CASE REPORT

Sudden hemichorea and frontal lobe syndrome: a rare presentation of unbalanced polycythaemia vera

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SUMMARY

Polycythaemia vera (PV) is an haematological neoplasm that frequently presents neurological symptoms. However, chorea is a rare complication of this disease, occurring in less than 5% of the patients. Cognitive impairment related to PV unbalanced is also a rare complication, and it can improve with proper treatment. We present a 96-year-old-man with acute-onset hemichorea and frontal lobe syndrome with no vascular pathology in the basal ganglia or frontal region. A clear relationship was observed between the onset of involuntary movements and the cognitive impairment and worsening of haematological parameters in the patient. After causal and symptomatic treatment, the patient's clinical status improved. In the elderly, PV must be considered as a cause of acute chorea and sudden cognitive impairment, as early diagnosis leads to effective treatment and prevention of complications.

BACKGROUND

Chorea is a hyperkinetic movement disorder occurring mostly due to infectious, vascular, hereditary, drug-induced or metabolic causes.¹

Polycythaemia vera (PV) is a myeloproliferative neoplasm characterised by elevated haemoglobin (Hb) and bone marrow panmyelosis.²

Neurological manifestations of PV such as headache, vertigo and transient ischaemic attacks are frequent (50%–78%), but chorea is rare and infrequently reported as a complication of the disease (1%–5%).³

Polycythaemic chorea manifest itself after the age of 50, and most cases have an acute onset or present as a sudden aggravation of a pre-existing symptom. The choreic syndrome is usually generalised with predominantly faciobrachial muscle involvement.^{4,5}

The frontal lobe syndrome in PV has been described once, and cognitive impairment with partial reversibility on lowering of the haematocrit (Ht) has been reported before.^{6,7}

Because of these uncommon symptoms, it is needed to investigate the presence of PV as a potentially treatable cause in elderly patients with new-onset movement disorder.

Our aim is to present a patient with hemichorea and frontal lobe syndrome as the main manifestation of unbalanced PV.

CASE PRESENTATION

A 96-year-old man presented to our emergency room with sudden-onset left-sided involuntary

movements involving the upper limb and face that had developed 4 days earlier. Concomitantly, he had demonstrated progressive behavioural change which family members described as social disinhibited behaviour that included inappropriate verbal comments and unrestrained laugh.

His medical history included hypertension, atrial fibrillation (treated with a direct anticoagulant agent, apixaban) and chronic lower limb ischaemia. He also has a PV that was treated with hydroxyurea for a long time, but the haematologist had decided to retire a month ago.

He was not being treated with chorea-induced drugs such antiparkinsonian drugs, antipsychotics, amphetamines or tricyclic antidepressants.

There was no family history of movement disorders, dementia or psychiatric illness.

His physical showed choreic movements involving orofacial and brachial muscles. These movements were random and fleeting from one part to the body to another, causing difficulty in day-a-day activities. Neuropsychological testing showed mild cognitive impairment, with predominantly extensive frontal symptomatology (tendency to imitate, disinhibition...).

There was no clubbing, splenomegaly or evidence of peripheral venous thrombosis.

Cardiovascular examination was unremarkable.

INVESTIGATIONS

On hospital admission, laboratory testing showed elevated Hb (15.5 gr/dl, reference 12–15), higher Ht than previous (46%, reference 35–45) and elevated mean corpuscular volume (MCV). In retrospect, Hb, MCV and Ht values had been rising since 1 month before presentation (figure 1).

Other laboratory tests were normal, including coagulation indicators.

The sudden appearance of left-sided hemichorea in a patient with a former Atrial fibrillation (AF) raised the suspicion of a vascular event in the basal ganglia region, however a CT of the brain was normal (figure 2).

The patient had a pacemaker that was not compatible with MRI, so it could not be performed.

The absence of vascular stenosis, both in supra-aortic trunks and in the polygon of Willis, was also confirmed by a neurosonological study that was normal.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of chorea is shown in table 1.

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Figure 1 Haematocrit levels.

TREATMENT

As the violence of involuntary movements required symptomatic treatment during follow-up, at first, haloperidol was initiated which was not effective. Then it was decided to start clonazepam 2mg every 8 hours which was not effective either. The improvement had to do with the establishment of hydroxyurea as previously.

OUTCOME AND FOLLOW-UP

Following the PV treatment, a decrease in the severity of involuntary movements was observed. Unfortunately, because of the symptomatic agents his level of consciousness decreased, and he suffered aspiration pneumonia.

DISCUSSION

Although neurological manifestations of PV, such as headache, dizziness, paraesthesias or stroke, are frequent, chorea is a rare symptom with a prevalence of 1%–5%.³ There are some case reports that described reversible cognitive impairment in PV, and in one of them the authors reported a frontal lobe syndrome similar to our patient.^{6,7}

Figure 2 CT of the brain.

Table 1 Differential diagnosis of chorea

Type of chorea	Characteristics
Inherited/degenerative choreas: <ul style="list-style-type: none"> ▶ Huntington disease. ▶ Benign hereditary chorea. ▶ Huntington-like syndromes. ▶ Neuroferritinopathy. ▶ McLeod. ▶ Neuroacanthocytosis. 	Onset typically >18 years Gradual onset
Autoimmune chorea: <ul style="list-style-type: none"> ▶ Antiphospholipid syndrome. ▶ Sydenham's disease. ▶ Paraneoplastic syndromes. 	Onset typically >18 years Acute onset
Other choreas: <ul style="list-style-type: none"> ▶ Vascular. ▶ Demyelinating. ▶ Pharmacological. ▶ Metabolic: polycythaemic chorea, disorders of the thyroid, glucose, sodium... 	Onset typically >18 years Acute onset Variable age of onset, acute onset Onset typically > 60 years Acute onset

Polycythaemic chorea manifests itself predominantly after the age of 50, making PV the first disorder to be considered in cases of senile chorea. Most cases have an acute onset, and the syndrome is usually generalised with predominantly orofacial and brachial involvement.⁴

Although the chorea caused by PV is due to metabolic cause, in most of the cases described in the literature, its presentation is unilateral, so in this case, the decompensation of the patient's base disease, the improvement with normalisation of the Ht levels and the absence of vascular alterations would justify the clinical presentation in this patient.^{3,4}

The majority of the choreic or cognitive manifestations appear after the exacerbation of haematological parameters. This supports the theory that hyperviscosity is the determining factor in the pathophysiology. In the case of chorea, neostriatal hyperviscosity in the brain leads to venous stasis, low brain blood flow and impaired oxygen and glucose metabolism, causing movement disorder. It is possible that the pathophysiology of frontal lobe syndrome is based on the same principle as polycythaemic chorea.⁸

Proper treatment of PV and restoration of Hb and Ht levels lead to a reversibility of choreic and cognitive symptoms. In the treatment, venesection appears to be the initial method of choice, but bone marrow suppressor agents should also be given.

In the present case, PV was diagnosed and successfully managed by hydroxyurea; it illustrated the need to investigate the presence of polycythaemia as a potentially treatable cause in elderly patients with new-onset movement disorder.

Learning points

- ▶ Chorea is an hyperkinetic movement disorder occurring mostly due to infectious, vascular, hereditary, drug-induced or metabolic causes.
- ▶ Neurological symptoms are frequent in some haematological diseases as polycythaemia vera (PV).
- ▶ Polycythaemic chorea manifests itself predominantly after the age of 50, making PV the first disorder to be considered in cases of senile chorea.

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