



Case report

Alien hand and complex regional pain syndromes during rehabilitation program

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ARTICLE INFO

Article history

Received 20 April 2020

Accepted 27 September 2020

Available online 7 April 2021

Keywords

Rehabilitation

Complex regional pain syndrome

Corticobasal degeneration

Alien hand syndrome

Limb fracture

Doi

<https://doi.org/10.29089/2020.20.00150>

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ABSTRACT

Introduction: Alien hand syndrome (AHS) belongs to the group of asymmetrical movement symptoms that are a characteristic picture of neurodegenerative diseases such as corticobasal degeneration syndrome (CBS). Changes in the musculoskeletal system such as dystonia, bradykinesia and myoclonus may also occur in the subacute stage of complex regional pain syndrome (CRPS) type I.

Aim: To learn about difficulties related to diagnosis and rehabilitation of a patient with AHS and CRPS type I complicated by an upper limb fracture.

Case study: A case of a patient admitted to the rehabilitation department with compulsive unilateral involuntary groping and grasping movements of the left hand for about half a year is presented. The woman has been suspected of CBS. A few months after the diagnosis, the patient was admitted to the rehabilitation ward, where she suffered an elbow fracture during exercise. Two months after fracture, type I CRPS was diagnosed.

Results and discussion: AHS in CBS and CRPS type I may have a similar clinical picture, which makes differentiation difficult. It is very rare that both diseases coexist with each other. They can also lead to a number of unwanted symptoms such as limb fractures.

Conclusions: CRPS may increase the symptoms of dystonia due to other causes. Patients with AHS and dystonia are more likely to break because of rapid movements alone or because of immobilization and osteoporotic changes. As a result, treatment and rehabilitation cannot be based on a questionable diagnosis of a neurological syndrome.

1. INTRODUCTION

Alien hand syndrome (AHS) is a rare movement impairment diagnosed in such diseases as cortico-basal syndrome (CBS), stroke, prion disease, tumors, etc.¹ It is characterized by involuntary movements of the patient's limb with a simultaneous feeling of losing the ability to regain the ability to 'control' his or her own limb¹. The characteristic symptoms of CBS are also asymmetrical movement symptoms such as stiffness, bradykinesia, dystonia, apraxia. CBS is a chronically progressive neurodegenerative disorders which affect the cerebral cortex and basal ganglia. Due to the complex pattern of disorders and their unstable occurrence, CBS remains a challenge for both diagnosis and treatment.

Complex regional pain syndrome (CRPS) is a disease which main symptom is persistent pain. In addition, pain is accompanied by autonomic and/or trophic disturbances. The factor indicating the occurrence of CRPS is often a minor or severe trauma to the limb, which was not accompanied by damage to the nerve structures.

In the course of the above disease entities there are symptoms that are common to them. These are usually motor changes such as dystonia, bradykinesia, myoclonus and tremor. Rehabilitation treatment in these syndromes can be difficult and must be based on the current symptoms of the disease.

2. AIM

Herein, we report a rare case of CBS with AHS complicated with upper limb fracture during rehabilitation program with overlapping CRPS symptoms.

3. CASE STUDY

A 71-year-old right-handed female patient about half a year before admission to neurological department noticed left hand grasping and groping of objects movements, for which she ultimately sought medical advice. A neurological examination revealed bradykinesia, postural instability, upper limb rigidity, myoclonic movements in the left hand with AHS and dysarthria (Table 1). The left hand movements aggravated during action and there was a stimulus-sensitive component. Apraxia was difficult to diagnose because of rigidity and left hand movements. The patient also had cognitive impairment, mostly affecting executive and visuospatial functions. She also presented symptoms of depression and behavioral changes such as irritability. An MRI revealed asymmetrical cortical atrophy in the fronto-parietal region that was more severe in the right hemisphere. CBS was suspected. Treatment with L-3,4-dihydroxyphenylalanine (L-DOPA) was started with no effect. The patient was hospitalized for the second time in a specialized movement disorder department, where single-photon emission computed tomography (SPECT) hexamethylpropyleneamine oxime (HMPAO) should have been performed as part of

Table 1. Symptoms of CBS and CRPS in presented patient.

Symptoms	CBS	CRPS
Presentation	asymmetrical	asymmetrical
Higher cortical features:	AHS in left hand, in right only motor sequence programming difficulties	no
Cognitive impairment	executive and visuospatial functions,	no
Rigidity	yes	permanently worsening after CRPS occurrence
Dystonia	mild	permanently worsening after CRPS occurrence
Myoclonus	mild	mild
Athetosis	no	yes, after CRPS occurrence
AHS	yes	no longer after CRPS occurrence
Pain	no	yes, severe after CRPS occurrence, allodynia

the CBS work-up. Unfortunately the patient stopped the examination immediately after the start because of anxiety. The probable CBS was also diagnosed according to criteria proposed by Armstrong et. al 2013.¹ The patient was admitted to a rehabilitation ward after 3 months, mostly in order to improve the performance of the upper limb. A few days after starting rehabilitation (non-weight bearing exercises, exercises according to physiotherapeutic methods), the patient reported severe pain in her left hand a few hours after a physical therapy session. An X-ray examination revealed a fracture of the styloid process of the left ulna. There was no evidence of osteoporosis which was also confirmed by laboratory tests. Initially, the limb was placed in an orthopedic cast; however, the cast had to be removed a day later due to intensification of involuntary left hand movements (AHS, myoclonus), which the patient could not tolerate. The patient was then fitted with a kind of soft orthosis for a period of 4 weeks, but she was still in considerable distress. A radiograph after 4 weeks revealed the presence of bone union, and the patient remained under the care of an outpatient neurologist (Figure 1). After 2 months she came to the outpatient clinic and reported paroxysmal pain, stiffness and erythema in her broken limb that had begun about 3 weeks before. A neurological examination showed a dystonic posture of the left upper limb (flexion of left elbow), skin erythema and oedema with mild sweating changes; intense pain visual analogue scale (VAS) pain score was 8/10, the pain was characterized by patient as continuous burning; allodynia was also diagnosed. There was no AHS, only slight myoclonic movements were noted (Table 1). A radiograph revealed osteoporosis. CRPS type I was suspected and gabapentin was prescribed. Because of severe dystonic flexion in the left elbow, the botulinum toxin was administered and produced improvement. Because of hostility and depression the psychotherapy was proposed, but the patient refused this kind of therapy. The patient was in considerable distress with chronic pain and depression and

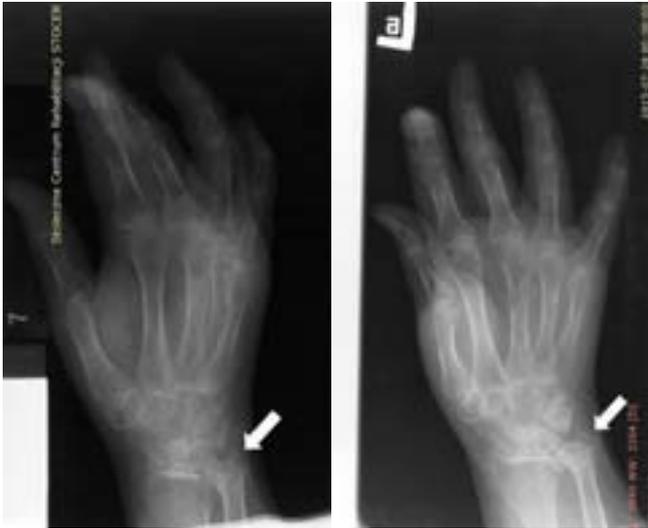


Figure 1. Radiographs showing fracture of the styloid process of left ulna in lateral (A) and anteroposterior (B) view. White arrows indicate the fracture site.



Figure 2. A few months after injury, edema and dystonia with athetosis in left hand, no AHS.

did not want to continue neither her rehabilitation program nor botulinum toxin therapy. A few months later, a neurological examination revealed a predominance of dystonic and athetotic movements (Figure 2). She discontinued her neurological outpatient care and was lost to follow-up.

4. RESULTS AND DISCUSSION

To our knowledge, this is the first report in the literature of AHS as a probable feature of CBS and complicated by an arm fracture during rehabilitation program. We found one case study of corticobasal degeneration presenting as CRPS type I.² We consider that our patient fulfilled the criteria of CBS proposed by Armstrong et al.¹ The development of AHS in CBS usually lasts from several months to several

years. It is usually associated with progressive neurodegeneration. In our patient, AHS progressed but after the appearance of type I CRPS, the symptoms systematically disappeared, as if 'blurred' by dystonic symptoms intensifying as a result of CRPS. Both CRPS and CBS may affect the same areas of the central nervous system (CNS): thalamus, basal ganglia (BG), and cerebral cortex. In our presented case, we can speculate, that these two processes overlapped and could damage both BG and thalami, which had an additive effect on the patient. In the literature we find a high incidence of dystonia in both these syndromes. Van Rjin sets it at 91% and 62%, respectively, in CRPS and CBS.³ We do not know the mechanism of dystonia in the course of CRPS. It seems that the neural structures mediating sensory-motor integration may be important. The factor that causes the development of CRPS is usually minor peripheral nerve damage. However, due to the more and more frequent clinical reports supported by neuroimaging, the BG are structures of significant importance in the mechanism of CRPS development.⁴ These considerations are largely based on the fact that BG are associated with the integration of information between the cortical and thalamus areas and in the processing of pain, especially in terms of its modification.⁵ BG and the basal-thalamic-frontal cortex circuit dysfunction can contribute to dystonia and proprioception deficiencies that sometimes occur in CRPS.

AHS treatment, despite being sometimes effective, often lasts from several months to several years. The best results are obtained in the group of patients who had AHS as a result of stroke. The management is based mainly on various rehabilitation programs, however, as part of therapy, pharmacological treatment or botulinum toxin may also be used.⁶ As part of the rehabilitation program, mirror therapy or visual-spatial training techniques are recommended.⁶ We do not know much about the effects of exercise on tauopathies or other Parkinson's diseases. There are casuistic reports indicating some efficacy of repetitive facilitation exercise (RFE) in combination with occupational therapy in patients with AHS in the course of CBS.⁶ In our case, the rehabilitation program ended a few days after the patient was admitted because of an upper limb fracture. We cannot be sure that the fracture was associated with rehabilitation program. To our knowledge, ours is the first report of a fracture related to these abnormal movements (AHS). The management of this fracture posed a challenge because the patient could not tolerate an orthopedic cast. In the literature, such injuries are not often described. We have found reports of injuries in the course of cervical dystonia, as well as one case report in which a metatarsal bone fracture occurred in the course of dystonia caused by levodopa treatment.⁷

Patients with CBS often experience behavior and cognitive disturbances.¹ Our patient showed signs of depression and behavioral changes in the form of increased irritability. The severity of changes was also correlated in time with the diagnosis of CRPS. A long time of treatment and a number of undesirable events meant that the patient was discouraged to health care and did not want further medical visits.

Diagnosis of CRPS is often a challenge, but we believe that our patient has met the clinical diagnostic criteria of the New International Association for the Study of Pain (IASP).⁸ The most visible symptom of our patient's CRPS was left hand pain (mainly from the elbow to the fingers). In post-traumatic patients – as in our case – these clinical criteria can help the doctor make the right diagnosis. Diagnosis of CRPS in other disease entities, including after a stroke, seems more complex. In stroke patients, symptoms such as edema, increased pain, a change in the warming of the limb, abnormal touch, or dystrophic changes in the skin may be associated with paresis or subsequent neglect syndrome. The difference in CRPS symptoms in stroke patients may also consist of the characteristic distribution of pain, which mainly affects the shoulder and wrist area, and does not involve the elbow area. Such a syndrome is commonly known as shoulder-arm syndrome (SHS). Three-phase bone scintigraphy may help to confirm this diagnosis. Options for effective CRPS treatment are limited. Both pharmacological treatment and physiotherapy are used. Therapy should begin with appropriately selected physiotherapy with elements of physical therapy. Our patient after rehabilitation did not want to have contact with physiotherapists, because a broken arm was associated with rehabilitation. Due to the fact that the patient's main complaint was pain, pharmacotherapies focused on its treatment and were based on gabapentin. The choice of gabapentin was due to the fact that it has been shown to be effective in treating neuropathic pain in CRPS.⁹ Despite strong evidence of the effectiveness of bisphosphonates, they were not used in our patient because of the refusal due to the high price. We did not use any oral muscle relaxants or spasmolytics, because of severity of dystonic movements and we decided to use botulin toxin with improvement. We did not consider intrathecal use of these drugs, because we were afraid of insufficient cooperation on the patient's side. In the light of still insufficient evidence for the effectiveness of the therapy used, the prevention of CRPS should be taken into account. Reducing the risk of CRPS can be obtained by using vitamin C immediately after injury or orthopedic surgery. Such prophylaxis should be used for 40–45 days.¹⁰ In patients who developed CRPS in the form of SHS after stroke, careful prophylaxis of the joint with shoulder recentralization, shoulder support, as well as in the case of subluxation of the brachial-ligamentous ligament electrostimulation of the deltoid and supracarpal muscles can be used as prophylaxis.

5. CONCLUSIONS

CRPS and CBS share common characteristics, including dystonia. In our patient, dystonic movements intensified significantly after limb fracture, resulting in the disappearance of continuous hand movements (AHS). Patients with AHS and dystonia are at risk of fractures due to rapid movements alone or due to immobilization and osteoporotic changes. We believe that CBS patients with AHS or dystonia are at risk of bone fractures, which should be considered during exercise programs.

Conflict of interest

None declared.

Funding

None declared.

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