Ferrrol-Besnier disease with associated recurrent meningitis

We report an unusual case of a recurrent cutaneous syndrome regularly associated with a lymphomocytary meningitis.

A 40 year old white man presented with a complaint of throat pain and a burning sensation on the inside of the hands and feet starting three days previously, followed a day later by fever and severe headache. Neurological examination was unremarkable except for meningism. On the inside of both hands and feet extensive reddening and early desquamation were noted. Examination of CSF showed normal protein, lactate, and sugar content, no local IgG synthesis, and 216 μl, 80% of which were lymphocytes and monocytes with a high proportion of large, fragile endothelial cells; 20% were polymuclear cells and about half of these were eosinophils. No infectious agent, including hepatitis B and herpes simplex viruses, was demonstrated serologically, in culture or by virus isolation. Antibody studies for connective tissue disease, T cell differential count, streptolysin titres, complement levels, and electrophoresis for immunoglobulin were normal.

Fever and headache subsided within four days. Two days later, extensive desquamation with almost complete shedding of palmar and plantar skin occurred (figure). The patient had had six similar attacks since 1986 with intervals from seven to 18 months. All episodes started with throat pain and a burning sensation on palmar and plantar skin, followed by fever and headache that subsided within days, accompanied by extensive desquamation of affected skin, on two occasions also of several nails. Neurological abnormalities other than meningism were not found. Brain CT and MRI were normal on the several occasions that they were done. No leak was demonstrated on CSF scintigraphy. During five episodes a lymphocytic CSF pleocytosis was documented. No infectious agent was isolated. On two occasions a slight transient proteinuria was noted. The patient was well between attacks, had no skin abnormalities in the intervals, and did not require regular medication.

The cutaneous syndrome conforms to the rare clinical entity of erythema scarlatiniforme desquamativum recidivans of Ferrrol-Besnier. It is characterised by recurrent attacks of a prodromal phase with head and muscle aches, gastrointestinal and enteric syndromes, and fever, followed by a macular erythema leading to the pathognomonic desquamation and scaling of palmar and plantar skin. Patients are symptom free during the intervals, which may last from weeks to several years. About 40 definite cases have been reported since the disease was described in 1878. Localised variants in which only the hands and feet are involved correspond precisely to the cutaneous syndrome found here. Throat pain and transient proteinuria, unusual for recurrent meningitis, are common. The aetiology is unknown, but abnormal cutaneous reaction to an infectious disease has been proposed as the cause in some cases. No infectious agent was isolated. Connective tissue diseases, immunosuppression, and uveomeninginitis syndromes were excluded by clinical presentation and appropriate laboratory studies.

Interestingly, the CSF cytology, with a mixed lymphomocytary pleocytosis with large fragile endothelial cells, conforms to the picture seen in benign recurrent aseptic meningitis (Mollaret's meningitis). An unusual feature was the high number of eosinophils, rarely reported in this disease. The clear association in this case of Ferrrol-Besnier disease with recurrent CSF lymphocytosis has no precedent in the literature. However, we located two patients presenting with an urticarial rash during episodes of recurrent meningitis. In these cases possible aetiologies were lymphoma and familial Mediterranean fever. These diagnoses should be borne in mind when confronting a patient with the rare picture of recurrent meningitis associated with cutaneous symptoms.

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Trigeminal neuralgia in pontine ischaemia

Trigeminal neuralgia occurs in several conditions involving slight damage of the trigeminal root entry zone into the pons. To our knowledge there are no reported cases of trigeminal neuralgia occurring after brainstem ischaemia. We report one such patient.

A 58 year old man had had trigeminal neuralgia in the territory of the second branch of the right trigeminal nerve for four years. Carbamazepine (200 mg twice daily) had been effective for the first two to three years of his pain, but was useless when we first saw him. Neurological examination showed slightly diminished superficial sensation in the territory of the second and third branch of the right trigeminal nerve, and was otherwise normal. Corneal reflex was normal bilaterally. The sensory loss was confirmed by quantitative sensory testing. Trigeminal evoked potentials (TEPs) were obtained after stimulation of the infraorbital nerve. On the left side they were normal, but delayed on the right side. A T2 weighted axial MRI. Patient's right side is on the figure. The left hand Myelin spot-like area (arrow) is present in the right lateral part of the pons, corresponding to the trigeminal root entry zone. This area was not seen in corresponding T1 weighted images, but was seen in proton-density images (not shown). These findings are indicative of a small ischaemic lacune.
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