Cerebral venous thrombosis in a child with Behçet's disease: a complication to bear in mind also in children

Sirs,

Behçet's disease (BD) is a rare multisystemic inflammatory disease primarily characterised by recurrent mucocutaneous along with ocular, musculoskeletal, vascular, gastrointestinal, and central nervous system (CNS) manifestations (1-3). BD is a vasculitis involving all size of vessels, but prominently vein. The onset in paediatric age is rare accounting to the 4–26% of cases (2-4). Even though the mucocutaneous involvement is the most common manifestation, the neurological and vascular involvement are among the most serious.

Our aim is to present a case of a boy with a diagnosis of BD which debuted with a cerebral venous thrombosis.

A 15-year-old-boy was referred to our hospital for a one-year history of recurrent fever, chest pain and oral and genital aphthous ulcers. Blood tests showed raised inflammatory markers (ESR 110 mm/h, CRP 9.21 mg/dl), and WBC 17.700/ml, whilst extensive infectious workup was negative. In the suspicion of BD, ophthalmological evaluation was performed and showed papillitis in the right eye. The pathergy test was positive and HLA B51 was negative. Based on the eye findings and persistent headache, a brain MRI with contrast was performed revealing an extensive thrombosis of the internal jugular, sigmoid sinus and transverse sinus up to the proximity of the straight sinus of the right side (Fig. 1A and 1C). Three pulses of methylprednisolone (1 g) were then administered, along with colchicine (1 mg/die) and infliximab (400 mg at 0, 2 weeks and every 4 weeks). Extensive thrombosis also required heparin at 100 UI/kg/day. Patient progressively improved, thus corticosteroids were slowly tapered. At 3 month-follow-up, he experienced a complete clinical and radiological recovery of the extensive thrombosis of the internal jugular and sinus (Fig. 1B and 1D). Although rare, physicians should be aware that BD may show several complications (3, 5). In order to avoid serious complications, cerebral venous sinus thrombosis (CVST) should be ruled out in a child complaining neurological symptoms, even when features of BD may be mild and incomplete (3, 6, 7). Although not totally clear, the aetiopathogenesis of thrombosis seems to be the consequence of a multifactorial process involving the inflammatory status, the hypercoagulability and the endothelial damage and dysfunction. As evaluated in several studies the most common initial manifestation are the mucocutaneous symptoms, while CVST occurs within 5 years from disease onset (8). Of note, in our patient CVST was one of the first and earliest signs and symptoms of BD, thus heralding the disease.

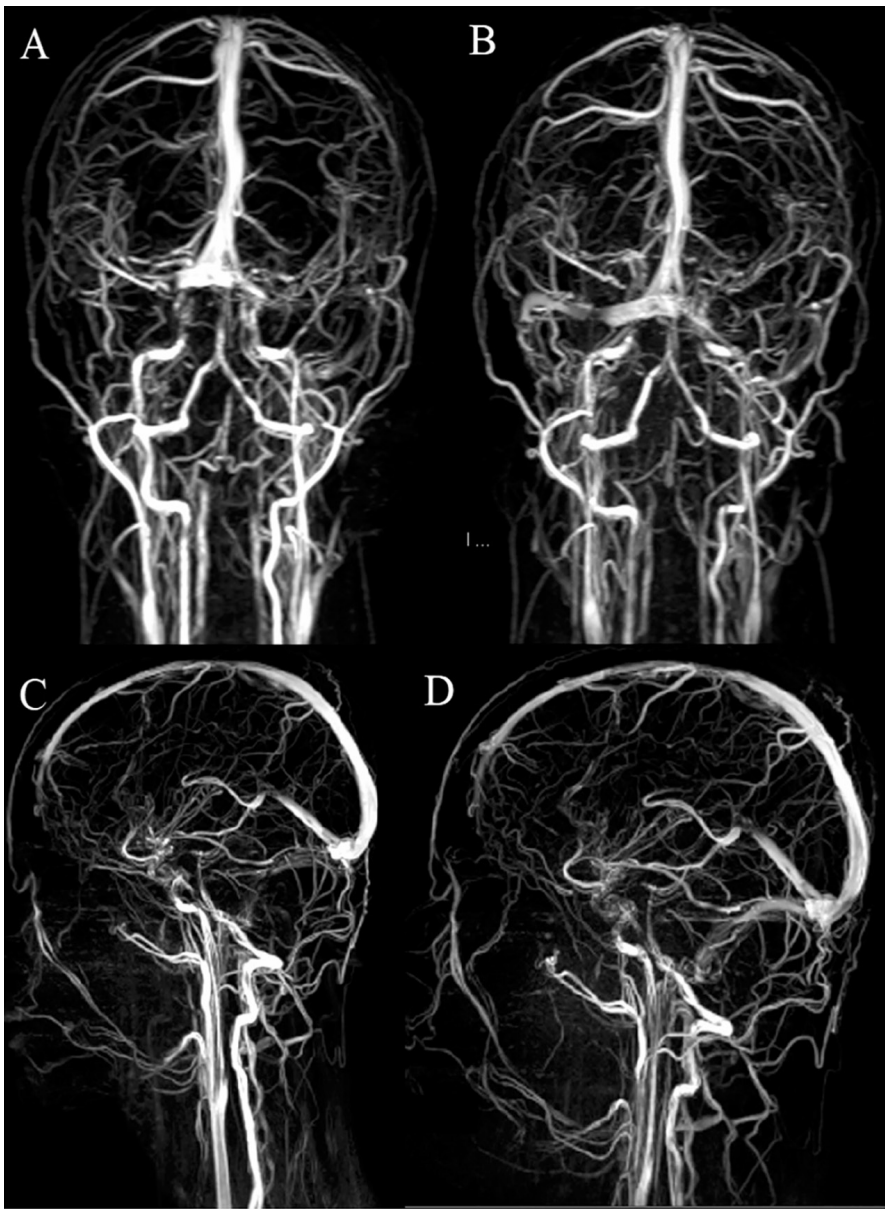

Fig. 1. Brain MRI with contrast and angio-MRI showed an extensive thrombosis of the internal jugular vein, the sigmoid sinus and transverse sinus up to the proximity of the straight sinus of the right side (**A** and **C**).
At 3 month-follow-up a total recovery of the extensive thrombosis of the internal jugular and sinus (**B** and **D**).

As evaluated by Rottenstreich *et al*. in a literature review, CSVT was more frequently in boys, the median age at onset was 12 years and was diagnosed simultaneously with BD in 75% of cases as in our patient (9). Moreover, as evaluated in several studies, the most common sites of CSVT involved were the superior sagittal, lateral and transverse sinus, even though the sigmoid and the internal jugular were also reported (5, 6, 9). The signs and symptoms that typically underlying the CNS involvement were more frequently the headache and the papilloedema at the ophthalmological evaluation (5, 6, 9). Even though the neurological symptoms may be vague, a complete work-up need to be performed. Nowadays there is not an established therapeutic approach for childhood CVST (38), since no prospective studies are currently available. In order to avoid additional complications and increase survival, anticoagulation therapy should be associated with an early immunosuppressive treatment aiming to damp inflammation and thus the risk of a new thrombosis (6).

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