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Hybrid intravascular management of pediatric complex tubular aortic coarctation in the shadow of SARS-CoV-2 pandemic

Hybrydowa przezskórna terapia złożonej koarktacji aorty w cieniu pandemii SARS-CoV-2

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Abstract

Introduction. Coarctation of the aorta (CoA) is a congenital heart defect defined as a narrowing in the region of aortic isthmus, clinically presenting with peripheral perfusion disturbances in physical examination. Newborns are majority of the affected and surgery is the management of choice in these patients. However, some may not present with CoA symptoms until later childhood, with mostly percutaneous interventions as a favorable method of treatment. Aneurysms, heart failure and stroke are the main complications of untreated CoA, which explains low survival rate of undiagnosed patients.

Case report. A 5-year-old female patient in good general condition presented with a heart murmur during a prophylactic pediatric control. The patient was referred to pediatric cardiologist, but unfortunately due to ongoing coronary disease 2019 pandemic only telemedical consultation was available, precluding physical examination. Finally, the patient was referred to hybrid pediatric cardiac surgery department as an urgent consultation of the dubious anamnesis.

The echocardiographic study revealed continuous, non-pulsatile flow in the abdominal aorta with a narrowed descending aorta behind the left atrium, and the CoA was a suspected diagnosis. The angio-computed tomography (CT) confirmed tubular (55 mm long) narrowed section of thoracic aorta (up to 2.5 mm diameter) with concomitant collateral circulation.

Due to the anatomy of the aortic lesion the patient was referred for transcatheter stent graft (Bentley BeGraft© 9 mm/ /57 mm) implantation with ECC-backup. An initial dilatation to 9 mm except the region around 6mm long narrowed to 5.2 mm was performed. After 2 months, the stent graft was dilated again, and echocardiography confirmed uniformed aortic lumen (9 mm).

The postprocedural course was uncomplicated; the patient was discharged home and referred for further follow-up.

Conclusions. The CoA rarely occupies a non-typical region and therfore may be diagnostically challenging without a profound phisical examination, particularly in later childhood and pandemic settings, and may result in serious complications, if untreated.

Key words: coarctation of aorta, congenital heart defects, interventional cardiology, pediatric cardiac surgery, hybrid treatment

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Introduction

Coarctation of the aorta (CoA) is a congenital heart defect defined as a narrowing in the region of aortic isthmus. Although the condition usually manifests itself as a discrete constriction of the aortic isthmus, it may also present as a tubular hypoplasia and different morphological variations between these two [1].

As the fetal echocardiographic diagnosis remains difficult, the postnatal transthoracic echocardiography along with physical examination are the most common diagnostic tools for CoA. Moreover, the magnetic resonance imaging is preferred as the advanced imaging method [2], if available. The physical examination findings usually include delayed or reduced femoral pulse, a supine arm-leg blood pressure gradient (> 20 mm Hg), or a murmur due to rapid blood flow across the CoA [2].

Newborns and infants are the majority of the presenting children, and the urgent surgical correction is the management of choice in these patients. However, some rare cases may present with mild CoA symptoms until later childhood or adolescence, with mostly percutaneous interventions as a favorable method of treatment [3]. Heart failure, aortic aneurysms and stroke are the main complications of untreated CoA, which explains the low survival rate of undiagnosed patients [1–3].

Case report

A 5-year-old female patient in a good general condition presented with a heart murmur during a routine prophylactic pediatric control. As the first case of coronavirus disease 2019 (COVID-19) was confirmed on 4th of March 2020 in Poland, the National Health Found Institution recommended the limitation of elective admissions and surgical procedures from the 23rd of March 2020 until further notice. [4] Therefore, the patient was referred to pediatric cardiologist. Unfortunately, due to ongoing COVID-19 pandemic, only the telemedical consultation was available which precluded physical examination. Finally, after several remote consultations, the patient was referred to hybrid pediatric cardiac surgery department as an urgent admission of the dubious anamnesis.

The echocardiographic study revealed continuous, non-pulsatile flow in the abdominal aorta with a narrowed descending aorta behind the left atrium, and the CoA was a suspected diagnosis (Figure 1). The angio-computed tomography (CT) confirmed tubular (55 mm long) narrowed section of thoracic aorta (to 2,5 mm diameter) with concomitant collateral circulation (Figure 2).

Due to the anatomy of the aortic lesion the patient was referred for hybrid transcatheter stent graft implantation (Bentley BeGraft © 9 mm/57 mm) in hybrid operating theatre with extracorporeal circulation — backup. The first procedure finished with an initial dilatation to 9 mm except the region of the thoracic aorta around 6 mm long narrowed to 5.2 mm (Figure 3). After 2 months, the stent graft was dilated again, and the echocardiography confirmed uniformed aortic lumen (9 mm) (Figure 4).

The postprocedural course was uncomplicated and the patient was discharged home and referred for further clinical follow-up. The 2-month follow up confirmed good result of the procedures without any residual restenosis.

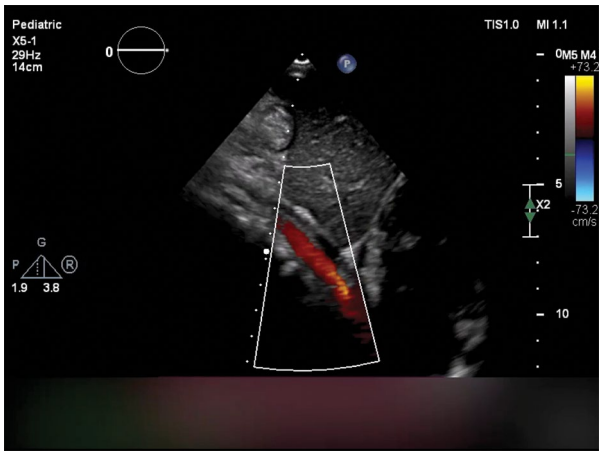

Figure 1. Echocardiographic study — continuous, non-pulsatile flow in the abdominal aorta

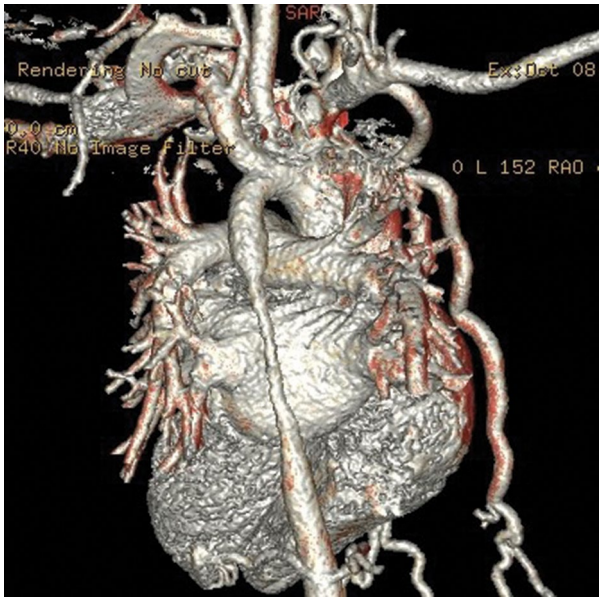

Figure 2. Computed tomography (CT) three-dimensional (3D) reconstruction — narrowing tubular (55 mm long) narrowed section of thoracic aorta (to 2.5 mm diameter) with concomitant collateral circulation

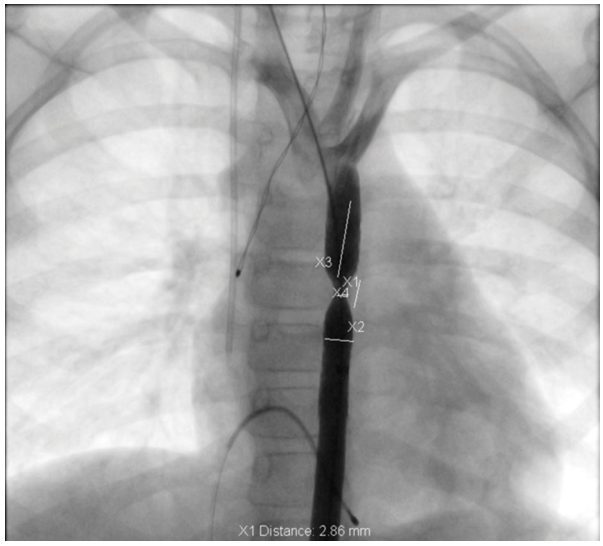

Figure 3. Angiography — the result after the first stent graft implantation. X1 distance: 2.86 mm, X2 distance: 6.94 mm, X3 distance: 18.56 mm, X4 distance: 6.86 mm

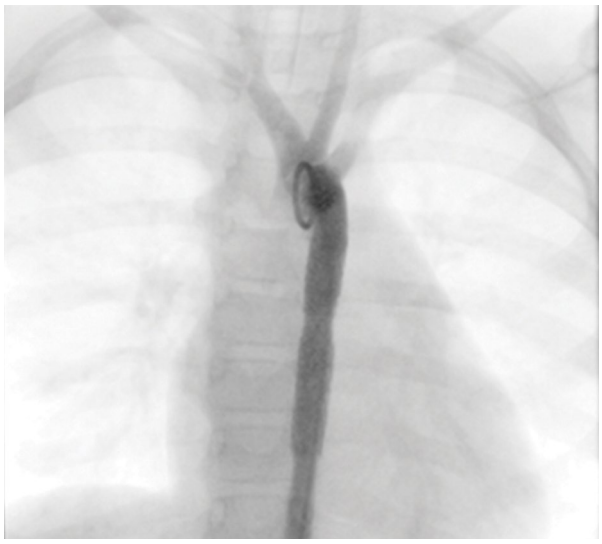

Figure 4. Angiography — the result after the final stent dilatation showing no residual stenosis

Conclusions

The CoA rarely occupies a non-typical region and therefore may be diagnostically challenging. A profound physical examination, particularly in a later childhood and COVID-19 pandemic settings, relevantly supplies the anamnesis in the patients who require dedicated strategy of treatment and may develop serious complications, if untreated.

Conflict of interest

None declared.

Streszczenie

Wstęp. Koarktacja aorty (CoA) to wrodzona wada serca definiowana jako zwężenie w okolicy cieśni aorty, klinicznie charakteryzująca się zaburzeniami perfuzji obwodowej w badaniu przedmiotowym. Większość pacjentów, u których rozpoznaje się CoA, stanowią noworodki, a korekcja chirurgiczna jest w tej grupie leczeniem z wyboru. Przezskórne interwencje kardiologiczne stanowią terapię rzadkich przypadków, w których objawy występują dopiero w kolejnych latach życia dziecka. Tętniak aorty, niewydolność serca oraz udar to główne powikłania nieleczonego CoA, co wyjaśnia niski wskaźnik przeżywalności niezdiagnozowanych pacjentów.

Opis przypadku. Podczas profilaktycznej kontroli pediatrycznej 5-letniej pacjentki w dobrym stanie ogólnym wykazano szmer nad sercem. Chorą skierowano do kardiologa dziecięcego, ale niestety ze względu na trwającą pandemię choroby koronawirusowej 2019 dostępna była tylko konsultacja telemedyczna, wykluczająca dokładne badanie przedmiotowe. Ostatecznie chorą skierowano na oddział kardiochirurgii dziecięcej w celu pilnej konsultacji.

Badanie echokardiograficzne wykazało ciągły, niepulsacyjny przepływ w aorcie brzusznej oraz zwężenie światła aorty zstępującej za lewym przedsionkiem. Angiografia tomografii komputerowej potwierdziła zwężenie światła aorty piersiowej (o długości 55 mm do średnicy 2,5 mm) z towarzyszącym krążeniem obocznym.

Ze względu na anatomię zmiany aorty pacjentkę zakwalifikowano do przezskórnej implantacji stent-graftu (Bentley BeGraft© 9 mm/57 mm) w hybrydowej sali operacyjnej z zapleczem krążenia pozaustrojowego. Wykonano wstępną dylatację aorty do 9 mm, z wyjątkiem obszaru o długości około 6 mm zwężonego do 5,2 mm. Po 2 miesiącach stent-graft ponownie poszerzono, a badanie echokardiograficzne potwierdziło dobry wynik zabiegu (średnica aorty piersiowej na całym odcinku 9 mm).

Przebieg pooperacyjny bez powikłań, pacjentkę wypisano do domu i skierowano na kontrolę ambulatoryjną.

Wnioski. Rzadko CoA obejmuje nietypowy obszar, wywołując trudne diagnostycznie objawy w warunkach pandemii i ograniczenia dostępności lekarzy specjalistów, szczególnie u starszych pacjentów pediatrycznych obciążonych ryzykiem wystąpienia poważnych powikłań bez wdrożenia odpowiedniego leczenia.

Słowa kluczowe: koarktacja aorty, wrodzone wady serca, kardiologia interwencyjna, kardiochirurgia dziecięca, leczenie hybrydowe

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