Neonatal cerebral thrombophlebitis revealing protein C deficiency

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INTRODUCTION

- Vascular thromboses are rare in the newborn but most often very serious. Their occurrence is partly conditioned by peculiarities of coagulation and fibrinolysis. Their diagnosis is better and better ensured by imaging techniques.
- We report a clinical case of cerebral thrombophlebitis; revealing a deficiency of protein C, hospitalized in the neonatal resuscitation department, CHU Mohammed VI, of MARRAKECH.
- **Objectives:**
 - Describe the diagnostic elements,
 - Discuss what to do in the neonatal period.

OBSERVATION

- It was a newborn female,
- Age: 21 days,
- Admitted for a state of convulsive illness in a feverish context,
- On examination: the newborn was not very responsive, with axial and peripheral hypotonia, the anterior fontanel was bulging and the suction reflex was absent,
- The lumbar puncture was sterile,
- The initial infectious balance was negative,
- The cerebral CT scan revealed multiple cerebral, hemorrhagic, bilateral frontal, thalamus and basal ganglia with intraventricular hemorrhage and cerebral thrombophlebitis at the level of the longitudinal sinus: (Fig.1,2).
- The search for thrombophilia revealed a partial defect in protein C with a rate of 40%.
- Magnetic resonance imaging showed extensive cerebral venous thrombosis with abnormalities of the parenchymal signal and Dandy Walker type malformation (Fig.2, 3).
- Therapeutic management consisted of initial probabilistic antibiotic therapy; with phenobarbital.
- The immediate evolution was marked by the disappearance of the convulsions and a clinical improvement.
- The follow-up at 5 months was marked by a delay of psychomotor acquisitions.

Figure (1; 2): Cerebral CT showing hemorrhagic stroke and cerebral thrombophlebitis

Figure (2,3): Cerebral MRI showing extensive CTV with parenchymal signal abnormalities

DISCUSSION

- Venous thromboembolism is a major cause of morbidity and mortality in pediatrics. In the neonatal period, thrombotic accidents cause constitutional abnormalities of hemostasis, including congenital protein C deficiency.
- The most common expression of the disease in the neonatal period is renal vein thrombosis, cerebral venous thromboses are rare (our case).
- Seizures are the most common mode of revelation of neonatal cerebral thrombophlebitis.
- The diagnosis of vascular thromboses is based on imaging.
- The etiological diagnosis is based on a decrease in the level of protein C. This rate varies between 0 and 30% in severe deficits and between 30 and 70% in partial deficits.
- The prevalence of the severe deficit is estimated at 1 / 500,000. Partial deficits are much more frequent (1/200 to 1/500).
- The treatment still escapes any codification.

CONCLUSION

- In the case of neonatal thrombosis, the search for a deficit in anticoagulant factor, in particular protein C, is required in the newborn.
- The therapeutic behavior is not codified.
- An individualized approach seems adapted to this very rare clinical situation.

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