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Ferréol-Besnier disease with associated recurrent meningitis

We report an unusual case of a recurrent cutaneous syndrome regularly associated with a lymphomonocytary 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 μ l, 80% of which were lymphocytes and monocytes with a high proportion of large, fragile endothelial cells; 20% were polynuclear 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 Ferréol-Besnier. It is characterised by recurrent attacks of a prodromal phase with head and muscle aches, gastrointestinal and enteritic syndromes, and fever, followed by a macular erythema leading to the pathognomonic desquamation and scalding of palmar and plantar skin.¹ 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.¹ 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.¹ No infectious agent was isolated. Connective tissue diseases, immunosuppression, and uveomeningitic syndromes were excluded by clinical presentation and appropriate laboratory studies.

Interestingly, the CSF cytology, with a mixed lymphomonocytary pleocytosis with large fragile endothelial cells, conforms to the picture seen in benign recurrent aseptic meningitis (Mollaret's meningitis).² An unusual feature was the high number of eosinophils, rarely reported in this disease.³

The clear association in this case of Ferréol-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 lymphoma⁴ and familial Mediterranean fever.⁵ 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.¹ 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.² Trigeminal evoked potentials (TEPs)³ were obtained after stimulation of the infraorbital nerve. On the left side they were normal,

T2 weighted axial MRI. Patient's right side is on the figure's left hand side. A hyperintense 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.

Patient's right foot on sixth day after admission, showing extensive desquamation.

whereas on the right side all components beyond W2 were delayed and reduced in amplitude, suggesting an impairment of afferent conduction at some site between the trigeminal root and the trigeminal nuclei.⁴ Increasing carbamazepine dosage to 400 mg three times daily caused the pain to disappear. Two years later his symptoms worsened. Neurological examination and TEPs were unchanged. Magnetic resonance angiography showed multiple ischaemic lesions in the cerebral hemispheres with widespread cortical atrophy. An ischaemic lesion was found in the right lateral part of the pons, in the trigeminal root entry zone (figure). Multiple sclerosis and Lyme disease were ruled out by the clinical history and by appropriate investigations. Radiofrequency selective thermal rhizotomy was followed by a slight, further decrease of tactile and pain sensation in the right trigeminal territory (second branch) and by disappearance of the pain.

In this patient the typical pain of trigeminal neuralgia was associated with an ischaemic lesion strictly localised to the ipsilateral trigeminal root entry zone. This association may have been, in theory, coincidental, but it seems unlikely. In fact, alterations in TEP showed a functional damage of the afferent pathway at the same site where altered morphology was detected by MRI. Furthermore, the ischaemic lesion was exactly at the trigeminal root entry zone, an area where most lesions causing secondary trigeminal neuralgia are located.¹ Neuralgic pain waxed and waned for many years: this is expected, as trigeminal neuralgia is known to show remissions and recurrences secondary to permanent trigeminal lesions.¹ We recommend that brainstem ischaemia is included in the differential diagnosis of trigeminal neuralgia, especially when the neurological examination discloses alterations in trigeminal nerve function. Measurement of TEP is useful in the search for such alterations. In our patient the pain was successfully treated with carbamazepine for several years, and was eventually relieved by radiofrequency selective thermal rhizotomy, a procedure that may not necessarily be confined to the treatment of "essential" trigeminal neuralgia.

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MRI demonstration of reversible impairment of the blood-CNS barrier function in subacute combined degeneration of the spinal cord

We report clinical, laboratory, and imaging findings in a case of subacute combined degeneration of the spinal cord. The 50 year

(A) Sagittal T1 weighted image after administration of 15 ml contrast agent. Apart from larger lesions at the thoracic and lower cervical level (large arrows), many smaller pearl-like contrast enhancing spots can be seen (small arrows).

(B) Sagittal T1 weighted image after administration of 15 ml of contrast agent. A nodular contrast enhancing lesion is present in the posterior cervical cord (arrow).

old woman presented with tickling sensations running down her back when bending the head. Five months before she had first noticed numbness of her feet which slowly ascended to the level of her nipples. During the past six months she had lost 7 kg in weight.

Forward flexion of her neck induced Lhermitte's phenomenon. The tendon reflexes of her legs and plantar responses were absent. Complete loss of light touches, vibration, and position sense was found below D5. Pain and thermal perception were not diminished. Her gait was unsteady due to a sensory ataxia.

Her mean red cell volume was 110 fl and serum vitamin B12 concentration was 44 pmol/l. Haemoglobin, packed cell volume, and folate were normal. The two stage "Schilling test" showed intestinal malabsorption of vitamin B12, not due to lack of intrinsic factor. Gastric endoscopy was non-specific. analysis of CSF was normal.

Somatosensory evoked potentials from tibial and sural nerves showed abnormalities in latency indicative of a lesion in the posterior columns of the spinal cord; the somatosensory evoked potentials from both median nerves were normal. Nerve conduction studies of the sural nerves disclosed a reduced nerve conduction velocity indicative of a demyelinating neuropathy.

Two weeks after treatment with 1000 µg vitamin B12 daily, there was an almost complete restitution of sensory functions.

T2 weighted MRI images of the thoracic spinal cord showed an ill defined hyperintense lesion in the posterior parts of the spinal cord. T1 weighted images after administration of gadolinium DTPA showed multiple slightly expansive, contrast enhancing lesions in the posterior column of the cervical and thoracic spinal cord (fig A, B). After 18 days of treatment the lesions had disappeared.

In summary, our patient had the classic clinical signs of spinal cord degeneration including Lhermitte's phenomenon, sensory impairment, dysaesthesia, and sensory ataxia of the lower limbs. In all cases reported so far, MRI studies were performed in spinal cord degeneration to exclude a spinal cord compression.¹⁻⁵ The lesions detected as hyperintensities in T2 weighted images were always located in the posterior columns of the spinal cord. The thoracic region was affected in all patients. Signal abnormalities in the cervical region were seen in two cases.^{4,5}

We found a very pronounced, multifocal contrast enhancement of the cervical and thoracic sections of the spinal cord indicating blood-CNS barrier disruption. The lesions were multifocal and located close to the spinal cord surface. A slight degree of expansion was noted. Thus a granulomatous inflammation, multiple sclerosis lesions, or tumour metastases had to be considered. The clue to spinal cord degeneration in this case was the location of the lesions exclu-