Status post thrombosis of the inferior vena cava (IVC), left common iliac vein and Phlebothrombosis of the left femoral vein
(Suspicion for the primary antiphospholipid syndrome - APS)

CASE REPORT

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Case report

• Romani girl at the age of 14 years
• At 22/02/2012. she was received to our Clinic under Dg: Thrombosis of the inferior vena cava (IVC), left common iliac vein and suspicion for the primary antiphospholipid syndrome (APS).
• The disease has begun on 17th February 2012.
• Main symptoms were: abdominal pain, swelling, paleness and soreness of the left leg.
• Vital parameters were normal. The values of blood pressure were ranged from 126/87mmHg to 141/95mmHg.
• During recent viral infection of upper respiratory tract, she had a swelling of the left salivary gland.
• FAMILY HISTORY: Mother has epilepsy.
• Laboratory findings: elevated CRP 104.5, ESR 55/, urea 7.6, creatinine 139.
• APTT 32.3, D-dimers 3150 (N <500). Proteinuria 5 +.
• Cystatin C 1.18 (reference interval 0.50-0.96)
• Homocysteine 12.0 (reference interval 4-11.5)
• Protein C 1.46 (reference interval 0.64-1.28)
• Protein S 1.30, Factor VIII 1.69, Activated protein C 2.8.
• Factor V Leiden mutation, Antithrombin III activity: normal findings.
• Mutation in the prothrombin gene G20210A and MTHFR mutation were not done due to objective reasons - the inability of parents to pay for the required searches.
• Thromboelastometry (TEM): Parameters of thromboelastometry (TEM) show unabated hemostatic capacity with limited reserves.
Diagnostic Imaging

- **MRI of the Abdomen (27.02.2012):** A comprehensive finding of multiorganic infarction, and difficult venous drainage with visible edema of parenchyma organs. Is noticeable discrepancy between overfilled hepatic veins and intra-abdominal inferior vena cava, which has a diameter up to 0.5 cm.

- Distinct discrepancy between the upper pole of the left kidney, which is voluminous and the lower pole of the left kidney, which is a consequence of left partial atrophy, the most likely on the basis of infarction. Along the upper pole of the left kidney is less visible subcapsular hematoma.

- Within the spleen, it is visible presence of subcapsular hematoma, in the lower pole of the spleen with diameter of 8 cm.

- Overfilled collateral veins, as in the abdominal, as well in the lumbar veins area. It is visible expressive atrophy of the right kidney, the left perirenal edema as multilocular hiperintense zone fluids, which completely surrounds the left kidney, also less visible subcapsular hematoma.

- Conclusion: This is a coagulopathy with massive venous thrombosis, including VCI, until the confluence of the hepatic veins. The described plexus of venous vessels with strong flow are collateral veins for drainage from the lower body through the vein azygos and lumbar veins.

- **ECHO (24.02.2012):** No effusion, vegetation and signs of thrombosis.

- Patient was on oral anticoagulant therapy, Sintrom by scheme (I, II and III day, 1/2-1/2-1/4) until further, a maximum of 6 months.

- We have no information how long she has been on the recommended therapy. She did not answer the scheduled controls.
Control laboratory after two years of diagnosis (the girl did not come under controls)

- albumin 0.51 (ref int 0.53-0.64), globulin 0.49 (ref int 0.36-0.47), A/G 1.04 (ref int 1.13-1.78), α1-globulin 0.03, α2 globulin 0.13 (ref int 0.07-0.10), β globulin 0.18 (ref int 0.09-0.13), γ-globulin 0.15.

- **Rheuma tests:** ASO 318 (ref int <200), CRP <3.14, RF <10.2, Waler Rose test is negative.

- **Immunoglobulins:** IgG 10.5, IgA 3.64 IgM 1:34, IgE 73.7.

- **Immunological tests:** Antiphospholipid antibodies:
  - Anti cardiolipin IgG 27.12 (20-40 middle pos.),
  - Anti cardiolipin IgM 15.23 (10-20 weakly pos.),

- Anti β2 glycoprotein IgG 1.94, Anti β2 glycoprotein IgM 0.70.

- **ENA 6 Profile:** Anti SS A negative, Anti SS B negative, Anti Sm negative, Anti Sm /RNP negative, Anti Jo 1 negative, Anti Scl 70 negative.

- **ANA-ANF IFpositive,**

- Anti dsDNA IF negative. C3 1.41, c4 0.438, C1q-CIC 39.84.

- **Color Doppler of the lower limb deep vein system (12/02/2014):** The finding at the lower limb deep vein system indicates the state after iliacofemoral deep vein thrombosis of the left leg, now almost completely recanalised with a secondary dilated varices at the left thigh.
Diagnostic Imaging

- **Ultrasound of the abdomen and pelvic (05.02.2014.):** indicates on splenomegaly with the infarction zone more towards the lower pole, atrophic right kidney, enlarged left kidney, which is uneven contour, echogenic parenchyma with unclear corticomedullary differentiation and dilated channel system, predominantly hypoechogenic heterogeneous liquid formation around the lower pole of the left kidney. Right and cranial from the bladder, it is the rounded heteroehcogenic formation with small cystic formations, 4x3.1 cm in diameter, which in the differential diagnosis could correspond to inflammatory changed right adnexa.

- **CT phlebography of the inferior vena cava and common iliac vein:** the finding indicates on the common iliac vein thrombosis, right after the confluence of external iliac vein and left internal iliac vein, distinctly reduced diameter of the inferior vena cava caudally of the hepatic portion, and distinctly low contrast opacification of this segment. Visible conical constriction of the left renal vein, right before the confluence to the inferior cava vein and proximal of the described constriction is vein dilation. Visible portal vein and splenic vein dilatation. Drainage of the lower parts is taken through highly developed porto - systemic and gastroenteric collateral network and tortuous and dilated vein azygos.
Conclusion

• We believe that in this case, the diagnosis is **Thrombophilia**, probably congenital thrombophilia with massive venous thrombosis, including IVC, to the confluence of the hepatic veins.
• We assume that the possible trigger for existing condition was a genetic predisposition to thrombophilia associated with infection.
• The findings at the lower limb deep vein system indicate the state after deep vein thrombosis of the left leg, now almost completely recanalised with a secondary dilated varices at the left thigh, and the development of collateral veins for drainage from the lower body through the vein azygos and lumbar veins.
• Also, there were diagnosed sactosalpinx, dysfunction and atrophy of the right kidney, arterial hypertension, status post infarction of the liver, spleen and kidney.
• In the further, the girl will be controlled by a multidisciplinary team (nephrologist, hematologist, immunologist and gynecologist).
• In this case, we did not confirm the diagnosis of congenital thrombophilia due to indifference of parents to conduct diagnosis until the end, ie. to do mutation in the prothrombin gene G20210A and MTHFR mutation.
• For us, this is an interesting case, because the child has been studied long and inexhaustible in the direction of antiphospholipid syndrome, which is clinically and diagnostically excluded.
• The girl is in a good general condition.
Thank you for your attention!
For additional information and questions you can contact me at the email address: a.tunic@yahoo.ca