CASE REPORT

Spontaneous intracranial hypotension associated with pachymeningeal enhancement in a patient with systemic lupus erythematosus (SLE): an extremely rare presenting feature

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SUMMARY

A 16-years-old girl presented to our institution with history of severe bilateral headache for 5 days associated with vomiting. She also had fever for 2 months without any localising symptoms and skin lesions for 1 month. Examination revealed erythematous rash over bridge of nose and ear lobes, ulcer over hard palate and tenderness of small joints of both hands. Systemic examination was unremarkable except for bilateral papilloedema. Investigations revealed anaemia, leucopenia and elevated erythrocyte sedimentation rate. Cranial imaging revealed diffuse pachymeningeal enhancement with subdural effusion. Lumbar puncture revealed no abnormal findings in cerebrospinal fluid except low opening pressure. Antinuclear antibody was 4+ with anti-Sm antibody positive. She was diagnosed to have spontaneous intracranial hypotension associated with pachymeningeal enhancement secondary to systemic lupus erythematosus. She showed dramatic improvement with steroid and azathioprine therapy. She continues to be asymptomatic after 2 years of follow-up.

BACKGROUND

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with a myriad of clinical manifestations and atypical presentations which makes the diagnosis challenging. This is further complicated by low index of suspicion and possibilities of various infectious aetiologies especially in tropical countries. A high index of clinical suspicion and early recognition alters the natural course of disease and improves response without significant sequelae. We came across a 16-years-old girl who presented to our institution with features suggestive of raised intracranial pressure. On evaluation, she was found to be having spontaneous intracranial hypotension associated with pachymeningeal enhancement as a presenting manifestation of SLE which is very rare. This case is reported not only because of its rarity but because of diagnostic as well as management dilemma we faced while treating her.

CASE PRESENTATION

A 16-years-old girl who was previously healthy presented with history of low grade fever for 2 months

without any localising symptoms and skin lesions for 1 month over root of the nose which started as a small vesicle and progressed to erythematous lesion, followed by similar lesions over periorbital areas, upper arm and trunk not associated with photosensitivity or itching. Five days prior to hospitalisation, she developed severe bilateral predominantly retro-orbital headache associated with vomiting. There was no fever or visual blurring associated with this headache. She also complained of pain and early morning stiffness involving the small joints of hand which improved with analgesics. Her medical and family histories were unremarkable.

On examination, she had mild pallor, erythematous rash over bridge of nose and ear lobes with a few purpuric lesions over the volar aspect of the forearm and deltoid region consistent with acute cutaneous lupus. Metacarpophalangeal and proximal interphalangeal joints of both hands were tender for palpation with no associated swelling. There was a 1 × 1 cm ulcer over the hard palate. Central nervous system (CNS) examination revealed no meningeal signs or focal neurological deficits. Fundus examination revealed bilateral papilloedema. Rest of the examination was unremarkable.

INVESTIGATIONS

She had normocytic normochromic anaemia (haemoglobin 8.6 g/dL), leucopenia (leucocyte count $2.7 \times 10^9/L$), elevated erythrocyte sedimentation rate (ESR; 55 mm at 1 hour). Kidney function and liver function tests were normal and urine examination did not reveal any abnormality. CT of the brain revealed diffuse cerebral oedema and MRI showed rim of subdural effusion with no mass effect over the right frontoparietal region and diffuse pachymeningeal enhancement over bilateral cerebral convexities and in the tentorium cerebelli (figures 1 and 2). The cerebrospinal fluid (CSF) spaces in bilateral periorbital areas were prominent as well. There were no parenchymal abnormalities. Cerebrospinal fluid analysis did not reveal any abnormalities except low opening pressure (6 cm H₂O). Antinuclear antibody (ANA) was 4+ and ANA blot for extractable antigens demonstrated positive anti Sm/RNP, anti Sm and anti PCNA antibodies. Antineutrophil cytoplasmic

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Figure 1 MRI (Coronal reconstruction of post-contrast 3D T1 gradient echo weighted sequence) reveals subdural effusion [*] overlying the right parietal convexity and mild diffuse uniform pachymeningeal enhancement (arrow) (left) which resolved completely with treatment (right).

antibody was negative. Bone marrow examination revealed normocellular active marrow without any abnormalities.

DIFFERENTIAL DIAGNOSIS

1. Tuberculosis.
2. Sarcoidosis.
3. Vasculitis.
4. Leukaemia.

TREATMENT

Patient was diagnosed to have spontaneous intracranial hypotension associated with pachymeningeal enhancement secondary to SLE (based acute cutaneous lupus, oral ulcer, arthritis, leucopenia, ANA and anti-Sm antibodies). She would also have fulfilled Systemic Lupus International Collaborating Clinics (SLICC) classification criteria for SLE.¹ After ruling out infectious causes, patient was given intravenous methylprednisolone pulse 1gm/day for 3 days followed by oral prednisolone 1mg/kg/day for 4 weeks after which the dose was gradually tapered. Azathioprine was added as a steroid sparing agent after ruling out thiopurine methyltransferase (TPMT) polymorphism.

OUTCOME AND FOLLOW-UP

Headache dramatically improved after methylprednisolone pulse. Fever and skin lesions also subsided subsequently and joint pain resolved. Repeat MRI of brain was done after 5 months of treatment which revealed complete resolution of subdural effusion and pachymeningeal enhancement (figures 1 and 2). After 2 years of regular follow-up patient has remained asymptomatic.

DISCUSSION

Spontaneous intracranial hypotension also known as cerebrospinal fluid hypovolaemia is a rare entity characterised by

Figure 2 MRI (Coronal T2 weighted TSE sequence) shows subdural effusion [*] overlying the right parietal convexity (left) which completely resolved with treatment (right).

orthostatic hypotension relieved by lying down. Spontaneous spinal cerebrospinal fluid leak due to simple dural rents or fragile arachnoid cysts often in the presence of underlying but previously unrecognised connective tissue disorder is the typical cause of spontaneous intracranial hypotension.²

Spontaneous intracranial hypotension associated with pachymeningeal enhancement is an extremely rare manifestation in patients with SLE. There are only two such cases described in the literature.^{3 4} One case which is similar to ours had generalised thickening of the dura mater and presence of subdural effusion suggestive of spontaneous intracranial hypotension. Meningeal biopsy was deferred and patient was treated with oral steroids, hydroxychloroquine and mycophenolate mofetil. Since dura mater thickening persisted with resolution of clinical and other radiological features authors felt that both hypertrophic pachymeningitis and CSF hypovolaemia coexisted in the patient. They postulated that persistent meningeal inflammation lead to meningeal disruption causing CSF leak and CSF hypovolaemia. In another case of spontaneous intracranial hypotension associated with SLE, CSF leak was demonstrated in the cervicothoracic junction bilaterally by integrated single-photon emission computed tomography (SPECT/CT).

Both the previous cases were more than 60 years old whereas our case is 16 years old girl. Spontaneous intracranial hypotension was the presenting manifestation in the first patient similar to our patient. First patient presented with episodes of fall and unconsciousness whereas in our patient the presenting symptom was headache. Proliferative and membranous nephropathy (class IV/V) was seen in first patient but our patient did not have other serious organ involvement. There was a very good response to immunosuppressive therapy in the first patient as seen in ours. Details of the second case are unavailable as it is published as an image.

To summarise, spontaneous intracranial hypotension associated with pachymeningeal enhancement is a very rare presenting manifestation of SLE. There are only two previous such cases reported in the literature. This diagnosis should be considered in patients with SLE presenting with headache. MRI is the investigation of choice. Steroids with or without immunosuppressive drugs are first line therapy for this condition. It has good prognosis with therapy.

Learning points

- ▶ Spontaneous intracranial hypotension associated with pachymeningeal enhancement is an extremely rare presenting manifestation of systemic lupus erythematosus.
- ▶ Infectious causes of pachymeningeal enhancement needs to be ruled out before immunosuppressive therapy.
- ▶ Spontaneous intracranial hypotension associated with pachymeningeal enhancement due to systemic lupus erythematosus responds dramatically to immunosuppressive therapy.

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