International Archives of Pediatrics & Neonatology

Case Report

doi: 10.39127/2575-8253/IAPN:1000107
Tahiri F, et al. In Arch Pedia Neon: IAPN-107

Cerebral Thrombophlebitis Revealing a Familial Seronegative Anti-Phospholipid Syndrome: About A Pediatric Case

F. TAHIRI[1.2], Z. ALAHIANE[1.2], H. NASSIH[1.2], R. EL QADIRY[1.2], A. BOURRAHOUAT[1.2], I. AIT SAB[1.2]

[1]Department of Pediatrics B, CHU Mohamed VI, MARRAKECH Morocco
[2]Faculty of Medicine and Pharmacy MARRAKECH Morocco

Corresponding author: Tahiri Fatima Ezzahra, Department of Pediatrics B, CHU Mohamed VI, MARRAKECH Morocco. Email: fatimaezzahratahiri29@gmail.com

Citation: Tahiri F, Alahiane Z, Nassih H, EL Qadiry R, Bourrahouat A, Sab IA (2021) Cerebral Thrombophlebitis Revealing a Familial Seronegative Anti-Phospholipid Syndrome: About A Pediatric Case. In Arch Pedia Neon: IAPN-107.

Received Date: 15 September, 2021; **Accepted Date:** 20 September, 2021; **Published Date:** 27 September, 2021

Summary

Cerebral thrombophlebitis remains a significant cause of stroke in children; antiphospholipid syndrome (APS) is exceptionally a cause.

Observation: We report the case of a 12-year-old patient, with a family history of thrombosis, admitted to our clinic with an ICD, cerebral CT and cerebral MRI were in favor of a recent cerebral venous thrombophlebitis in the right lateral and sigmoid venous sinus as well as in the superior, with a right temporo-occipital hematoma. An etiological workup did not reveal any abnormalities. Treatment was based on heparin therapy.

Conclusion: Antiphospholipid syndrome (APS) or Hughes syndrome remains a fairly rare cause of thrombophlebitis in children but should be systematically sought in the presence of family history.

Introduction

Cerebral thrombophlebitis (CT) is related to the occurrence of a thrombus within a cerebral venous structure. It is a rare condition, particularly in children, but a serious one with an incidence of 0.4 to 0.7 per 100,000 children per year. It is characterized by a great diversity in its clinical presentation as well as by its numerous etiologies. Advances in imaging, particularly cerebral magnetic resonance imaging associated with magnetic resonance venous angiography, have led to diagnostic and therapeutic advances. Heparin therapy combined with treatment of intracranial hypertension (ICHT) is the cornerstone of treatment [1,2,3].

Medical Observation

A 12-year-old patient from a non-consanguineous marriage, with a father who died at age 35 from an undocumented DVA, two paternal cousins with anti-phospholipid syndrome secondary to lupus, a paternal uncle with renal artery thrombosis, and two paternal aunts with recurrent miscarriages.

Our patient was initially admitted with an ICD, a cerebral CT scan showed cerebral thrombophlebitis of the right transverse sinus complicated by a right temporal intra parenchymal hematoma. A complementary cerebral MRI was in favor of a recent cerebral venous thrombophlebitis of the right lateral and sigmoid venous sinuses as well as the superior one, with a right temporo-occipital hematoma. A haemostasis work-up was done and no abnormalities were found. A complete autoimmune workup, including anti-phospholipid antibodies, was performed on our patient and was negative. A homocysteine assay was normal and the ophthalmological examination was normal. The genetic study was not done because of the unavailability in our country. The treatment was based on heparin therapy then relayed by anti-vitamin K after obtaining a correct INR. No complication was noted in our patient after the initiation of anticoagulant treatment.

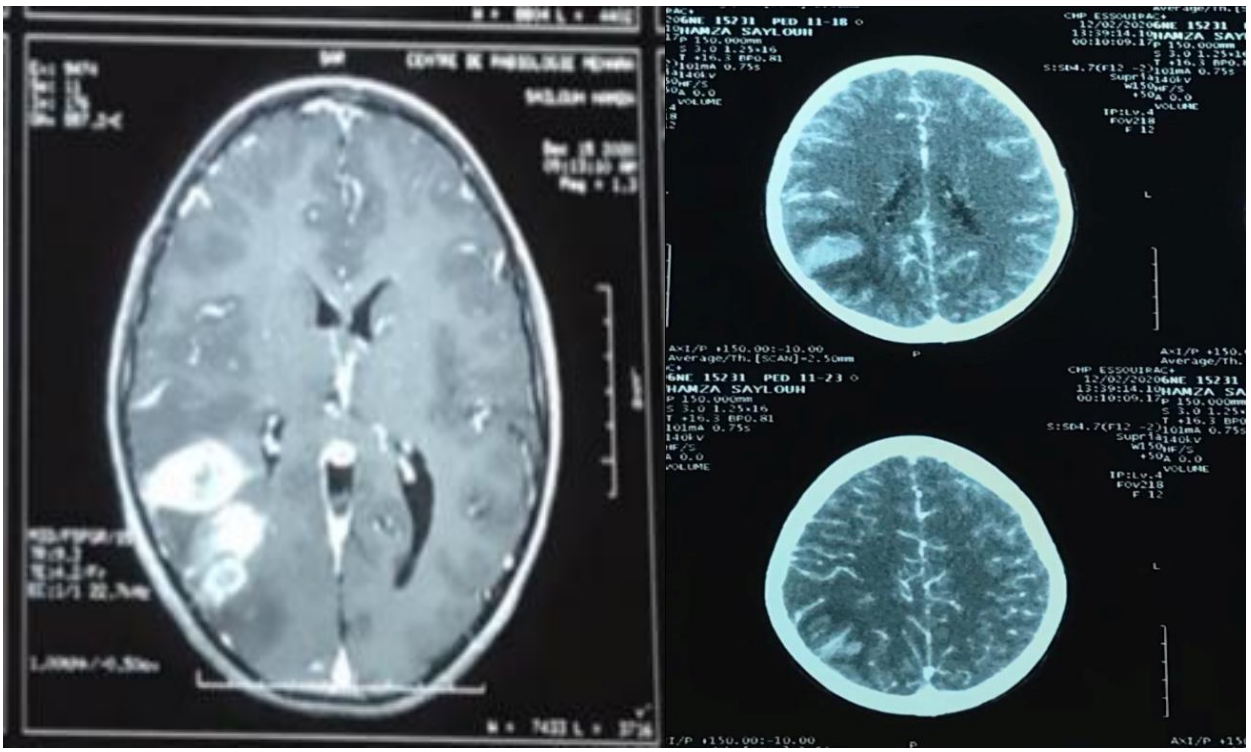
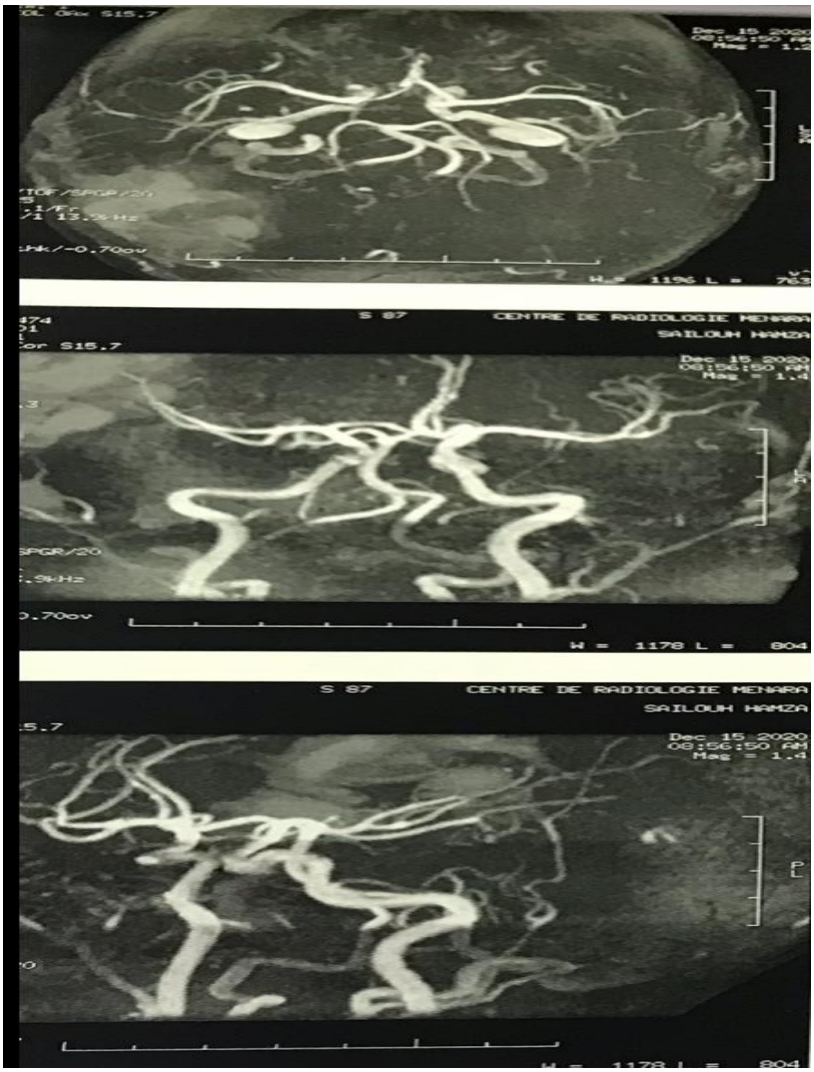


Figure 1: cerebral scan: cerebral thrombophlebitis of the right transverse sinus complicated by right temporal intraparenchymal hematoma.



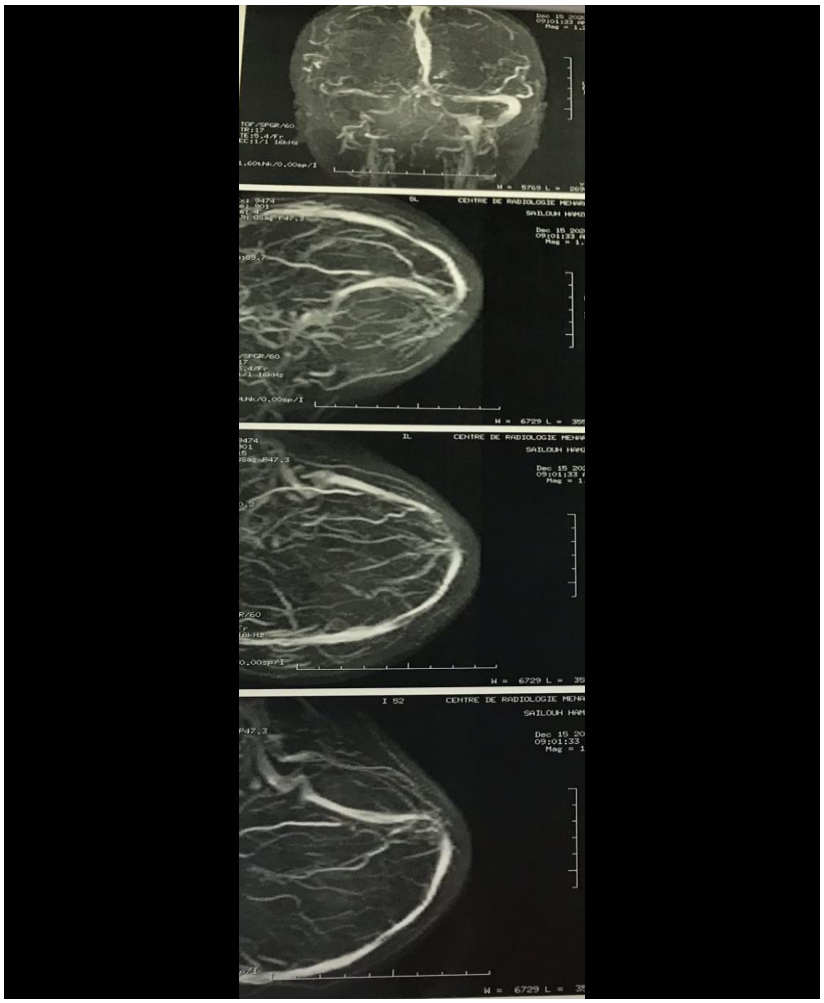


Figure 2: Angio-Mri: Recent Cerebral Venous Thrombophlebetis in The Right Lateral and Sigmoidal Venous Sinues As Well As The Superior One, With A Right Temporo-Occipatal Hematoma.

Discussion

Cerebral venous thrombosis (CVT), also known as *cerebral thrombophlebitis, is the* obstruction of a vein or venous sinus of the brain by a clot (thrombus). It is the only type of stroke defined by the vascular lesion itself, and not by its consequence in the brain parenchyma, which can be ischemic and/or hemorrhagic [1,2]. This multifactorial condition manifests itself through a wide range of signs and symptoms making its diagnosis difficult.

Pathophysiology: The cerebral venous drainage network is divided into two major systems: a superficial and a deep system, the second being generally more involved. This condition of multifactorial origin can be differentiated into cerebral venous thrombosis of infectious and non-infectious origin. Sinusitis, otitis or mastoiditis can cause cerebral venous thrombosis by continuity. These infections appear to be the most common cause in children. Other factors of non-infectious origin may promote the occurrence of venous thrombosis by affecting at least one of the parameters of Virchow's triad (hemodynamic variations, alteration of the endothelium or hypercoagulability) [5].

Cerebral lesions may be induced by hypoxia and ischemia, parenchymal edema, secondary intracranial hypertension or even hemorrhagic transformation. The numerous and varied clinical presentations that result can therefore confuse the clinician. Diagnosis is based on CT or MRI with arteriography, which are currently the reference methods for the investigation of cerebral venous thrombosis.

Etiologies: The discovery of familial or constitutional thrombophilia dates back to 1950. In 1965, Egeberg described the first case of antithrombin III deficiency, and in 1969 Mc Cully associated hyperhomocysteinemia with an abnormal frequency of DVT. In 1980, protein C and protein S deficiencies were described. In 1992, Dahlback described activated protein C resistance (APCR) in a family; APCR is responsible for about 20% of DVTs. In 1996, Sun confirmed that factor V is involved in RCPA and the addition of an abnormal factor V confers RCPA. Bertina was responsible for the discovery of the mutation of a nucleotide, codon CGA coding for arginine 506 to CAA coding for glutamine. This Arg 506/Gln mutation is called factor V Leiden, anti-phospholipid syndrome being a very rare cause in pediatrics [1].

ANGIO-MRI: Recent cerebral venous thrombophlebitis in the right lateral and sigmoidal venous sinuses as well as the superior one, with a right temporo-occipital hematoma.

APS is an autoimmune disease (related to a disorder of the immune system) characterized by recurrent blood clots in the vessels (arterial and/or venous thrombosis), repeated miscarriages in women and the presence of particular antibodies in the blood, the anti-phospholipid antibodies. Other clinical manifestations, such as skin, neurological and renal disorders, are often associated with it. When thrombosis and the presence of anti-phospholipid antibodies occur in isolation, without any other associated autoimmune disease, it is called primary antiphospholipid syndrome. In some cases, these manifestations are associated with other autoimmune diseases (such as systemic lupus): this is called secondary APS. APS is sometimes called Hughes syndrome. It is very difficult to estimate its prevalence in the pediatric population due to its rarity [6].

Antiphospholipid antibodies (aPL) represent a complex of autoantibodies (lupus-like circulating anticoagulant ACC, anticardiolipin (ACL) and anti-b2 type I glycoprotein (anti-ß2GPI).

The particularities of our patient were the negativation of the immunological assessment with family history allowing to retain the diagnosis of seronegative anti-phospholipid syndrome, a very rare form in the reviews of the literature.

The genetic study remains the only means for the diagnosis of certainty but it is not done because of the lack of availability in our country.

Therapeutically, anticoagulation is essential to stop the extension of the thrombus, promote its fibrinolysis and avoid pulmonary embolism. Intravenous unfractionated heparin is generally preferred in the acute phase because its effects may be reversible in the event of hemorrhage. Subcutaneous LMWH is then started for 3 to 6 months. Thrombolysis and thrombectomy have also been used successfully in this indication but are currently confined to the second-line therapeutic arsenal [6].

The treatment of APS still raises many questions. It is mainly based on antiplatelet agents and anticoagulants [7].

Conclusion

Cerebral thrombophlebitis remains 100 times rarer in children than in adults. It often occurs in the context of infection, neoplasia, or cardiac malformations; autoimmune diseases are very rare. Radiological diagnosis is currently based on non-invasive examinations, notably angioscan and angiomicrograph [3].

APAS in children should be considered in the setting of an ischemic stroke once the usual causes have been ruled out.

References

1. Cerebral thrombophlebitis in the pediatric intensive care unit (About 16 cases), THESIS NUMBER 231 Faculty of Medicine and Pharmacy Marrakech Morocco 2019.
2. Cerebral thrombophlebitis: clinical aspects, diagnosis and treatment C. Arquizan* Neurology Department, Sainte-Anne Hospital, 1, rue Cabanis, 75674 Paris cedex, France.
3. RP-WS-28 Deep vein thrombosis in pediatrics (about 52 cases) Author links open overlay panel R.SaouabR.Dafiri
4. Cerebral *venous thrombosis in children: about a series of 12 cases.*
5. CEREBRAL VENOUS THROMBOSIS IN CHILDREN C. Barrea1, S. Vaessen2, P. Leroy 2, J.P. Misson3 CHR Citadelle de Liège Hospital, (1) Assistant in Pediatrics, (2) neuropediatrician at the CHR Citadelle de Liège, (3) professor and head of the Pediatrics Department at the CHR Citadelle de Liège.
6. Antiphospholipid syndrome: a new pediatric case report
I. Chabchoub [a, A]. Ben Thabet [a], I. Maaloul [a], H. Aloulou [a], T. Kamoun [a], H. Daoued [b], Z. Mnif [b], M. Hachicha [a]
[a] Department of general pediatrics, Habib-Bourguiba hospital, Sfax, Tunisia
[b] Radiology Department, Habib-Bourguiba Hospital, Sfax, Tunisia
7. Antiphospholipidsyndrome thesis number 16 Faculty of Medicine and Pharmacy MARRAKECH MOROCCO 2018.
8. Anticoagulant treatment for cerebral venous thromboses in children and newborns. Les recommandations de la Société' franc¸aise de neurologie pe'diatrique (SFNP) Anticoagulant treatment for cerebral venous thromboses in children and newborns. French Society of Pediatric Neurology guidelines A. Lebasa,b , S. Chabrierc, *, M. Tardieub , M. Kossorotoffd.
9. Cerebral venous thrombosis in children : S.Chaouki S.AbourazzakF. Z. Souilmi M. Lakhdar Idrissi S. Atmani M. Hida.