Pseudotumour cerebri presentation of systemic lupus erythematosus: more than an association

SIR, We describe a 22-yr-old female admitted with a 1-month history of blurred vision. Five weeks prior to presentation, the patient started to experience headaches, diplopia and squint followed a week later by blurred vision. The headaches and diplopia gradually subsided, whereas her blurred vision worsened progressively. Prior to admission the patient was being treated with oral multivitamins, including B1, B6, B12 and folic acid, for anaemia. On review of systems, the patient reported an itchy skin rash on the trunk and lower extremities. On physical examination, the patient had severe haemorrhagic papilloedema, a partial right sixth cranial nerve palsy and mild esotropia (convergent squint) of the right eye. The neurological examination was otherwise within normal limits. General examination revealed a malar rash and plaques of follicular pits with blackheads in both conchae in addition to multiple erythematous annular purpuric skin lesions over the trunk and lower extremities. Laboratory work-up revealed normocytic normochromic anaemia with a haematocrit of 29%, a mean cell volume of 84.4 μm³ and a ferritin of 73 μg/l (normal: 6–81). White blood cells (WBCs) were mildly reduced at 3200 cells/μm³ with 960 lymphocytes/μm³. Erythrocyte sedimentation rate was markedly elevated at 120 mm/h. Magnetic resonance imaging of the brain revealed an empty sella with abundance of cerebrospinal fluid (CSF) and mild brain atrophy. Magnetic resonance venography (MRV) was normal with no evidence of cerebral venous...
thrombosis. A lumbar puncture revealed an opening pressure of >55 cmH₂O, 3 mononuclear cells/mm³, 28 red blood cells (RBCs)/mm³, protein of 0.22 g/l and glucose of 3.9 mmol/l with serum glucose of 5.7 mmol/l. The CSF IgG ratio (CSF IgG/CSF albumin) was elevated at 40% with normal IgG index (CSF IgG × serum albumin/CSF albumin × serum IgG). Oligoclonal bands were not detected.

Further work-up revealed highly positive antinuclear antibodies (ANA), anti-double-stranded DNA (ds-DNA), anti-SM, anti-SS-A, anti-SS-B, anti-SM/RNP, anti-Scl-70 and anti-Jo antibodies. Rapid plasma reagin was reactive. IgG antinuclear antibody was positive at 23 U/ml and IgM antinuclear antibody was negative. Lupus anticoagulants were negative. Urinalysis showed 20 WBCs, 30 RBCs and 1+ protein with no casts.

Our patient fulfils the diagnostic criteria for pseudotumour cerebri with (i) an elevated CSF opening pressure of more than 25 cmH₂O, (ii) normal CSF sugar, total protein and cells, (iii) no radiological evidence of an underlying pathology and (iv) all signs and symptoms being related to increased intracranial pressure (ICP). An empty sella on neuroimaging and a sixth nerve palsy on physical examination are both due to increased ICP. She fulfils the criteria of systemic lupus erythematosus (SLE) as well. She has (i) a malar rash, (ii) discoid lupus (in the concha), (iii) anaemia and leucocytopenia, (iv) proteinuria, (v) positive antinuclear antigen antibodies (ANA) and (vi) positive anti-ds-DNA along with many other ANA. The skin and haematopoietic systems were mainly targeted, whereas the renal function was relatively preserved, with normal creatinine and urea at 48 and 4.1 mmol/l respectively, in spite of a quite severe autoantibody flare-up. It is uncertain as to how long the patient had experienced the signs and symptoms of SLE. Discoid lupus affecting the concha may precede the diagnosis of systemic lupus by many years in a completely asymptomatic patient. However, it is more indicative of concurrent systemic disease [1]. Plaques of black-headed follicular pits in the concha are quite typical and highly specific for lupus [2].

Although she displayed findings upon presentation sufficient to make the diagnosis, or at least raise the suspicion, of SLE, the patient interestingly presented with a full-blown picture of pseudotumour cerebri. To the best of our knowledge, there has been only one case of SLE presenting initially with pseudotumour cerebri described in the literature [3]. In our patient, the elevated ICP was not due to cerebral venous thrombosis given the normal MRV. Cerebral venous thrombosis, presumably due to a hypercoagulable state, has been described in patients with SLE [4]. Severe anaemia (not as seen in our patient, who had only a mild reduction in haematocrit) has been associated with increased ICP and papilloedema [5]. Brain atrophy has been reported in some children with SLE and central nervous system involvement [6]. Although the significance of brain atrophy in our patient is uncertain, it may point to a more chronic process. The markedly elevated CSF IgG ratio, which to our knowledge has not previously been reported in pseudotumour cerebri, correlates with the diffuse autoantibody flare-up seen in the blood rather than being a reflection of intrathecal IgG synthesis. This high CSF IgG ratio, to a certain extent, questions the integrity of the blood–brain barrier [7]. The known antibodies associated with lupus cerebritis, such as anti-ribosomal P and anti-neuronal antibodies, were all negative. The significant antinuclear antibody response that was noted in this patient, which is consistent with an SLE flare-up, drew our attention to the possible trigger for the development of pseudotumour cerebri. The association of pseudotumour cerebri with SLE disease flare-up has been reported in several other cases as well [8]. Our patient, similar to other patients with SLE-associated pseudotumour cerebri [9], responded dramatically to steroids, which is quite atypical for ‘idiopathic’ pseudotumour cerebri. The high CSF IgG ratio and the dramatic response to steroids may both point towards an underlying process. From a clinical point of view, we argue that pseudotumour cerebri in this patient does not represent an idiopathic process, although the pathophysiology remains unknown. Rather than a simple association, we suggest that in this case pseudotumour cerebri developed secondary to SLE flare-up. We further suggest extending the work-up of pseudotumour cerebri to include screening for connective tissue diseases and IgG ratio in the CSF.

S. SBEITI, D. M. KAYED, H. MAJURI
Division of Neurology, Department of Internal Medicine, Tawam Hospital, Ministry of Health, Al Ain, United Arab Emirates
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Correspondence to: S. Sbeiti, PO Box 15258, Tawam Hospital, Al Ain, Abu Dhabi, United Arab Emirates.
E-mail: saleshsbeiti@hotmail.com