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Short communication

A case of Parry–Romberg syndrome and alien hand

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ABSTRACT

Parry–Romberg syndrome (PRS) is a rare condition characterised by progressive hemi-facial atrophy. Here we present a PRS case with alien-hand syndrome, which has not previously been described in adult onset disease. On the basis of the presumed auto-immune pathology of PRS we justify the treatment strategy we successfully used in this patient. A review of the literature was extensively done for understanding the history of alien hand sign over the years.

1. Introduction

Parry–Romberg syndrome (PRS; also known as Romberg's disease or progressive hemi-facial atrophy) is a rare disease, causing progressive wasting of the skin, subcutaneous fat, muscle and occasionally bones of the face. It most commonly involves dermatomes of one or more branches of the fifth cranial nerve [1]. Occasionally it can extend to involve the ipsilateral or contralateral limbs [2,3]. The clinical course typically begins in early childhood, with females being more commonly affected than males [4]. It can progress for 2–20 years [1].

Many associations have been documented in the literature. Headaches, facial pain and seizures (particularly partial seizures) are relatively common [5,6]. A survey of 205 patients in 2003 reported the presence of these symptoms in 52%, 46%, and 11% (4%) respectively [7]. Eye and visual problems were reported by 46% of the patients and 17% had history of uveitis. Other specific ophthalmic complications reported from the literature include enopthalmos with various eyelid modifications (ptosis, retraction, atrophy) [8]. Amblyopia and diplopia may be due to ocular nerve involvement, muscle paresis or restrictive oedema [9,10]. Visual acuity can also be affected by retinal vasculitis [11], optic atrophy [12] and uveitis [13].

Arteriovenous malformations and intracranial aneurysms have also been reported [14,15], however these findings occur less commonly in the literature.

Only one case of alien hand syndrome with PRS has been described and this was a paediatric patient [16].

The exact pathophysiology of the disease is unknown though many support the theory that it has an autoimmune aetiology [17], as there are features that overlap with scleroderma and en coup de sabre syndromes [18]. Hence treatment strategies involving steroids and immunosuppressants have been used [6,19,20] in PRS with some success.

Here we describe the case of a 36 year old patient with PRS from childhood, who presented with new-onset headache, seizures and unilateral alien hand syndrome. We discuss the neuroradiology, investigations and treatment.

2. Case report

A 36-year-old Caucasian left handed lady with previously diagnosed PRS, and a history of bulimia, depression and alcohol abuse (50 units/week), was admitted (day 1) after an episode of loss of consciousness. The day after admission she suffered status epilepticus

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which was treated with Lorazepam and Phenytoin and required intubation. Her prior medication was Fluoxetine and Disulfiram only. Her relatives commented that a few days before the event she complained of intermittent right temporal headache with vomiting, episodes of confusion, visual hallucinations consisting of seeing heads or bodies of people disproportionately large, and auditory hallucinations where she could hear music for hours not heard by other members of the family.

Brain MRI (Fig. 1A) was performed on day 2 and showed right cranio-facial atrophy which was obvious on clinical examination and had been documented previously. It also showed previously undocumented left cerebral hemisphere atrophy consistent with alcohol abuse, subtle thickening and signal alteration on T2-weighted images of the cortex of the right cerebral hemisphere and ipsilateral thalamus, consistent with changes related to the previous status epilepticus.

On day 3, once Glasgow Coma Scale returned to normal following extubation, neurological examination revealed ideo-motor apraxia only with the left hand. Indeed she was unable, when asked, to perform with the left hand some activities as brushing teeth or the hair or showing how to put a picture on the wall. It was also noticed that the first three digits of the left hand were held at times in a flexed position with modest bradykinesia but no clear dystonia or dyskinesia was observed. The assessment of orientation in time and space, strength, tone, language (understanding and speech), cerebellar system, cranial nerves, was normal but at that time visual field evaluation was not done. Tendon reflexes were present in the upper and lower limbs with equivocal plantars and no clonus. She reported apallesthesia of the left big toe and reduced proprioception on the left side.

Her initial tests, taken in intensive care revealed only mild neutrophilia with a normal biochemical profile. Her ANA, nDNA, ANCA, ACA and lupus anticoagulant were negative. Wilson's disease was excluded with normal ceruloplasmin, serum and urinary copper levels.

She had further visual hallucinations, noted on day 8, when she reported seeing people from her past. EEG performed on day 9 showed that the right hemisphere appeared epileptogenic without a clear focus.

Lumbar puncture was performed on day 13. Cerebrospinal fluid (CSF) appeared blood-stained with 7 lymphocytes/cm^2 but no organisms seen or cultured. CSF protein was slightly raised at 0.43 g/L (NR 0.2 — 0.4 g/L). Serological tests for syphilis, Lyme disease (IgG and IgM) and HIV were negative. HSV, varicella zoster, enterovirus and parechovirus were not detected by PCR. On the basis of the episodes of confusion with hallucinations, seizures and 100% CSF lymphocytosis with a slightly raised protein, she was initially treated with acyclovir for possible viral encephalitis (this was stopped on day 20 when the PCR for HSV result came back negative).

On day 14, the patient stated that she wanted to discharge herself from hospital against medical advice. During examination of capacity to make this decision, through a thorough neurological examination a left homonymous hemianopia was incidentally observed during the assessment of the visual field test. The patient did not notice it and did not want to look into. She discharged herself but was given Carbamazepine to prevent further seizures and vitamin B strong given her alcohol history and bulimia.

She represented 3 days later, day 17 in relation to initial presentation, with further headache and vomiting.

Minimental state examination score on day 20 was 27/30 making one mistake in orientation by giving the wrong month, unable to write a sentence and copying the design just with her left hand (Fig. 1B, C).

A head CT angiogram on day 21 to explore a possible subarachnoid haemorrhage was negative, but revealed a hypodense region in the right parieto-occipital hemisphere (not shown). The left homonymous hemianopia was confirmed by visual field testing on day 24 and was not attributable to disease of the optic nerve or retina. The neurological

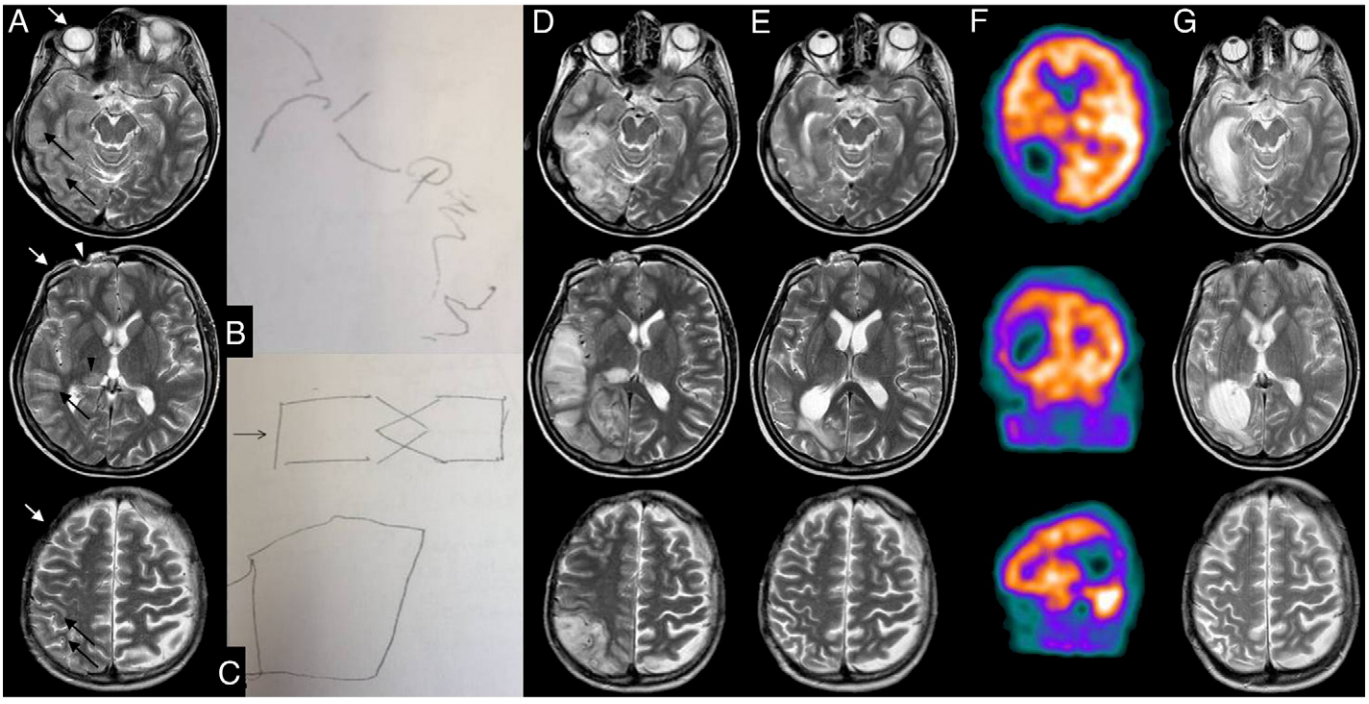

Fig. 1. Neuroimaging of the brain and patient's left hand activity. Serial non-consecutive T2-weighted axial magnetic resonance (MR) images (A) obtained during status epilepticus on day 2 show the right cranio-facial brain atrophy (white arrows), and a subtle thickening and high-intensity signal (long black arrows) of right temporal, occipital, and fronto-parietal cortex, as well as high-intensity signal (black arrowhead) of the pulvinar of the ipsilateral thalamus. Note also some image distortion (white arrowhead) in the right frontal region, consistent with metallic foreign body. When status epilepticus had been treated, the patient's left hand does not provide understandable writing (B), and does not repeat the drawing of intersected pentagons (short black arrow) below them (C). At brain MRI obtained at re-admission (D) on day 24, the right cerebral hemisphere shows marked cortical–subcortical swelling and thalamus signal alteration. On day 57 (E), brain MRI shows clear-cut atrophy, and reduced signal alteration. On day 83, axial, coronal and sagittal single photon emission computed tomography images (F) show the absence of perfusion in the right temporo-parieto-occipital lobes that were found further involved at brain MRI on day 96 (G).