third and nineteenth day from the beginning of the symptoms, the first one being normal, and the second one showing an increase in protein (0.95 g/l) with 11 cells and negative titres to brucella, syphilis, toxoplasma, herpes simplex, varicella-zoster, and a negative antinuclear antibody determination.

The patient's clinical pattern began to improve two weeks after admission; she was discharged a week later. Follow up a month later revealed only a generalised areflexia and a slight dysmetria on the heel-toe test.

Electrophysiological studies (table) were carried out on the sixth and eighteenth day, which revealed absent sensory potentials. Motor NCS showed less abnormality than that of sensory nerves. There was delay in the distal motor latency in all the nerves tested and there was also reduction in motor amplitude in both median nerves, with a 25-3% drop in peak-to-peak amplitude between wrist and elbow, and a 39-2% drop in amplitude between wrist and axilla in the first right median nerve conduction study. Neither temporal dispersion nor motor conduction velocities of the nerves were within normal limits. EMG studies were normal. A third electrophysiological study performed a month after her discharge showed the recovery of the sensory nerve action potentials in the median and upper extremities, although the amplitude and sensory conduction velocity of the nerves studied were lower than the normal values. The motor NCS, however, were similar to previous results. The EMG study of the right anterior tibial and right first dorsal interosseous muscles showed compatible changes with minimum denervation.

Our patient had Guillain-Barré syndrome as an entity whose nosological characterisation relied upon a purely descriptive base, with relatively widely accepted diagnostic criteria. Among these, the presence of muscular weakness is the most noticeable, indicating the main affection of the motor roots. However, a purely sensory clinical variant of this illness is also believed to be possible and the following features are necessary for it to be accepted: rapid onset, distribution widespread and symmetrical, complete or near recovery, high CSF protein content, with few or no cells, and an electrophysiological study compatible with a demyelinating process in the peripheral nervous system.

There is still a certain controversy as to whether this clinical variant occurs. In a recent review of 42 patients with acute sensory neuropathy, the authors concluded that this condition is not part of the spectrum of inflammatory demyelinating neuropathies,2 nevertheless, it is noticeable that only 2 of the 42 patients had complete remission of symptoms and that the course of their disease was very protracted—from six to nine months.

Cerebral vascular thrombosis in paroxysmal nocturnal haemoglobinuria

Paroxysmal nocturnal haemoglobinuria (PNH) is a rare acquired disease of the haematopoietic system manifested by chronic haemolytic anaemia, leukopenia, and thrombocytopenia. Patients with PNH have an increased risk of developing systemic venous thrombosis, and obstruction of the hepatic veins, or Budd-Chiari syndrome (BCS) which is a highly fatal complication in about 30% of cases. PNH is also a well-established, though extremely uncommon cause of cerebral venous thrombosis (CVT).1,4 We describe a patient with PNH and BCS in whom CVT was angiographically documented. We comment upon the risks of anticoagulants in this unusual situation and the need to monitor PNH among the possible causes of CVT.

A 34 year old woman was admitted to hospital because of malaise, low-grade fever, pleural effusion, hepatomegaly and ascites. The diagnosis of PNH was confirmed by the presence of haemosiderinuria, positive Ham acid haemolysis and sucrose lysis tests. Hepatic venogram confirmed the existence of BCS. She received repeated blood transfusions and a year later a portacava shunt was performed. Three years later she complained of intensive fronto-occipital pain and clumsiness of the left limbs. Examination showed a mild distal paresis and a left leg with only minimal slowness of alternating movements in the left arm. There was impairment of position and pain perception with sensory extinction on the left side. The optic discs were slightly blurred. The visual fields were full with a moderately enlarged blind spot on both sides. CT scanning showed a right parietal infarction. Cerebral angiography revealed thrombosis of superior sagittal sinus, straight sinus, lateral sinuses, and internal cerebral veins. The patient made a complete recovery with dexamethasone and coumarin therapy.

In a comprehensive review of 38 cases, Bousset et al4 failed to list PNH among the possible causes of CVT. PNH is commonly undiagnosed for a period of months or years, and its true incidence among patients with CVT is probably underestimated because detailed coagulation studies are not performed in most cases. This may be of utmost importance with cerebral venous thrombosis, as there is no better treatment than heparin under consideration. The recent German randomised trial demonstrated the benefit of high-dose heparin in patients with CVT.1,2 Conversely, therapy with heparin has caused exacerbation of haemorrhage in some patients with PNH,1,2 and thus might be deleterious in this particularly difficult situation.

PNH should be considered among other possible causes of CVT and ruled out by the appropriate laboratory investigations, especially when treatment with heparin is contemplated.
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