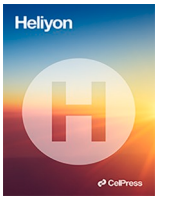
Heliyon 9 (2023) e13022

Contents lists available at ScienceDirect

Heliyon

journal homepage: www.cell.com/heliyon



Case report

A rare middle aortic syndrome with celiac trunk, superior mesenteric and bilateral renal artery involvement

Qi Guo, Huijia Liu, Xue Li, Menglin Wu, Jiang Li, Xuening Zhang *

Department of Radiology, Second Hospital of Tianjin Medical University, No. 23, Pingjiang Road, Hexi District, Tianjin 300211, PR China

ARTICLE INFO

Keywords:
Middle aortic syndrome
Computed tomography angiography
Atypical aortic coarctation
Thoracoabdominal aortic bypass grafting
Follow-up

ABSTRACT

Middle aortic syndrome (MAS) is a rare atypical aortic coarctation (AC), often accompanied by refractory renal hypertension, which eventually leads to death from congestive heart failure, stroke or hypertensive encephalopathy. Computed tomography angiography (CTA) has unique advantages in assessing aortic stenosis and splanchnic artery abnormalities. Prompt aortic bypass surgery can relieve symptoms and improve quality of life. In this study, we report a patient with MAS diagnosed by CTA and follow-up after thoracoabdominal aortic bypass grafting.

1. Introduction

Aortic coarctation (AC) is a segmental narrowing of the aortic lumen, accounting for 5%–10% of congenital heart disease, which is typically located at the aortic isthmus between the left subclavian artery and the ductus arteriosus [1]. Middle aortic syndrome (MAS), first proposed by Sen [2], refers to an atypical AC characterized by a long segmental or diffuse narrowing of the distal thoracic aorta and upper abdominal aorta, often involving with the bilateral renal arteries and other visceral arteries. It is a rare cause of AC, accounting for less than 2% [3]. The etiology is still unclear, including congenital abnormal development or obtained from Takayasu arteritis, neurofibromatosis and so on [4,5].

MAS patients usually present with severe refractory renal hypertension, lower extremity claudication, as well as some other atypical symptoms, such as headache, dizziness and fatigue. Untreated patients mostly die from congestive cardiac failure, stroke, or hypertensive encephalopathy due to progressive hypertension. Management of MAS includes medical, endovascular intervention and surgical treatment [6,7]. As symptomatic supportive treatment, medication can achieve the effects of lowering blood pressure, dilating blood vessels, improving microcirculation and so on [7]. Endovascular interventional therapy and surgical artificial bypass surgery can alleviate hypertension that is difficult to control with drugs, and reduce the incidence of its associated fatal complications [6,8,9]. Therefore, timely and effective treatment can alleviate patient's symptoms and improve quality of life.

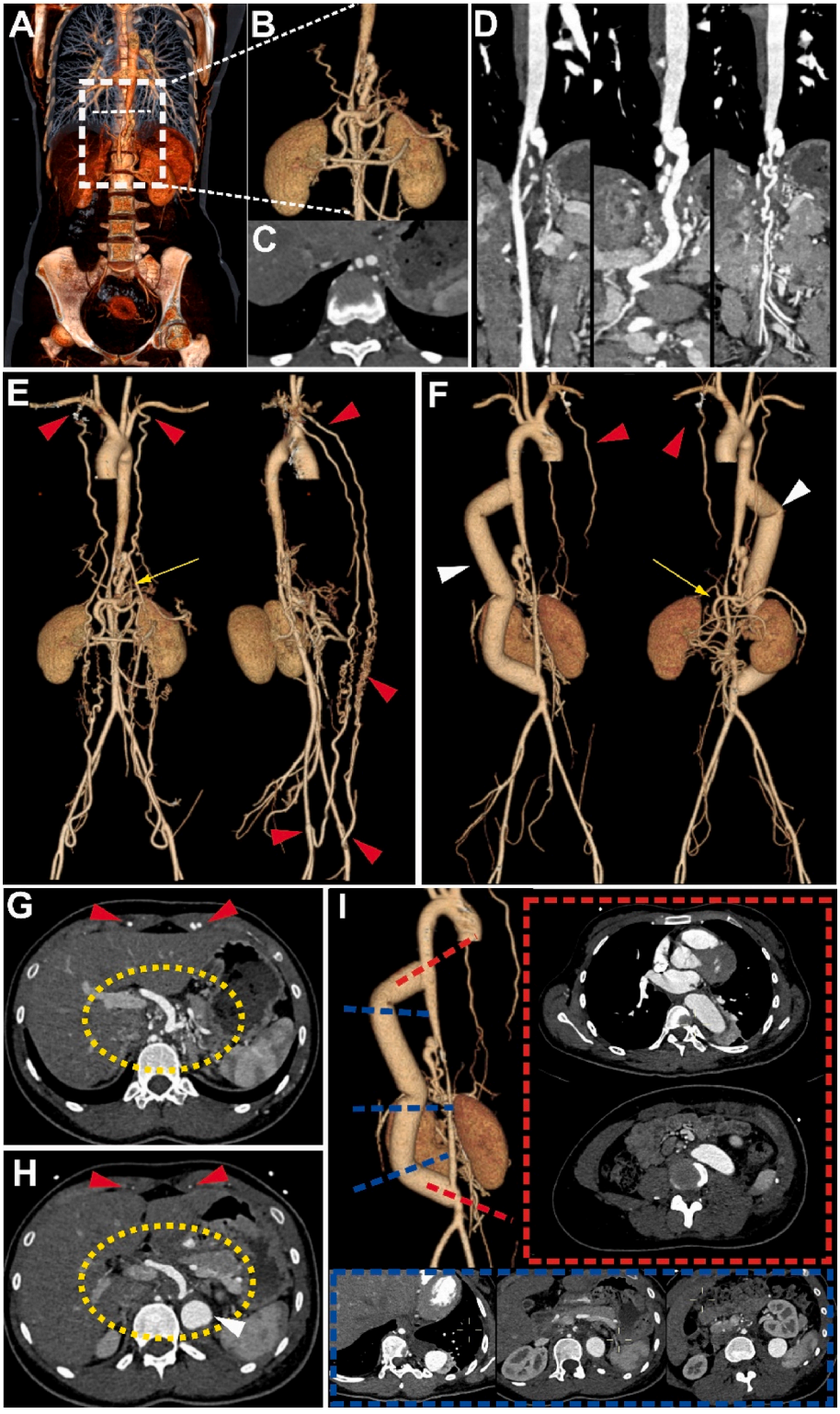
In this report, we present a case of a young woman with MAS which associated with celiac trunk, superior mesenteric and bilateral renal artery stenosis, who was first diagnosed by CTA and successfully relieved existing symptoms by undergoing endoscopic-assisted thoracoabdominal aortic bypass grafting.

2. Case presentation

A 22-year-old woman who had hypertension (149/72 mmHg) for 3 years with a maximum systolic blood pressure of 160 mmHg,

* Corresponding author. ,
E-mail address: luckyxn_tianjin@163.com (X. Zhang).

https://doi.org/10.1016/j.heliyon.2023.e13022
Received 28 September 2022; Received in revised form 12 January 2023; Accepted 12 January 2023
Available online 18 January 2023



(caption on next page)

Fig. 1. (A)is a VR pseudo-color image of the patient visually showing vascular stenosis, (B)is an enlarged 3D imaging of the stenotic segment of the blood vessel, and multiple tortuous small blood vessels can be seen, and (C) is the axial image of the narrowest vessel; (D) is the reconstructed images for the curved surfaces of the three branches of the lower thoracic aorta (from left to right); Preoperative (E and G) and postoperative (F and H) three-dimensional and axial images of the patient's vessels, showing that multiple collateral vessels in the preoperative abdominal cavity (yellow arrows and ovals) and abdominal wall (red arrowheads) were significantly reduced postoperatively, and surgical artificial blood vessel (white arrowheads); (I) is an image of the proximal and distal anastomosis of the graft to the native aorta (red line and box), the suspected kink and its distal segment (blue line and box) after surgery, respectively.

had been taking valsartan during this period, but her blood pressure did not return to the normal blood pressure range. She recently felt numbness of lower limbs after prolonged sitting, which was relieved after physical activity. In addition, she had frequent nosebleeds. Physical examination showed a discrepant blood pressure (BP) between the upper and lower limbs (left upper extremity: 139/78 mmHg; right upper extremity: 136/82 mmHg; left lower extremity: 113/81 mmHg; right lower extremity: 128/85 mmHg). Muscle tension and strength of extremities were normal, and oxygen saturation was between 97% and 99%. No murmurs were detected in each valve auscultation area and abdomen. Laboratory analysis showed the complete blood count, serum electrolytes, urea, creatinine, liver function, renal function, coagulation tests and inflammatory markers were normal. Therefore, the preliminary clinical consideration is AC.

Aortic CTA was performed and confirmed the diagnosis of MAS, which demonstrated the lumens of the distal thoracic aorta and abdominal aorta markedly narrowed, with the narrowest point being approximately 3 mm above the diaphragm (Fig. 1A–C). Moreover, the diameter of the abdominal aorta lumen was not exceed 10 mm (Fig. 1D). Two branches were seen in the lower thoracic aorta, both passing through the diaphragm and descending into the celiac trunk and superior mesenteric artery respectively (Fig. 1D). The left renal artery was supplied by both the abdominal aorta and celiac trunk branches, while the right renal artery was supplied by both the abdominal aorta and diaphragmatic artery branches. **In addition, the bilateral internal thoracic arteries (also named the mammary arteries), which were branches of the bilateral subclavian arteries, have clearly shown to anastomose with the inferior epigastric artery, branches of the external iliac arteries (**Fig. 1E**).**

Subsequently, the patient successfully underwent descending aorta-abdominal aortic bypass surgery. On the third day after surgery, the blood pressure dropped to 111/64 mmHg. The patient recovered well, and was examined again by CTA on the 12th day after surgery. The examined CTA showed artificial blood vessels next to the narrowed distal thoracic aorta and abdominal aorta (Fig. 1F). Furthermore, CTA also found that the superior mesenteric artery, the branches of the celiac trunk and the diaphragmatic artery which reach the renal artery were thinner than before. **While the bilateral internal thoracic arteries and the inferior epigastric arteries were significantly thinner than before surgery** (Fig. 1E–H). Written informed consent was obtained from the patient for the publication of any potentially identifiable images or data included in this article.

3. Discussion and conclusions

As an atypical AC, MAS is a rare clinical condition, typically diagnosed in childhood or adolescence with previously undiagnosed systemic hypertension, blood pressure difference of extremities, and weak or absent femoral pulses [10]. Studies have reported that the average age of MAS patients is about 9 years, the incidence of male and female is equal, and the most common finding symptom is high blood pressure [11]. **The pathogenesis of most MAS is mainly speculative, which may be a congenital anomaly due to failure of paired dorsal aortic fusions around the 25th day of fetal development or an acquired anomaly due to atherosclerosis, Takayasu's arteritis, or nonspecific inflammation and so on** [12,13]. In this present case, the pathology was speculated to be congenital hypoplasia because the patient had no specific signs, no evidence of an inflammatory process in the blood sample, and no history of any systemic signs of inflammation [14].

Aortography is the gold standard for MAS diagnosis, but it cannot be observed the extramural structure of the artery. CTA is increasingly being used as the preferred alternate imaging modality for diagnosis, especially in assessing the location and length of the narrowed segment and evaluating the visceral artery abnormalities and collateral circulation [15,16]. In general, congenital and acquired MAS have different locations of aortic narrowing, with the most common anatomic site of congenital MAS being the infrarenal, often involving the renal and visceral branches of the aorta [17]. For congenital MAS, the stenosis results from hypoplasia without inflammatory response. Therefore, CTA usually shows significant narrowing of the lumen, with normal wall thickness, and no inflammatory changes around the vessel. Since the arterial lumen in this case was significantly narrowed without markedly circumferential arterial wall thickened, which was further presumed to be congenital.

MAS generally has a poor prognosis in untreated cases. There are several treatment options available to suit personalized treatment. **Angioplasty is the most common endovascular treatment and can be used even in pediatric patients with mild to severe vascular stenosis, as well as in younger patients for temporary relief of hypertension prior to surgical repair** [9,13]. Surgical treatment using bypass grafts is the main treatment for MAS complicated with visceral artery stenosis (renal and mesenteric vessels involvement) [18]. Hypertension symptoms improved or were cured in more than two-thirds of patients after surgery [19]. In this case, the patient's blood pressure recovered to normal after surgery, and the collateral circulation of the abdominal wall was closed during postoperative CTA examination.

Our case highlights (1) AC should be suspected in young patients with refractory hypertension during a comprehensive physical examination, such as a marked difference in upper and lower limb pressure; (2) imaging examinations such as CTA can assist in diagnosing the location and extent of AC, and help to distinguish MAS from congenital or acquired; and (3) surgical revascularization techniques can alleviate hypertension, reduce the incidence of fatal complications, and make hypertension that was difficult to control

with drugs to be controllable or normalized.

In conclusion, MAS cases are rare, and the narrow range and variation degree with abundant collateral circulation reported in this case are even rarer. This report provides the whole process of diagnosis, surgical treatment, and postoperative review, indicating that it is feasible for such patients to be diagnosed and treated in a timely manner.

This item belongs to the item group IG000035.

Author contribution statement

All authors listed have significantly contributed to the investigation, development and writing of this article.

Funding statement

Dr Qi Guo was supported by the Scientific Research Project of Tianjin Municipal Education Commission [2019KJ165].

Data availability statement

Data included in article/supp. Material/referenced in article.

Declaration of interest's statement

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Additional information

No additional information is available for this paper.

References

[1] T. Celik, H. Kursaklioglu, A. Iyisoy, H. Turhan, B. Amasyali, M. Kocaoglu, E. Isik, Hypoplasia of the descending thoracic and abdominal aorta: a case report and review of literature, J. Thorac. Imag. 21 (4) (2006 Nov) 296–299, https://doi.org/10.1097/01.rti.0000213555.61752.3a.
[2] P.K. Sen, S.G. Kinare, S.D. Engineer, G.B. Parulkar, The middle aortic syndrome, Br. Heart J. 25 (5) (1963 Sep) 610–618, https://doi.org/10.1136/hrt.25.5.610.
[3] R.K. Rumman, C. Nickel, M. Matsuda-Abedini, A.J. Lorenzo, V. Langlois, S. Radhakrishnan, J. Amaral, L. Mertens, R.S. Parekh, Disease beyond the arch: a systematic review of middle aortic syndrome in childhood, Am. J. Hypertens. 28 (7) (2015 Jul) 833–846, https://doi.org/10.1093/ajh/hpu296.
[4] F. Daghero, N. Bueno, A. Peirone, J. Ochoa, G.F. Torres, J. Ganame, Coarctation of the abdominal aorta: an uncommon cause of arterial hypertension and stroke, Circ Cardiovasc Imaging 1 (1) (2008 Jul) e4–e6, https://doi.org/10.1161/CIRCIMAGING.108.767947.
[5] X. Meng, J. Xue, J. Cai, H. Zhang, W. Ma, H. Wu, X. Zhou, Y. Lou, L. Wang, A single-center cohort of mid-aortic syndrome among adults in China: etiology, presentation and imaging features: mid-aortic Syndrome Among Adults, Am. J. Med. Sci. (2022 Apr 12), https://doi.org/10.1016/j.amjms.2022.04.004. S0002-9629(22)00148-3.
[6] M. Rabellino, L. Garcia-Nielsen, G. Gonzalez, S. Baldi, T. Zander, M. Maynar, Middle aortic syndrome percutaneous treatment with a balloon-expandable covered stent, J. Am. Coll. Cardiol. 56 (6) (2010 Aug 3) 521, https://doi.org/10.1016/j.jacc.2010.01.066.
[7] Q. Pellenc, A. Roussel, P. Mordant, M.P. Chauveheid, T. Papo, Y. Castier, A rare case of acquired thoracic aortic coarctation treated medically, Circulation 133 (2) (2016 Jan 12) 234–235, https://doi.org/10.1161/CIRCULATIONAHA.115.018177.
[8] S. Indriani, S. Erriyanti, R. Dewangga, S. Adiarto, T. Siddiq, I. Dakota, Late presentation of middle aortic syndrome complicated with severe aortic regurgitation; the role of endovascular intervention as a bridging for Bentall surgery, J Vasc Surg Cases Innov Tech 8 (1) (2021 Dec 8) 48–52, https://doi.org/10.1016/j.jvscit.2021.11.005.
[9] S. Indriani, S. Erriyanti, R. Dewangga, S. Adiarto, T. Siddiq, I. Dakota, Late presentation of middle aortic syndrome complicated with severe aortic regurgitation; the role of endovascular intervention as a bridging for Bentall surgery, J Vasc Surg Cases Innov Tech 8 (1) (2021 Dec 8) 48–52, https://doi.org/10.1016/j.jvscit.2021.11.005.
[10] K. Yokoyama, T. Minami, M. Seki, Y. Okada, H. Kumagai, T. Yamagata, A boy with Alagille syndrome coexisting with mid-aortic syndrome and renovascular hypertension, J Cardiol Cases 21 (1) (2019 Sep 27) 28–31, https://doi.org/10.1016/j.jccase.2019.09.010.
[11] R.K. Rumman, C. Nickel, M. Matsuda-Abedini, A.J. Lorenzo, V. Langlois, S. Radhakrishnan, J. Amaral, L. Mertens, R.S. Parekh, Disease beyond the arch: a systematic review of middle aortic syndrome in childhood, Am. J. Hypertens. 28 (7) (2015 Jul) 833–846, https://doi.org/10.1093/ajh/hpu296.
[12] F. Daghero, N. Bueno, A. Peirone, J. Ochoa, G.F. Torres, J. Ganame, Coarctation of the abdominal aorta: an uncommon cause of arterial hypertension and stroke, Circ Cardiovasc Imaging 1 (1) (2008 Jul) e4–e6, https://doi.org/10.1161/CIRCIMAGING.108.767947.
[13] P.A. Patel, A.M. Cahill, Renovascular hypertension in children, CVIR Endovasc 4 (1) (2021 Jan 7) 10, https://doi.org/10.1186/s42155-020-00176-5.
[14] N. Forman, J. Sinskey, A. Shalabi, A review of middle aortic syndromes in pediatric patients, J. Cardiothorac. Vasc. Anesth. 34 (4) (2020 Apr) 1042–1050, https://doi.org/10.1053/j.jvca.2019.07.130.
[15] L.L. Geyer, U.J. Schoepf, F.G. Meinel, J.W. Nance Jr., G. Bastarrika, J.A. Leipsic, N.S. Paul, M. Rengo, A. Laghi, C.N. De Cecco, State of the art: iterative CT reconstruction techniques, Radiology 276 (2) (2015 Aug) 339–357, https://doi.org/10.1148/radiol.2015132766.
[16] S. Ma, K. Zhou, Y. Ma, Q. Ma, Y. Hou, Coronary CTA would facilitate invasive angiography in patients with acute coronary syndrome and coronary bypass grafting history, Front Cardiovasc Med 9 (2022 Mar 29), 751527, https://doi.org/10.3389/fcvm.2022.751527.
[17] C. Lazea, C. Al-Khzouz, C. Sufana, D. Miclea, C. Asavoaie, I. Filimon, O. Fufezan, Diagnosis and management of genetic causes of middle aortic syndrome in children: a comprehensive literature review, Therapeut. Clin. Risk Manag. 18 (2022 Mar 16) 233–248, https://doi.org/10.2147/TCRM.S348366.
[18] S.M. Kim, I.M. Jung, A. Han, S.I. Min, T. Lee, J. Ha, S.J. Kim, S.K. Min, Surgical treatment of middle aortic syndrome with Takayasu arteritis or midaortic dysplastic syndrome, Eur. J. Vasc. Endovasc. Surg. 50 (2) (2015 Aug) 206–212, https://doi.org/10.1016/j.ejvs.2015.04.032.
[19] Д.О. ДзюрМан, М.П. Радченко, Е.О. АртеМенко, Б.Б. Серденко, М. Мець, Successful surgical treatment of supradiaphragmatic descending thoracic aorta coarctation with anomalous origin of truncus coeliacus, Ukrainian Journal of Cardiovascular Surgery (2018) 85–87, https://doi.org/10.30702/ujcvs/18.33/21(085-087.