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HYPERTROPHIC PACHYMENINGITIS AS AN UNUSUAL CAUSE OF HEADACHE AND SPHINCTER DYSFUNCTION IN SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT

Hypertrophic pachymeningitis (HP) is an uncommon condition characterised by focal or diffuse thickening of the dura mater. An increasing number of cases have been reported of its association with underlying connective tissue diseases. It is a rare complication in systemic lupus erythematosus (SLE) and might be the initial and sole clinical manifestation. We report a case of a 21-year-old man presenting with febrile meningeal syndrome and sphincter dysfunction. Physical examination showed malar rash and joint pain. Biological assessment revealed a regenerative normocytic normochromic anaemia, a leucopenia and a lymphopenia. The 24-hour urine protein was positive at 0.6 g. Immunological evaluation revealed positive antinuclear, anti-Sm and anti-dsDNA antibodies. Brain and spinal magnetic resonance imaging showed hypertrophic pachymeningitis. Cerebrospinal fluid biochemistry was within normal limits. Renal biopsy revealed a mesangial proliferative lupus nephritis. The diagnosis of SLE with neurologic and renal involvement was established, and the patient was treated with intravenous methylprednisolone pulse, followed by oral prednisone in association with azathioprine and hydroxychloroquine. Considering the persistence of symptoms and MRI lesions after 6 months, a treatment with rituximab was initiated with good evolution.

KEYWORDS

Lupus, headache, urinary retention, hypertrophic pachymeningitis

LEARNING POINTS

- Hypertrophic pachymeningitis is a rare condition of diverse aetiologies.
- A workup including search for infectious, autoimmune and neoplastic aetiologies should be performed.
- It is an extremely rare complication in systemic lupus erythematosus and might be the initial and sole clinical manifestation.

INTRODUCTION

Hypertrophic pachymeningitis (HP), a focal or diffuse thickening of the dura mater, is an unusual condition. It can be idiopathic or due to some disorders such as infections, tumoral and autoimmune diseases; the histology demonstrates non-specific chronic inflammation[1]. The

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presence of HP underlying systemic lupus is extremely rare. We report a case of systemic lupus erythematosus (SLE) revealed by febrile meningeal syndrome and sphincter disorders, due to HP.

CASE PRESENTATION

A 21-year-old man, with a history of treated lymph node tuberculosis four years prior to his admission, presented with a severe headache, photophobia and fever. He also had a history of urinary retention and constipation.

No signs of meningeal irritation nor neurological deficits were observed at the physical examination. Initial brain and spinal MRI were unremarkable. Fundus examination was normal. Lumbar puncture revealed a normal pressure. Cerebrospinal fluid (CSF) biochemistry was within normal limits: white cell count at 3/mm^3, protein at 0.4 g/l, glucose at 0.4 g/l. The CSF polymerase chain reaction, bacterial and tuberculous culture were negative.

A diagnosis of meningitidis-retention syndrome, a peculiar combination of acute urinary retention and aseptic meningitis, was suspected. The patient was lost to follow-up and came back after 3 years with the same symptomatology. Physical examination showed an apyretic patient (37 °C) with malar rash and joint pain (knees and ankles). Laboratory tests showed a regenerative normocytic normochromic anaemia at 9 g/dl, leucopenia at 1500/mm^3 and lymphopenia at 480/mm^3. The direct Coombs test was positive. The C-reactive protein was elevated at 50 mg/l and the erythrocyte sedimentation rate was at 85 mm. Tests of renal and liver functions were normal. The 24-hour urine protein was positive at 0.6 g.

The immunological evaluation revealed positive antinuclear antibodies by indirect immunofluorescence method with a speckled pattern at a titre of 1/1280, positive anti-Sm and anti-double-stranded DNA antibodies. Anticardiolipin, anti-β2 glycoprotein I antibodies, rheumatoid factor and ANCA testing were negative. The calcium, angiotensin-converting enzyme and IgG4 levels were normal.

Retrograde urethrocystography showed a weak urine stream and urinary hesitancy. Urodynamic tests revealed detrusor hypocontractility.

Brain magnetic resonance imaging (MRI) showed a generalised thickening of the dura matter with a hyperintense signal on fluid attenuated inversion recovery (FLAIR) sequence and gadolinium contrast enhancement *(Fig. 1)*. MR venography showed no venous sinus thrombosis. Renal biopsy revealed a mesangial proliferative lupus nephritis.

The diagnosis of SLE with neurologic and renal involvement was established, and the patient was treated with intravenous methylprednisolone pulse, followed by oral prednisone 60 mg daily (1 mg/kg/day) in association with azathioprine 150 mg/day and hydroxychloroquine 400 mg daily. The headache gradually resolved by the third day.

After 6 months, the headache was still persistent and the MRI remained unchanged, while the 24-hour urine protein was negative. Thus, a treatment with rituximab 1 g was initiated with two infusions, 2 weeks apart.

The follow-up MRI showed regression of lesions *(Fig. 2)*, and the patient was asymptomatic six months later.

DISCUSSION

HP is an extremely rare entity which can be associated with varied conditions. Clinical presentation depends on the site of involvement; the most common sign is headaches. Other clinical presentations include cranial nerve palsies, cerebellar ataxia, recurrent visual loss, venous and arterial occlusion, obstructive hydrocephalus and hypopituitarism complicated by diabetes insipidus[2].

The pathogenesis of HP remains largely unknown. It can be related to an underlying autoimmune condition resulting in inflammation and fibrosis of the dura mater. Many case reports suggested an association between HP and autoimmune disorders such as ANCA-positive vasculitis, connective tissue diseases and antiphospholipid syndrome.

Lumbar puncture may show lymphocytic pleocytosis but may be normal in up to one-quarter of patients, as was the case with our patient[3]. MRI imaging shows enhancing pachymeningeal thickening, as seen in our case. Dural biopsy confirms the diagnosis by showing severe interstitial fibrosis, diffuse inflammatory cell infiltration and non-specific inflammatory cells[4].

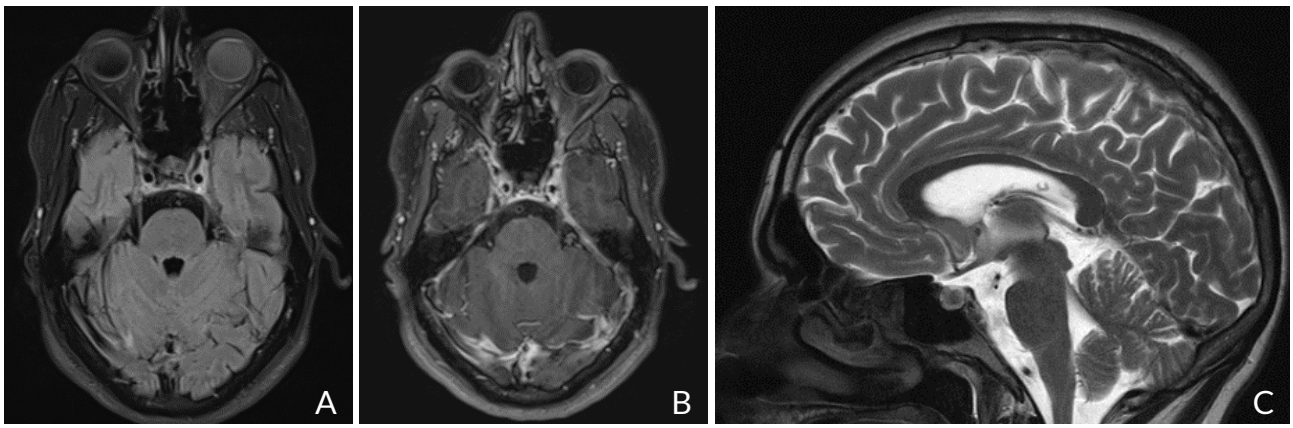

Figure 1. Brain magnetic resonance imaging showing pachymeningeal thickening over temporal lobes and the vertex with diffuse enhancement after injection. A) Axial T2 FLAIR sequence, B) sagital T2-weighted sequence, C) axial T1-weighted sequence after gadolinium injection

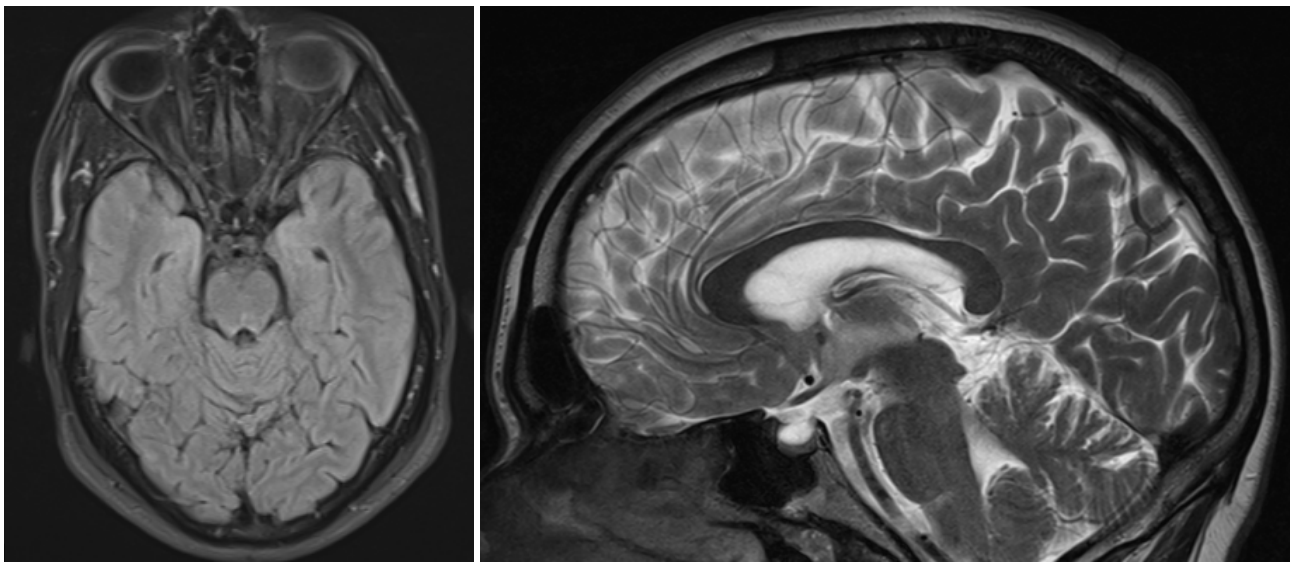

Figure 2. Follow-up brain magnetic resonance imaging, 6 months after rituximab therapy, showing total resolution of the pachymeningeal thickening

The diagnosis of HP can be difficult, especially if it is the sole manifestation. Thus, dural biopsy is necessary to exclude other causes and to establish the diagnosis of idiopathic HP.

The association between SLE and HP is extremely rare. To our knowledge, there are only ten case reports suggesting this association.

In our case, HP was the first manifestation of the SLE. The patient presented with a severe headache and sphincter dysfunction. Based on the negative bacteriological examinations and rapid clinical improvement with immunosuppressive drugs, we considered that HP is caused by SLE itself.

There is currently no consensus on the treatment of HP associated with SLE, because of the paucity of cases. As noted in all the previous case reports, the use of steroids showed good results. Jang et al.[5] suggest that rituximab is a good treatment for steroid-refractory HP, even with idiopathic cases.

CONCLUSION

HP is an extremely rare complication in SLE and might be the initial and sole clinical manifestation. Due to its rarity, additional studies are required to establish a guideline treatment. Steroids are the cornerstone of treatment for HP in SLE, with positive results. Immunosuppressive agents can be added depending on the initial response. A high index of suspicion for HP should be maintained in patients with SLE presenting with neurological disorders, to prevent further neurological sequelae.

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