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Opercular syndrome due to non-convulsive status epilepticus in an adult

The Foix–Chavany–Marie opercular syndrome (FCMS), a severe form of pseudobulbar palsy due to bilateral anterior opercular lesions, may be congenital or acquired, persistent or intermittent.[1] FCMS due to epilepsy has been described nearly exclusively in childhood.[1] We report the case of an adult patient in whom non-convulsive status epilepticus (NCSE) manifested with opercular syndrome, and which was completely reversible with treatment for epilepsy.

Case report

A 55-year-old patient with chronic renal failure on haemodialysis was admitted to the orthopaedic surgery department for the treatment of a bilateral humerus fracture. Cognitive and mental functions of the patient were normal before admission. She started to receive oxycodone–acetaminophen four times a day and later oxycodone 10 mg twice a day (total dose of oxycodone 60 mg over 48 h) for pain control. Over the course of 3 days she became confused and later obtunded. Oxycodone was discontinued. She became more alert and was able to communicate with gestures. Neurological examination, however, showed anarthria and inability to swallow, chew, or move her lips and tongue on command. Comprehension was retained during the whole episode, a fact that was proved after recovery, as the patient remembered specific details and events that had occurred during the entire incident. No focal signs were observed. Corneal, gag and jaw reflexes were preserved. Reflexive buccofacial movements such as yawning or coughing were present. Limb praxis was normal and eye movements were intact. Deep tendon reflexes were weak and no pyramidal signs were elicited. Routine blood tests disclosed mild normocytic anaemia and chronic renal failure, with no change in her haemodynamic status. Infective and inflammatory screens were negative. Computed tomography of the brain showed moderate to severe brain atrophy and bilateral subcortical lacunar lesions. These findings were similar to those observed 1 year earlier. No evidence of a new subcortical infarction was seen. The electroencephalograph (EEG) showed continuous rhythmic delta activity mixed with sharp waves and long periods of spike and wave ictal discharges (fig 1, left panel), consistent with NCSE. Intravenous valproic acid was initiated. Regular haemodialysis was continued. During the next few days she was able to initiate speech, move her tongue and buccooral muscles. She progressively regained her ability to swallow. On EEG performed 24 h after initiation of treatment, the spiky activity seen earlier had disappeared. Diffuse slowing of the background with gradual improvement was observed over a few days. An EEG performed 9 days later was normal (fig 1, right panel).

Two weeks after admission, owing to complete recovery, the treatment for epilepsy was gradually discontinued. Since then, the patient's neurological status and repeated EEGs have been normal.

Discussion

The clinical signs in this patient were consistent with FCMS. This condition, which may be congenital or acquired, persistent or intermittent, includes severe anarthria, loss of voluntary muscular functions of the face and tongue, and impaired mastication and swallowing, with preservation of reflex and autonomic functions.[1] The aetiology of FCMS is heterogeneous: vascular insults in adulthood, such as bilateral subsequent strokes; infections of the CNS, such as herpes simplex encephalitis or acute disseminated encephalomyelitis. FCMS can be congenital, owing to bilateral dysgenesis of the perisylvian region. FCMS as a manifestation of epilepsy has been described nearly exclusively in childhood.[2 3]

The clinical picture of our patient was initially attributed to an overdose of oxycodone. Only consciousness and not the facial, pharyngeal and lingual and mastication movements, however, improved with discontinuation of the treatment. Uraemic aetiology could not be implicated, as there was no change in the renal status during the whole episode because the patient was regularly haemodialysed.

NCSE was diagnosed only by EEG, which showed an electroencephalographic pattern compatible with the diagnosis. NCSE seems to be associated with a high mortality and morbidity, justifying aggressive treatment.

Oxycodone hydrochloride is an opiate derivative. Oxycodone and its metabolites are excreted primarily through the kidneys. In cases of renal failure, precautions should

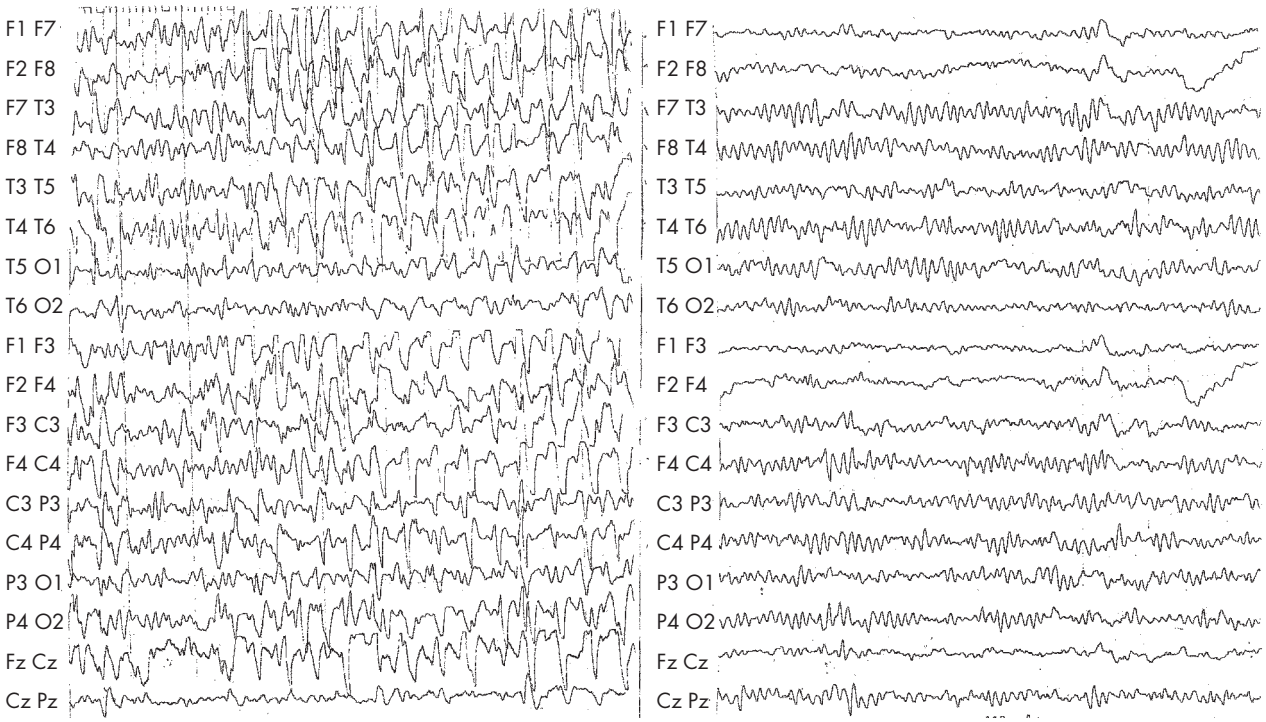

Figure 1 Left panel: electroencephalograph (EEG), recorded during the florid phase of the opercular syndrome, shows diffuse slow-wave activity and generalised repetitive synchronous sharp-wave complexes. Right panel: 9 days later, the EEG shows a 9–10 Hz background rhythm and disappearance of the sharp-wave complexes.

be taken to avoid an overdose that can be reached at lower doses.[4] Impaired consciousness, stupor, coma and seizures are all described as side effects of this drug, especially in the case of overdose or intoxication. In our case, the addition of oxocodone–acetaminophen may have caused an additive CNS depression,[4] lowering the threshold for epileptic activity, in a patient at risk because of the underlying silent CNS pathology. Treatment for epilepsy reversed both the clinical opercular signs and the abnormal EEG recordings. Our case recommends exercising caution in the use of oxycodone in patients undergoing dialysis.[4]

The association of an opercular syndrome with an NCSE in an adult patient is highly unusual. In their report on three cases of opercular syndrome, Thomas *et al*[2] describe an adult patient with myoclonic status epilepticus. Surface EEG, however, showed no evidence of cortical epileptic activity. The aetiology of these cases was either vascular or tumoral. Sasaguri *et al*[5] described a patient with epilepsy, with a corpus callosum defect, who developed FCMS at the age of 27 years. In this case, however, FCMS was due to chronic herpes simplex encephalitis.

FCMS has only rarely been described as a manifestation of epileptic activity, and has been explained as a para-ictal phenomenon, similar to the Landau–Kleffner syndrome by Shafrir and Prensky.[3]

The present case should draw attention to the possibility that an opercular syndrome in an adult may be due to NCSE and that in instances of FCMS the possibility of reversible (treatable) electrical activity should be ruled out.

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Successful immunosuppressive treatment and long-term follow-up of anti-Ri-associated paraneoplastic myelitis

Antineuronal antibody-associated paraneoplastic neurological syndromes (PaNSs) result from tumour-stimulated autoimmune attacks against components of the nervous system. The rare antineuronal antibody anti-Ri (ANNA-2) was initially thought to be closely associated with paraneoplastic opsoclonus–myoclonus syndrome. Recently, however, it has been found in several other PaNSs.[1] First-line treatment in PaNSs is removal of the underlying tumour. Second-line treatment is immunosuppression, which, although extensively used, especially when no tumour is detected, is often ineffective. Often, patients die from relentlessly progressive PaNS rather than the underlying neoplasm.[2] Here, we report the 2-year follow-up of an anti-Ri-positive steroid-responsive myeloneuropathy. No tumour was detected. Immunosuppressive treatment was tailored on the basis of clinical relapses, inflammatory changes in cerebrospinal fluid (CSF) and somatosensory evoked potentials (SEPs).

A 65-year-old woman, a retired administrator, was referred to our department with a 10-month history of progressive gait difficulties and ascending sensory loss in her legs, eventually having become wheelchair bound. Her medical history was unremarkable apart from enlarged axillary lymph nodes that were excised 9 months earlier. Histological examination showed only inflammatory changes. She had received hormone replacement therapy for 11 years. She never smoked.

On neurological examination, deep tendon reflexes were absent. She had spastic paraparesis with bilateral extensor plantar responses. Bilateral hypaesthesia for light touch up to the knees and diminished vibration sense were observed distally in her legs. Magnetic resonance imaging (MRI) showed symmetrical T2-hyperintense multisegmental (C6–TH3/TH8–TH12) cervicothoracal lesions of the spinal cord, with gadolinium enhancement restricted to the lateral parts (fig 1A). Cerebral MRI showed only minor microangiopathic changes. Nerve conduction studies showed normal tibial nerve conduction velocities (NCV), slightly reduced right median NCV (forearm 41 m/s, normal >41 m/s) and moderately reduced sural NCV (32 m/s, normal >40 m/s). Right median (32 ms, normal 28 ms) and tibial (61 ms, normal 52 ms) nerve F-wave latencies were prolonged, in keeping with polyneuropathy. SEPs after median nerve stimulation were normal; tibial SEP was absent bilaterally. An examination of the CSF showed pleocytosis (80% lymphocytes) and disturbed blood–CSF barrier (fig 1B). Microbiological serology of serum and CSF showed no evidence of acute infection. Routine laboratory tests were normal. Rheumatoid factor, anti-neutrophil and anti-nuclear autoantibodies were not found. Immunofluorescence screening for onconeural antibodies using monkey cerebellum, jejunum and peripheral nerve showed high-titre antineuronal nuclear antibodies in serum (1:1920) and CSF, with a pattern suggestive of anti-Ri antibodies. Subsequent immunoblots with recombinant targets of anti-Ri, anti-Hu, anti-Yo and amphiphysin antibodies (serum or CSF) confirmed anti-Ri specificity. In addition, no anti-Ma2, anti-CRMP5, anti-ANNA-3, anti-PCA2, anti-PCA-Tr, anti-N or anti-P/Q-calcium channel, anti-striated muscle or anti-acetylcholine receptor antibodies were found. Whole-body fluor-deoxyglucose-positron emission tomography (FDG-PET) was inconspicuous. Mammography showed bilateral mastopathy. Carcinoembryonic antigen, CA15-3, CA125, α-fetoprotein, human chorionic gonadotropin, and β2-microglobulin were not raised.

The patient was treated with high-dose steroids for 14 days. In weeks, she regained the ability of walking with support. Sensory disturbances improved. Plantar responses became flexor. A first relapse (relapse 1, fig 1B) led to a recurring inability to walk unaided, reappearance of extensor plantar responses and sensory deficits for all qualities below TH12. Spinal MRI showed increased signal abnormalities and gadolinium enhancement. The number of white blood cells in the CSF increased and blood–CSF barrier dysfunction became pronounced. Although a high-dose steroid treatment and slow tapering led to improved muscle strength, reduced sensory disturbances and increased walking distance as well as normalisation of abnormalities of the CSF, in the ensuing year, two more relapses (fig 1B) were witnessed, each after tapering steroids below 20 mg prednisolone and associated with the reappearance of inflammatory CSF (fig 1B). Increased prednisolone was followed by clinical improvement and alleviated abnormalities of the CSF. Peroneal SEP latencies were established as an additional parameter. Every relapse led to increased residual deficits. Eventually, oligoclonal bands appeared and remained detectable, mild pleocytosis persisted and peroneal SEP continued to deteriorate. As steroidal side effects were no longer tolerable, monthly cyclophosphamide-pulsed treatment (650 mg/m^2 body surface) was started. During the following 9 months, the patient's condition stabilised and CSF findings normalised. Six-monthly FDG-PET did not disclose an underlying malignancy. Owing to side effects, cyclophosphamide was eventually discontinued. As azathioprine and mycophenolate mofetil led to lymphopenia, intravenous immunoglobulins were started recently. So far, there has been no relapse.

This patient with anti-Ri antibodies presented with myeloneuropathy. The non-classical clinical presentation in combination with a well-characterised onconeural antibody fulfils the criteria of a definite PaNS.[3] That no tumour was detected during 36 months is compatible with anti-Ri PaNS. Only 3% of patients have not developed cancer within 3 years of follow-up.[1 3] The history of enlarged lymph nodes, oestrogen treatment and mastopathy is suggestive of