Letter to the Editor

Coarctation of the aorta, carotid artery stenosis and aberrant right subclavian artery as a rare cause of cerebral ischemia in a primigravid woman

Magdalena Wawak[1,2,3]^, Piotr Pieniążek[1,4,5]^, Łukasz Tekieli[1]^, Piotr Paluszek[4]^, Mariusz Trystuła[4]^, Tadeusz Przewłocki[1,5]^, Anna Kabłak-Ziembicka[3,6]^

[1]Department of Interventional Cardiology, the St. John Paul II Hospital, Kraków, Poland; [2]Jagiellonian University Medical College, Doctoral School of Medical and Health Sciences, Kraków, Poland; [3]Department of Interventional Cardiology, Institute of Cardiology, Jagiellonian University Medical College, Kraków, Poland; [4]Department of Vascular and Endovascular Surgery, the St. John Paul II Hospital, Kraków, Poland; [5]Department of Cardiac and Vascular Diseases, Institute of Cardiology, Jagiellonian University Medical College, Kraków, Poland; [6]Noninvasive Cardiovascular Laboratory, The St. John Paul II Hospital, Kraków, Poland

Correspondence to: Magdalena Wawak, MD. Department of Interventional Cardiology, the St. John Paul II Hospital, Prądnicka 80, 31-202 Kraków, Poland; Jagiellonian University Medical College, Doctoral School of Medical and Health Sciences, Kraków, Poland; Department of Interventional Cardiology, Institute of Cardiology, Jagiellonian University Medical College, Kraków, Poland. Email: wawakmagda@interia.pl; Prof. Anna Kabłak-Ziembicka, MD, PhD. Department of Interventional Cardiology, Institute of Cardiology, Jagiellonian University Medical College, Św. Anny 12, 31-008 Kraków, Poland; Cardiovascular Laboratory, The St. John Paul II Hospital, Kraków, Poland. Email: kablakziembicka@op.pl.

Submitted Jun 02, 2023. Accepted for publication Sep 26, 2023. Published online Nov 16, 2023.
doi: 10.21037/qims-23-792
View this article at: https://dx.doi.org/10.21037/qims-23-792

Case report

We present a case of a 24-year-old hypertensive primigravid woman operated on coarctation of the aorta (CoA) during her infancy, in whom severe stenosis of left common carotid artery (LCCA) and aberrant right subclavian artery (ARSA) were diagnosed. The CoA was managed with a Gore-Tex patch in her second month of life. She had repeated procedures with balloon angioplasty at the age of 4 and 18 that led to the residual pressure gradient of 20 mmHg. Computed tomography angiography (CTA) showed one of the aortic arch (AA) variants according to Adachi-Williams Classification (*Figure 1*) (1). In addition, LCCA showed severe (85%) stenosis 0.7 cm above ostium, while the minimal diameter of the aorta at CoA site was 11 mm.

The patient, in the 10th week of pregnancy, was referred to our outpatient clinic to offer her cardiovascular counseling. Color Doppler ultrasonography showed a peak systolic and the end-diastolic velocities of 600 and 143 cm/s respectively in LCCA, which was consistent with severe stenosis (*Figure 2*). She reported no neurologic symptoms and additional tests were within the normal range, including blood tests and neurologic examination, rest and a 24-hour electrocardiogram monitoring. Blood pressure values were 120/75 mmHg on bisoprolol 5 mg o.d.

The NeuroVascular Team, that included vascular and endovascular surgeons, radiologists, neurologist, cardiologist, and psychologist, referred the patient to carotid artery stenting (CAS). The consulting neurologist and obstetrician suggested postponing CAS until after the delivery. The patient was given aspirin in a dose of 75 mg o.d. since the 11th week of pregnancy.

According to the modified World Health Organization (mWHO) classification, the patient was in class II–III which is associated with an 8–22% maternal risk of cardiovascular

^ ORCID: Magdalena Wawak, 0009-0005-6457-7436; Piotr Pieniazek, 0000-0002-7688-7328; Lukasz Tekieli, 0000-0002-3431-1013; Piotr Paluszek, 0000-0003-1937-0928; Mariusz Trystuła, 0000-0002-9667-5082; Tadeusz Przewlocki, 0000-0002-9575-7270; Anna Kablak-Ziembicka, 0000-0002-8006-7840.

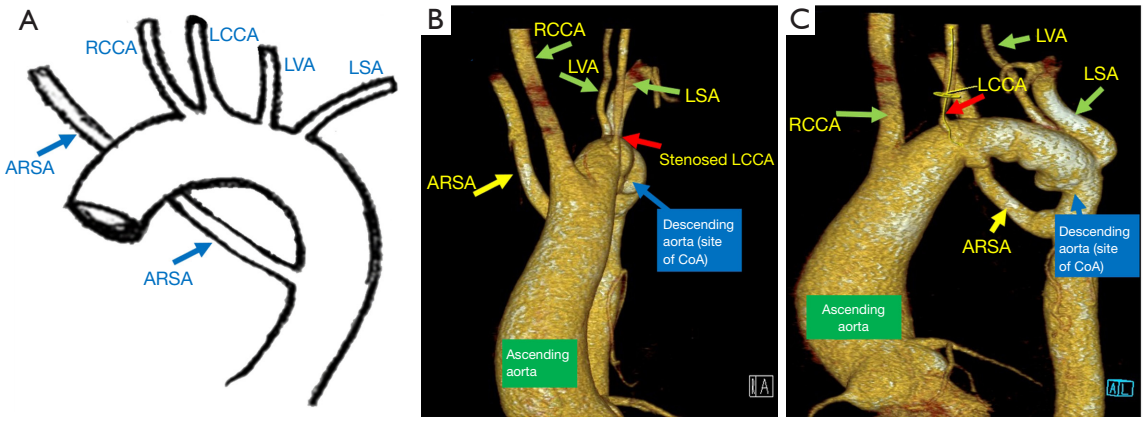

Figure 1 Configuration of the aortic arch identified in the patient. (A) Scheme of the aortic arch branching in the patient. In this configuration, the brachiocephalic trunk is absent. The RCCA arises as the first vessel from the aortic arch, followed by LCCA, LVA, LSA and ARSA. (B,C) Patient's CTA images. Site of the operated CoA (blue arrow). The sequence of the aortic arch branching is as follows: RCCA, LCCA which is stenosed (red arrow), LVA, LSA, and finally ARSA indenting the esophagus posteriorly. RCCA, right common carotid artery; LCCA, left common carotid artery stenosis; LVA, left vertebral artery; LSA, left subclavian artery; ARSA, aberrant right subclavian artery; CoA, coarctation of the aorta; CTA, computed tomography angiography.

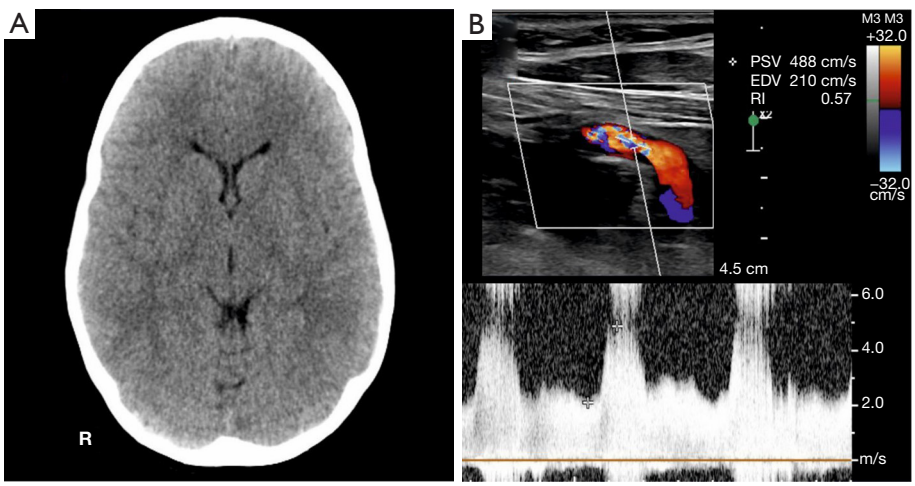

Figure 2 Diagnostic images obtained before the intervention on the LCCA. (A) Brain computed tomography scan confirming absence of infarct zone and excluding other non-neurovascular pathology; (B) color Doppler ultrasound showing abnormal turbulent flow in the proximal segment of LCCA with the peak systolic and end-diastolic velocities of 488 and 210 cm/s respectively, consistent with severe stenosis of the proximal LCCA. LCCA, left common carotid artery; PSV, peak systolic velocity; EDV, enddiastolic velocity; RI, resistive index.

event (2). Consistently, as a result of consensus between a cardiologist and an obstetrician, a caesarean section was recommended in the 37th week of pregnancy. The child was born without any complications.

A week after child delivery, the mother patient developed an episode of transient left hemisphere ischemia with symptoms of amaurosis fugax and aphasia. Brain CT scans showed no ischemic lesions (*Figure 2*). Neurological symptoms resolved within 2 hours. Subsequent neurologic examination showed scores of zero points for the modified Rankin Score (mRS) and the National Institutes of Health Stroke Scale (NIHSS). Repeated carotid Doppler ultrasonography confirmed former findings of severe common carotid artery stenosis in its proximal segment

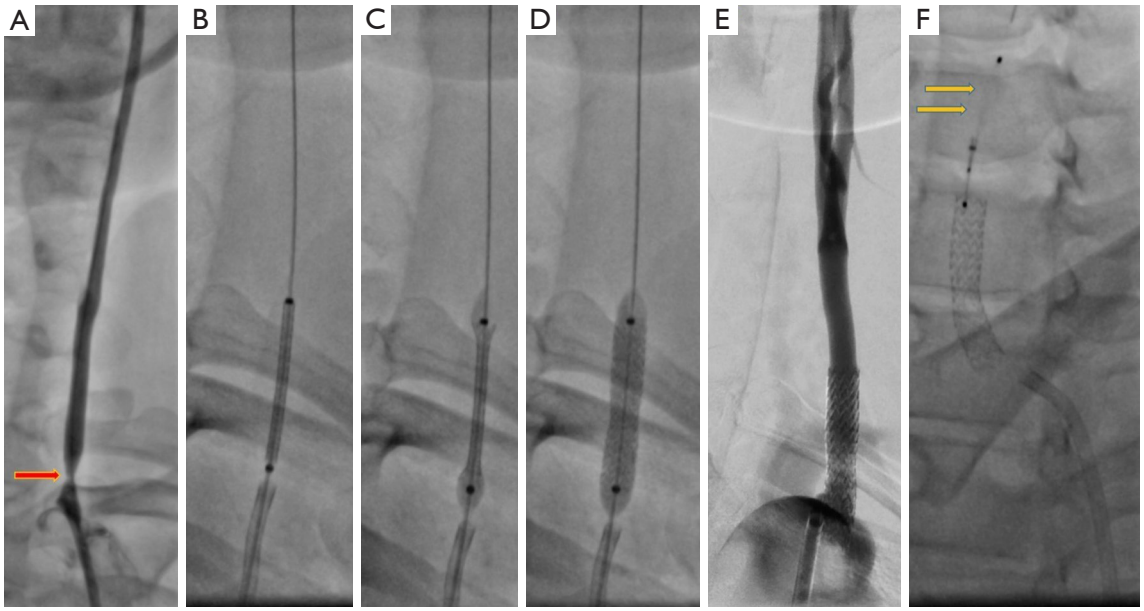

Figure 3 CAS of the LCCA. (A) Image before the procedure—stenosis of LCCA (arrow); (B-D) stent positioning and stent deployment; (E) the final result immediately after stent implantation (Omnilink Elite 6 mm ×29 mm); (F) the removal of the neuroprotection system (Spider FX 6 mm)—filter visible in the distal portion of the LCCA (arrows). CAS, carotid artery stenting; LCCA, left common carotid artery.

exceeding 90% according to Psychogios *et al.* criteria (*Figure 2*) (3).

Urgent CAS was performed with a typical dual antiplatelets therapy (a loading dose of clopidogrel 300 mg, followed by clopidogrel and aspirin 75 mg each). A standard heparinization at the time of procedure was given. In adherence to the guidelines, a neuroprotection system and direct stenting with a balloon-expandable cobalt-chromium stent were performed (*Figure 3*). The chrome cobalt stent (a balloon-mounted stent) was preferred in the common carotid artery because it was characterized with the highest radial force. Furthermore, it can be precisely positioned in ostial lesions, as opposed to the self-expandable stents that have significantly lower radial force and there are prone to dislocation during positioning.

At 6- and 12-month follow-up visits, a follow-up carotid Doppler ultrasonography confirmed the optimal result of CAS. The patient recovered without any residual neurologic deficits, and no new ischemic episodes were recorded during a follow-up period.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

CoA accounts for 4–8% of all congenital heart diseases (CHDs) and occurs in 4 out of 1,000 live births. CoA can occur as an isolated lesion, but is often associated with other cardiovascular defects, such as AA hypoplasia, aortic and mitral valve abnormalities, ventricular and atrial septal defects.

Here, we presented a young female patient who underwent several interventions on CoA. In addition to residual CoA, a severe stenosis of the LCCA was recognized in the first trimester of pregnancy. This finding is clinically relevant as even repaired CoA is associated with increased risk of cardiovascular events during pregnancy (2,4,5).

The reported patient with congenital AA anomaly and CoA represents one of 5 young female patients (0.065%), who underwent urgent endovascular procedure during the peri-partum and post-partum period in our registry of 82 (1.07%) out of 7,645 young patients who had endovascular procedure performed in our tertiary referral hospital in the department of vascular and endovascular surgery over the time period of 20 years (6). This demonstrates the rarity of

such cases. Nevertheless, stroke can complicate pregnancy, occurring in 30.0 per 100,000 pregnancies (7). Among pregnancy-related strokes, aortic dissection Stanford type A and type B contribute significantly to stroke cases (7). However, in this case the aortic dissection or rupture had not contributed to cerebral ischemia, despite the fact that CoA was reported a major risk factor for aortic rupture in peripartum.

Among stroke patients between 18 to 50 years old, also other vascular diseases, like congenital or iatrogenic artery stenosis, Takayasu disease, fibromuscular dysplasia, premature atherosclerosis, moyamoya disease, cerebral venous thrombosis, and reversible cerebral vasoconstriction syndrome have been more frequently found compared to non-young adults (6,8,9).

The LCCA stenosis imposes additional risk. Carotid artery stenosis is predominantly attributed to atherosclerotic etiology, which is responsible for 15–30% of total strokes, therefore it is an unexpected finding in a young woman. Whereas, approximately 5% to 15% of all strokes occur in adults aged 18 to 50 years, including 3–5% of all ischemic strokes in patients below the age of 50 years that are attributed to large AA branches pathology, therefore the risk they impose should not be neglected (10).

Risk of stroke recurrence is at its highest shortly after the first episode, within 2 weeks, then it decreases to 15% during the first 3 months. Therefore, when the neurological symptoms occurred in our female patient, the intervention on carotid stenosis had to be done without further delay. Of course, it can be debatable whether revascularization of LCCA was reasonable during pregnancy. In the described case, the risk of invasive procedure and the risk of watchful waiting policy should be balanced to avoid complications in mother and child (11). CAS procedure, with antiplatelet treatment and periprocedural heparin can increase the risk of bleeding and miscarriage. Moreover, mother and child's exposure to radiation during CAS is a main drawback of the procedure, unless there are life-threatening conditions that justify the risk. Increased pressure and hormonal changes during delivery can be associated with significant risk of dissection or stroke for the mother.

Conclusions

The coexistence of the AA anomaly, LCCA stenosis, and CoA is a rare condition, but nevertheless an important risk factor of complications in pregnant women. CAS is feasible and safe for patients when it is performed in tertiary centers. However, in gravid mothers, the risk of intervention versus risk of stroke accompanying child delivery must be considered. The awareness of risk factors management, a consensus on the way of the childbirth and time of intervention must be agreed upon by a multidisciplinary team including cardiovascular and obstetrician consultants.

Acknowledgments

Funding: This work was supported by the science fund of the St. John Paul II Hospital, Kraków, Poland (No. FN15/2023 to AKZ). Also, this work received external funding from the Jagiellonian University Medical College (No. N41/DBS/001247 to AKZ).

Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-23-792/coif). A.K.Z. reports that she has obtained funding from the St. John Paul II Hospital, Kraków, Poland and from the Jagiellonian University Medical College, Kraków, Poland. The other authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Hanneman K, Newman B, Chan F. Congenital Variants and Anomalies of the Aortic Arch. Radiographics 2017;37:32-51.
2. van Hagen IM, Roos-Hesselink JW. Pregnancy in congenital heart disease: risk prediction and counselling. Heart 2020;106:1853-61.
3. Psychogios K, Magoufis G, Kargiotis O, Safouris A, Bakola E, Chondrogianni M, Zis P, Stamboulis E, Tsivgoulis G. Ultrasound Assessment of Extracranial Carotids and Vertebral Arteries in Acute Cerebral Ischemia. Medicina (Kaunas) 2020;56:711.
4. Silversides CK, Grewal J, Mason J, Sermer M, Kiess M, Rychel V, Wald RM, Colman JM, Siu SC. Pregnancy Outcomes in Women With Heart Disease: The CARPREG II Study. J Am Coll Cardiol 2018;71:2419-30.
5. Mandalenakis Z, Rosengren A, Lappas G, Eriksson P, Hansson PO, Dellborg M. Ischemic Stroke in Children and Young Adults With Congenital Heart Disease. J Am Heart Assoc 2016;5:e003071.
6. Wawak M, Tekieli Ł, Badacz R, Pieniążek P, Maciejewski D, Trystuła M, Przewłocki T, Kabłak-Ziembicka A. Clinical Characteristics and Outcomes of Aortic Arch Emergencies: Takayasu Disease, Fibromuscular Dysplasia, and Aortic Arch Pathologies: A Retrospective Study and Review of the Literature. Biomedicines 2023;11:2207.
7. Swartz RH, Cayley ML, Foley N, Ladhani NNN, Leffert L, Bushnell C, McClure JA, Lindsay MP. The incidence of pregnancy-related stroke: A systematic review and meta-analysis. Int J Stroke 2017;12:687-97.
8. Van der Niepen P, Robberechts T, Devos H, van Tussenbroek F, Januszewicz A, Persu A. Fibromuscular dysplasia: its various phenotypes in everyday practice in 2021. Kardiol Pol 2021;79:733-44.
9. Wang N, Shen X, Zhang G, Gao B, Lerner A. Cerebrovascular disease in pregnancy and puerperium: perspectives from neuroradiologists. Quant Imaging Med Surg 2021;11:838-51.
10. Putaala J, Metso AJ, Metso TM, Konkola N, Kraemer Y, Haapaniemi E, Kaste M, Tatlisumak T. Analysis of 1008 consecutive patients aged 15 to 49 with first-ever ischemic stroke: the Helsinki young stroke registry. Stroke 2009;40:1195-203.
11. Tekieli Ł, Musiałek P, Kabłak-Ziembicka A, Trystuła M, Przewłocki T, Legutko J, Dzierwa K, Maciejewski D, Michalski M, Pieniążek P. Severe, recurrent in-stent carotid restenosis: endovascular approach, risk factors. Results from a prospective academic registry of 2637 consecutive carotid artery stenting procedures (TARGET-CAS). Postepy Kardiol Interwencyjnej 2019;15:465-71.

Cite this article as: Wawak M, Pieniążek P, Tekieli Ł, Paluszek P, Trystuła M, Przewłocki T, Kabłak-Ziembicka A. Coarctation of the aorta, carotid artery stenosis and aberrant right subclavian artery as a rare cause of cerebral ischemia in a primigravid woman. Quant Imaging Med Surg 2024;14(1):1261-1265. doi: 10.21037/qims-23-792

