abdominal aortic aneurysm that was successfully treated with surgery is described.

Patient concerns: A 33-year-old Chinese male suffered from Marfan syndrome combined with giant abdominal aortic aneurysm, and presented with back pain, fever, nausea, vomiting, abdominal distention, and constipation. After undergoing numerous tests, the patient underwent an abdominal aortic aneurysm resection coupled with artificial graft bypass. The patient's recovery was smooth, and he was discharged 14 days after the operation in stable condition.

Diagnosis: Abdominal aortic aneurysm.

Interventions: The patient underwent a surgery involving mass resection and artificial graft bypass.

Outcome: The patient was discharged 14 days after surgery in stable condition.

Lessons: Giant abdominal aortic aneurysms are rarely seen, and aneurysmectomy associated with prosthetic vascular graft repair is an effective and standard treatment for such aneurysms.

Abbreviations: AAA = abdominal aortic aneurysm, CPR = curved planar reconstruction, CTA = computed tomography angiography, EVAR = endovascular abdominal aortic aneurysm repair, MIP = maximum intensity projection, MRA = magnetic resonance angiography, MRI = magnetic resonance imaging, PTFE = polytetrafluoroethylene, SSD = surface shading display, VR = volume rendering, WBC = white blood cell.

Keywords: abdominal aortic aneurysm, artificial graft bypass, artificial vascular replacement, prosthetic vascular graft repair, vascular surgical procedures

1. Introduction

Aortic aneurysm is characterized as an increase of >50% of the expected normal diameter, which presents as a permanent localized dilation of the artery. Abdominal aortic aneurysm (AAA) is a common disease, which seriously threatens patients' lives. AAA occurs in the infrarenal portion of the abdominal aorta and approximately accounts for 80% of all aortic aneurysms.[1] There are not subjective symptoms in most patients with AAA, which is occasionally detected during physical examination. A common sign is a pulsatile mass located in the peri-umbilicus or epigastrium.[2] Majority of the patients suffer from abdominal distention, indicating an oppression of adjacent organs. Hypotensive patients feel a sudden severe pain immediately before or after the rupture of AAA. The cause and pathogenesis of AAA remain unknown, and open repair or endovascular stent-grafts are the predominant treatment options. Herein, we describe a case of giant AAA that underwent successful aneurysm resection and reconstruction.

1.1. Ethical statement and consent

This study was approved by the Ethics Committee of Second Affiliated Hospital of Harbin Medical University. The clinical and imaging data were obtained with the patient's consent for publication of this report.

1.2. Case presentation

A 33-year-old Chinese male, 7-feet tall, weighing around 10 stone, suffering from Marfan syndrome, presented at our hospital with complaints of back pain, fever, nausea, vomiting, abdominal distention, and constipation. Physical examination showed a giant 20×11 cm pulsatile abdominal mass (Fig. 1) combined with an abnormal groin pulsation and arachnodactyly. Further tests were performed after admission. Laboratory examination showed an elevated white blood cell (WBC) count. Computed tomography angiography (CTA) of abdominal aorta (Fig. 2) and

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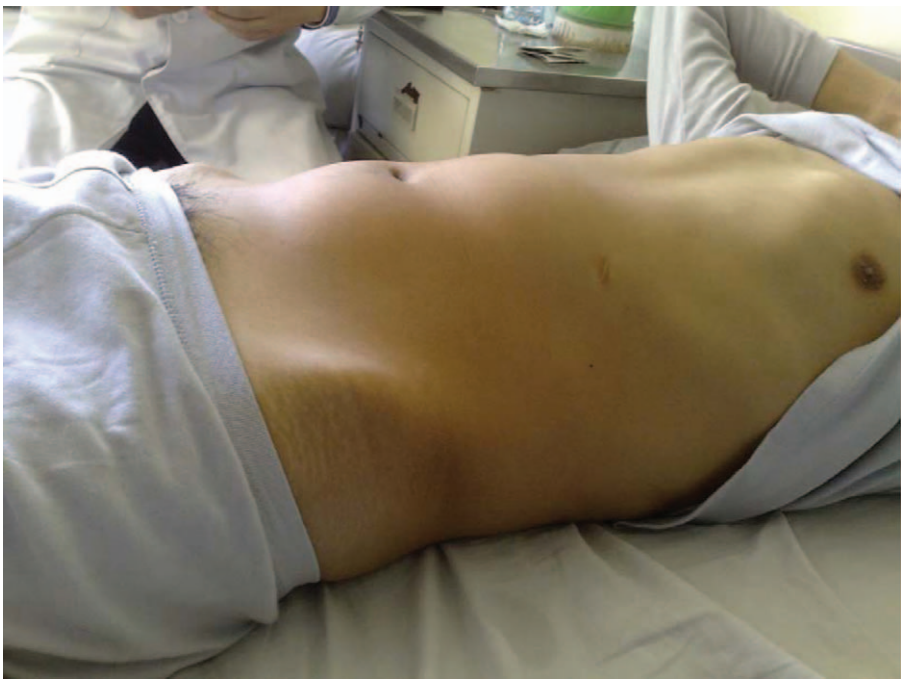

Figure 1. A large pulsatile mass located in the peri-umbilicus (patient in supine position).

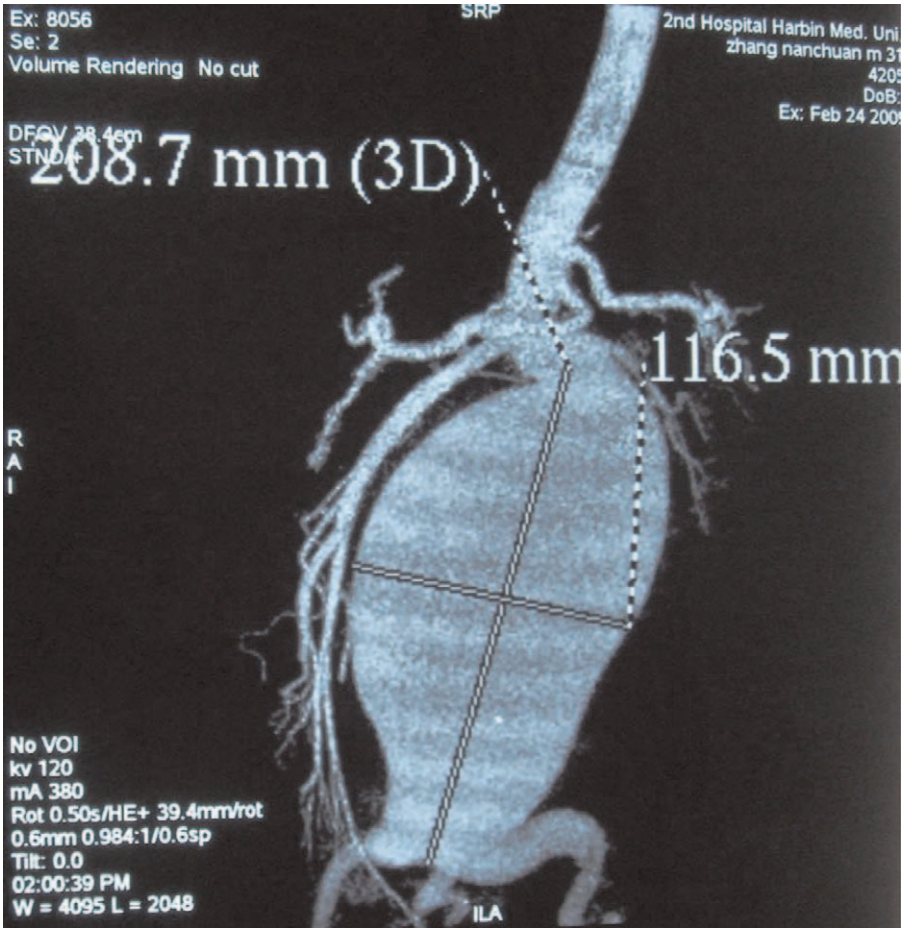

Figure 2. Computed tomography angiography (CTA) of abdominal aorta revealed a giant abdominal aortic aneurysm (AAA), approximately 20 × 11 cm in diameter.

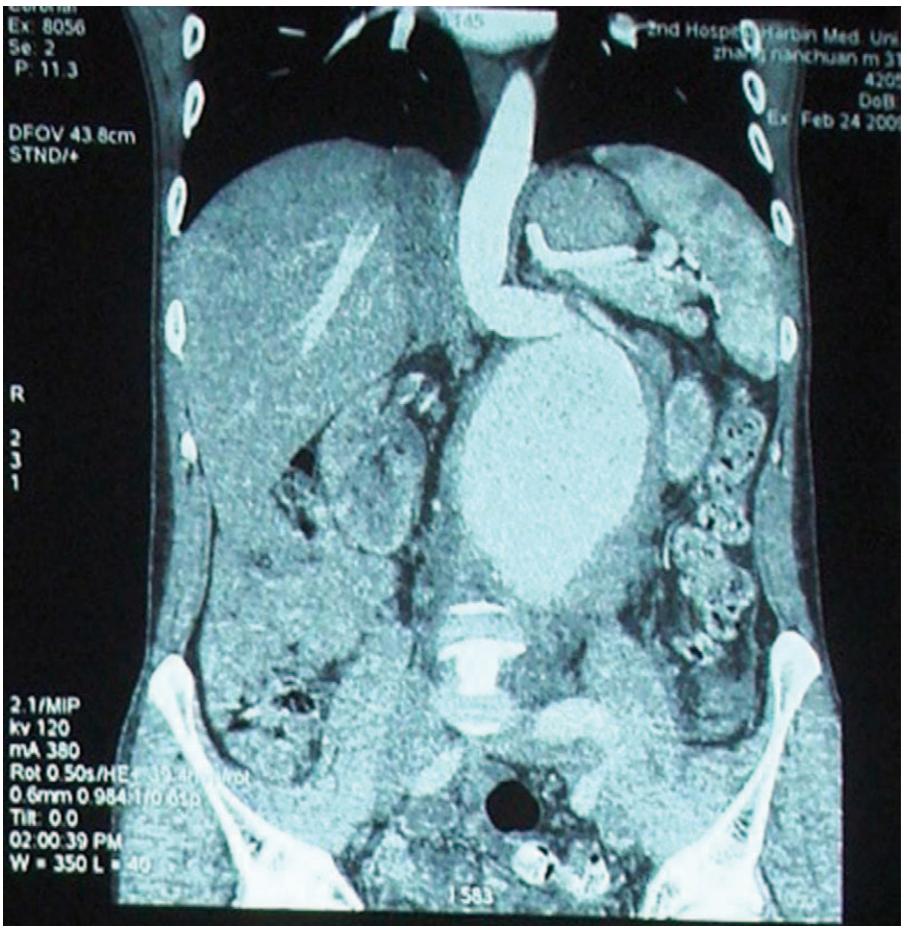

Figure 3. Intravenous contrast enhanced computed tomography reconstruction of the abdomen indicated that the abdominal aortic aneurysm (AAA) was associated with external hematoma, intramural calcified plaque, and oppression of adjacent organs (coronal frontal view).

intravenous contrast enhanced coronal (frontal) computed tomography (CT) reconstruction of the abdomen (Fig. 3) subsequently demonstrated the presence of an AAA of approximately 20 × 11 cm, with external hematoma, intramural calcified plaque, and pressure on adjacent organs.

Because of arachnodactyly and a giant AAA, the patient underwent surgery without any interventional treatment. During laparotomy, a giant AAA was detected (Fig. 4) and freed from the neck and bilateral common iliac arteries by temporarily blocking the arteries after promptly injecting heparin (30 mg) into the vein. Then the mass was opened, reconstructed with end-to-end anastomosis by grafts in polytetrafluoroethylene (PTFE) (Fig. 5). Pathology of abdominal aorta wall revealed fibrous connective tissue proliferation, hyalinosis associated with inflammation, cholesterol crystals, and calcification. The patient's vital signs and other monitored parameters were normal without colonic ischemia after returning to the ward, and he was discharged 14 days after surgery in stable condition.

2. Discussion

AAA is a potentially fatal disease with an incidence of 2% to 9% among older adults aged >65 years, but its causes remain unknown.[3,4] The pathogenesis of AAA may be related to familial risk,[5] inflammation,[6] and several signals.[7,8] There are no subjective symptoms in most patients with AAA. A common sign is a pulsatile mass located in the peri-umbilicus or epigastrium.[2] Patients feel abdominal distention usually indicating an oppression of adjacent organs. The present case also suffered from the above-mentioned symptoms and was confirmed as arteriosclerosis by pathology.

AAA is a very malignant disease characterized by a high incidence of mortality (70%) after rupture.[9] A sudden severe pain may indicate peritoneal or retroperitoneal rupture. A growing tumor could result in compression of adjacent organs such as duodenum, the proximal jejunum and ureter, which was observed in our patient who developed hydronephrosis due to an oppression. In some cases, if the thrombosis in the AAA breaks, it could lead to an arterial embolization.

AAA can be noninvasively diagnosed by B-ultrasound, CT, and magnetic resonance imaging (MRI). However, none of them clearly display the vascular anatomical structure around the aneurysm. In recent years, the application of spiral CT, magnetic resonance angiography (MRA), and some new techniques, such as curved planar reconstruction (CPR), maximum intensity projection (MIP), volume rendering (VR), and surface shading display (SSD) are very useful in preoperative diagnosis. To identify collateral circulation and small branches of AAA before operation, selective arteriography of abdominal aorta is the gold standard.

This patient had a giant AAA based on known criteria. Open repair was performed instead of interventional

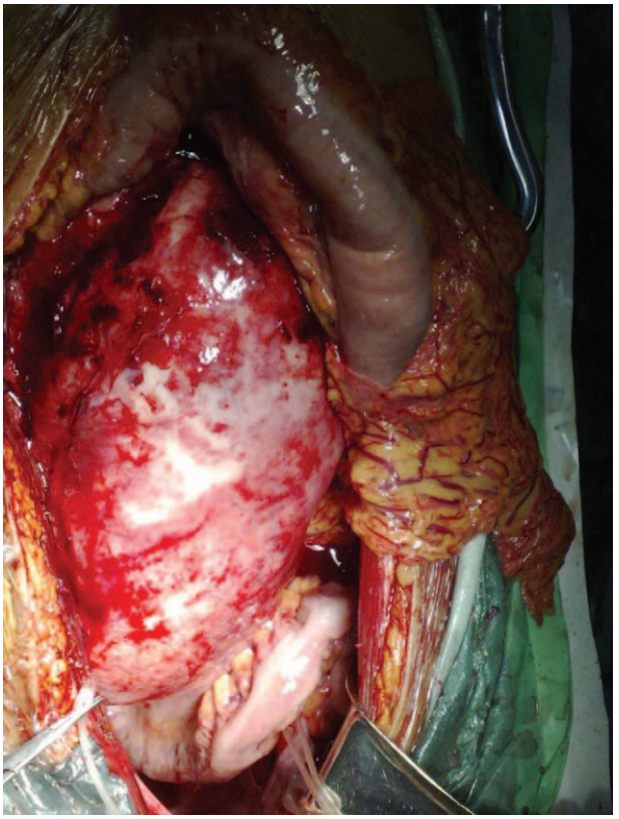

Figure 4. A giant abdominal aortic aneurysm (AAA) (intraoperative view).

treatment given his Marfan syndrome and the size of the mass. Open repair is typically more appropriate for large infrarenal AAA in people <65 years of age. However, endovascular AAA repair (EVAR) is the first choice for frail elderly patients, although there is no significant difference between the 2 approaches. In Lovegrove's[10] study comprising 21,178 patients, neither endovascular repair nor open surgery offered a clear long-term benefit in all-cause mortality. Prinssen et al[11] conducted a randomized trial to compare operative (30-day) mortality with open repair versus endovascular repair in 345 patients. The endovascular repair was superior to open repair in patients with AAA ≥5 cm in diameter. Similarly, a systematic review by Pei et al[12] included 27 clinical studies and 7226 patients, and suggested that patients who received EVAR had significantly lower 30-day mortality, less blood loss or blood transfusion, less colonic ischemia, fewer cardiac and respiratory complications, shorter hospital and intensive care unit stay, and fewer overall operative morbidities. However, EVAR involved an obviously higher frequency of early secondary procedures and graft-related complications, with a higher cost. Therefore, EVAR offers apparent benefit to aneurysm patients in the early postoperative period but in the mid- and long-term, the 2 techniques have no remarkable difference.

It is imperative to strictly understand the proper indications, achieve sufficient preoperative preparation, standardize the steps, and prevent and manage postoperative complications. These are key factors for improving operation and survival rate to obtain satisfactory treatment efficacy. There were only 5 reported cases[13–17] (retrieved from the PubMed database) of true AAA in adult patients with maximum diameter of 13.5, 14, 15, and 18 cm, respectively, as AAA can rarely reach giant dimensions, and the present case must be considered exceptional.

3. Conclusion

With the gradual improvement of living standards and increase in the average life expectancy, incidence of aneurysms has been steadily increasing every year, which demands an advancement in its diagnosis and treatment. In this report, open repair was successfully used for a giant AAA, which is important for learning and reference.

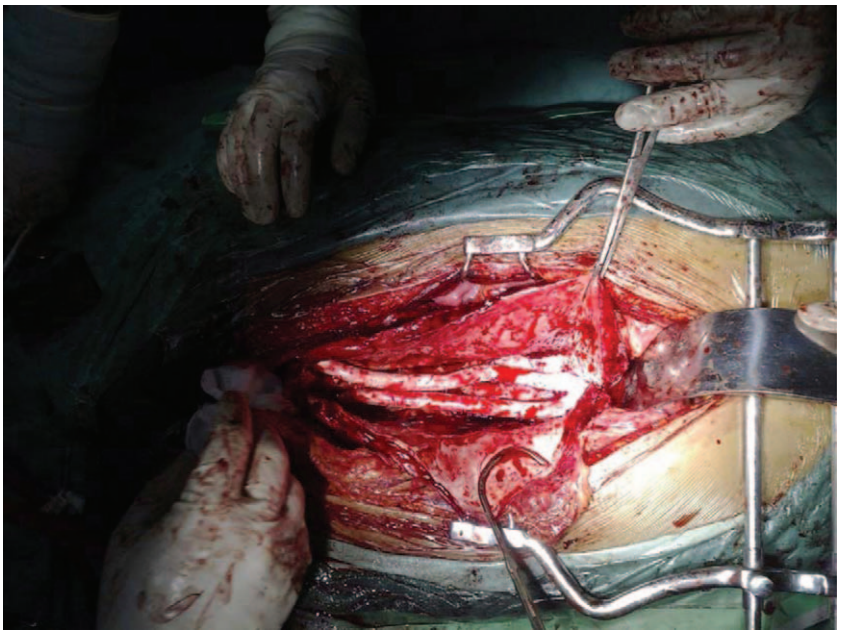

Figure 5. A reconstruction with end-to-end anastomosis by graft in polytetrafluoroethylene (PTFE).

Author contributions

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Writing – original draft: Shuai Qiang.

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