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S. Di Filippo, R. Riva, J. C. Lega, A. Belot

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Queries for progressive cerebral and systemic vasculitis

Laura Olivier-Gougenheim, MD¹, Caroline Freychet, MD¹, Sophie Collardeau-Frachon, MD, PhD², Chantal Roure-Sobas, MD³, Sylvie Di Filippo, MD, PhD⁴, Roberto Riva, MD⁵, Jean-Christophe Lega, MD, PhD⁶, Alexandre Belot, MD, PhD¹

¹ Pediatric Rheumatology, Nephrology and Dermatology Department, Hôpital Femme-Mère-Enfant, Hospices civils de Lyon, Lyon, France. Université Claude Bernard Lyon 1, France.

² Pathology Department, Hôpital Femme-Mère-Enfant, Hospices Civils de Lyon, Lyon France ; Université Claude Bernard Lyon 1, France.

³ Microbiology Department, Hopital Croix Rousse, Hospices Civils de Lyon, Lyon, France, Université Claude Bernard Lyon 1, France.

⁴ Paediatric and Congenital Cardiology Department, Louis Pradel Hospital, Hospices civils de Lyon, Lyon, France. Université Claude Bernard Lyon 1, France.

⁵ Interventional neuroradiology Department, Pierre Wertheimer Hospital, Hospices civils de Lyon, Lyon, France. Université Claude Bernard Lyon 1, France.

⁶ Internal and Vascular Medicine Department, Centre Hospitalier Lyon Sud, Hospices Civils de Lyon, Lyon, France. Claude Bernard University Lyon 1, France.

Corresponding author:

Pr Alexandre Belot

Adresse Pediatric Nephrology, Rheumatology, Dermatology Unit, Hopital Femme Mère Enfant, Hospices Civils de Lyon, INSERM U1111, Lyon, France

Email: alexandre.belot@chu-lyon.fr

Tel + 33 4 27 85 64 81

A 9-year-old girl was admitted for a status epilepticus; neuroimaging revealed an intra-parenchymal hematoma and intraventricular hemorrhage. She was born with a complex congenital cardiopathy requiring ventriculopulmonary tube (RV-PA) within the first week of life. CNS arteriography showed a right sylvian artery aneurysm (Figure 1A). Despite being afebrile, endocarditis with mycotic CNS aneurysm was considered due to the presence of intracardiac prosthetic material and elevated inflammatory markers. There was no phenotypic trait for genetic collagen disorders. Echocardiography, and thoraco-abdomino-pelvic scans were normal. Successive blood cultures, universal PCR as well as Human Herpesvirus 1-5 PCR were all negative. The aneurysm was embolized; The hematoma was drained. Empiric antibiotic therapy was administered for six weeks. She recovered with a mild left residual hemiparesis.

One month later, she presented with diabetes, followed by an acute hepatitis. A systemic inflammatory disease was considered but specific autoantibodies (anti-GAD, anti-IA2, anti-ZnT8, ANA, anti-LKM, ANCA, AMA) were negative. Liver biopsy revealed features of non-specific subacute hepatitis without fibrosis nor granuloma. Of note, peripheral naïve T cells were low as a possible consequence of early thymectomy and an inflammatory complication of immunodeficiency. 22q11.2 deletion syndrome was ruled out.

On three months follow-up, a new cerebral aneurysm was detected; this time involving the contralateral left sylvian artery (Figure 1B). This aneurysm was again embolized and the RV-PA conduit was changed.

Cultures of the explanted material remained sterile. Infectious evaluation was repeated and this time, included Q fever serology, which was positive as well as the PCR in the liver biopsy. The diagnosis of chronic Q fever was made. A one-year treatment with doxycycline and hydroxychloroquine was started. The girl made a complete recovery.

There are numerous causes of non-traumatic cerebral aneurysms in children comprising Marfan, vascular Ehlers-Danlos syndrome or fibromuscular dysplasia, primary cerebral / systemic vasculitis or infections. Negative blood cultures imply also consideration for fungi, staphylococcus and streptococcus variants, the gram-negative bacilli of the HACEK group, Bartonella species, Tropheryma whipplei and Coxiella burnetii.

Coxiella Burnetii is the intracellular pathogen causing the “query” fever, a zoonotic disease transmitted by dust aerosol of some contaminated animals (goat, sheep and cattle). Q Fever should be considered when querying multisystem disease with cerebral vasculopathy, even in the absence of fever.

Question:

A 9-year-old girl with a past medical history of congenital heart disease and early cardiac surgery presents with

intra-parenchymal hemorrhage associated with an isolated cerebral aneurysm. She is afebrile, successive blood cultures and universal PCR remain negative but inflammation markers were increased (CRP=45mg/l and ESR=50mm/h). Three months later, the child represents with acute hepatitis and a contralateral cerebral aneurysm.

Query: What is the diagnosis?

Staphylococcal infection

Q Fever

Systemic Poly/Panarteritis nodosa (PAN)

Kawasaki disease

Contributors

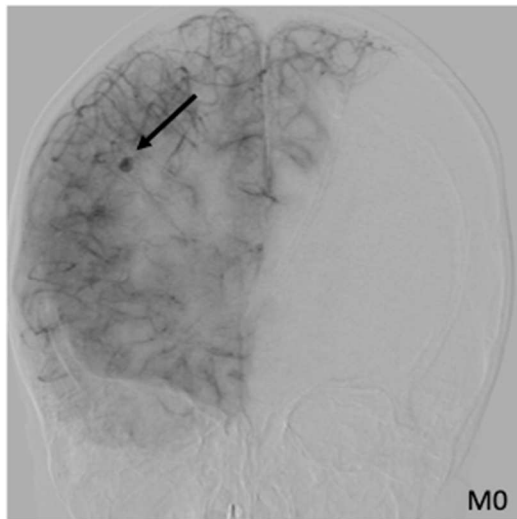
We each substantially contributed to the clinical management of the patient and to the writing up of the manuscript.

Declaration of interests

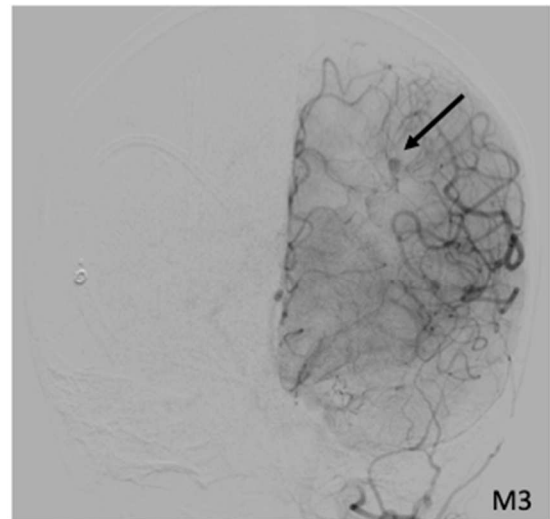
We declare no competing interests.

Figure 1: Cerebral Angiographies

(A) Cerebral arteriography at admission. Frontal cut of the brain revealing a right sylvian artery aneurysm. (B) follow-up cerebral arteriography three months later. Frontal cut revealing a new aneurysm of the posterior-parietal branch of the left sylvian artery.



A



B