CASE REPORT

An unusual psychiatric presentation of polycythaemia 'Difficulties lie in our habits of thought rather than in the nature of things' Andre Tardieu

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SUMMARY

Psychiatric symptoms are not always best explained in the context of psychological stress. The same mental, emotional and behavioural changes also arise from various medical conditions. For a clinician this dual origin of psychiatric symptoms creates an ongoing diagnostic challenge. Our patient is a 50-year-old gentleman who had been working in a company for around 33 years and always had good appraisals. He presented to mental health services with a 5-year history of persecutory beliefs, convinced that his employers were out to damage his reputation. Apart from a diagnosis of polycythaemia, a few months before the onset of abnormal beliefs, there is no personal or family history of psychiatric disorder or medical illness. His delusions did not respond to conventional treatment with psychotropic medication possibly due to non-adherence because of side effects. However, a series of venesections lead to an improvement in mental state.

treatment with psychotropic medication and to minimise delay of appropriate treatment.

CASE PRESENTATION

Our patient was referred to mental health services at age 50 by occupational health due to stress at work. He had been working in a company for 33 years and always had good appraisals. His difficulties appear to have started around 5 years ago when he noticed rumours were being spread that he may be gay. He reported that these rumours stopped when he denied them. Later he believed stories were being spread indicating that he was not a team player and that he was a difficult character. He became increasingly concerned when reports in his laptop were being changed and disappearing to discredit him and he managed to identify specific people in the company who were involved much to his distress. He also felt that he was being tracked on his mobile phone, that there were people following him, sent by employees of the company to intimidate him.

Unable to tolerate the torments he moved to a new company a year later but soon noticed a similar victimisation, which convinced him that his previous employers were in some way to blame. Consequently he resigned from the job and remains unemployed.

Recently during a visit to see his brother in Scotland, he disposed of an old GPS system in a public bin, as he felt unsafe to discard it locally. He was convinced that a controlled explosion destroyed the device as he saw a plain-clothes policeman observing his whereabouts at a bus stop on the following day. On returning home he was certain his house was broken into as he heard from neighbours there was a power cut in spite of which electronic clocks in the house had the correct time which convinced him that the break in was by a professional. He became distressed enough to seek the help of the police.

The patient has no previous psychiatric history. However before the onset of frank persecutory beliefs there was an incident on a plane when he became suspicious of a fellow passenger and informed an flight attendant but nothing came of this. In interactions with his niece and her partner, who was from the Middle East he became uneasy and uncomfortable.

Of note in his medical history was a diagnosis of polycythaemia made a few months before the onset of abnormal beliefs. This was identified when he

BACKGROUND

Psychiatric symptoms are not always best explained in the context of psychological stress. The same mental, emotional and behavioural changes also arise from various medical conditions.¹ For a clinician this dual origin of psychiatric symptoms creates an ongoing diagnostic challenge.

Polycythemia vera (PV) may not cause signs or symptoms for years and the disease is often found during blood tests for other reasons. Signs and symptoms occur as a result of the thick blood, which slows the flow of oxygen rich blood reaching all parts of the body leading to headaches, shortness of breath, abdominal discomfort due to an enlarged spleen, visual disturbances, pruritus and fatigue. Blood clots both venous and arterial are the most serious complications. Coronary and cerebral events are prominent and microvascular disturbances can also occur.² Together with transformation to myelofibrosis, acute leukaemia or both, thrombosis is the main cause of death.

Given the varied presentations of PV including neuropsychiatric symptoms across the spectrum it is important to keep an open mind about organic factors for all psychiatric symptoms particularly when there is no previous personal or family psychiatric illness, during late onset of psychiatric symptoms and when patients do not respond to conventional treatments. This is crucial to prevent iatrogenic harm by unnecessary pharmacological

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presented with a number of headaches. He is under the care of a haematologist and also consulting an urologist for a simple kidney cyst. He takes aspirin 75 mg daily. He is a non-smoker.

There is no family history of psychiatric illness. He was previously married for 22 years but separated around the time when he was diagnosed with polycythaemia. The reasons for their separation were unclear. There is no history of alcohol or illicit drug misuse. In terms of his premorbid personality, he has always focused on his family, loved his job, is conscientious and gets on well with people.

On mental state examination he was neatly presented, smiled appropriately and spoke at a normal rate. His face was red due to polycythaemia. He was not depressed. Concerning abnormal beliefs he displayed a delusional system involving his ex-employers but there were no abnormal perceptions. He was oriented in time, place and person. The patient did not think he suffered from a mental illness but was willing to consider treatment.

INVESTIGATIONS

A MRI brain scan did not reveal any abnormalities. Although his JAK-2 (a gene which is the main cause for primary polycythemia) was negative it was still felt he had primary polycythemia because of a low erythropoietin (secondary polycythemia is known to be associated with normal or high erythropoietin levels and not related to the JAK-2 gene).

DIFFERENTIAL DIAGNOSIS

- ▶ The patient's employers are indeed behaving in a conspiratorial way.
- ▶ He is suffering from paranoia, in this case a delusional disorder International Classification of Diseases (ICD) 10 Code F 22.0.
- ▶ PV is known to be associated with neuropsychiatric symptoms.

TREATMENT

The patient initially agreed to start an atypical antipsychotic medication and olanzapine was started at a dose of 5 mg, gradually increased to 10 mg. Because of the side effects of drowsiness he discontinued treatment after 2 months. However he had regular reviews with haematologist, receiving a series of venesections in succession which lead to an improved mental state. Although he still held a view about a conspiracy, he was not distressed as in the past.

OUTCOME AND FOLLOW-UP

*Guidelines for the diagnosis, investigation and management of polycythemia/erythrocytosis, British Journal of Haematology July 2005*³ recommend venesection to keep haematocrit below 0.45, aspirin 75 mg/day and cytoreduction (reduction of cells using one of the following agents: interferon α , hydroxycarbamide, anagrelide or busulfan). The choice of drug depends on age of the patient. Cytoreduction is indicated if patients do not tolerate venesection and if there is indication of disease progression.

DISCUSSION

The clinical presentation of our patient who has no personal or family history of psychiatric disorder was apparently at low risk for the development of a delusional disorder. Although the MRI brain scan was normal, his presentation may be best explained by abnormal cerebral blood flow (hyperviscosity) resulting in an organic psychosis. This view is supported by the

gradual reduction of symptoms and distress following successive venesections.

Erythrocytosis is used to identify a group of varied disorders with an increase in circulating red blood, that is, typified by a raised haematocrit. The term polycythemia is used when all three-cell lines are involved. Once a diagnosis is established it is desirable to identify any underlying aetiology and establish if the disorder is primary or secondary, although this may not be possible even after prolonged investigations. This is crucial as treating the underlying cause may improve secondary polycythemia³ (*Guidelines for the diagnosis, investigation and management of polycythemia/erythrocytosis British Journal of Haematology July 2005*).

A MEDLINE literature search using the words polycythaemia and psychosis showed several reports of PV associated with neuropsychiatry symptoms including acute psychosis⁴ delirium,⁵ mania,⁵ sudden onset of cognitive impairment,⁶ cerebral ischaemia,⁷ depression with psychotic symptoms,⁷ agoraphobia⁸ and progressive neurological deficits.⁹ In some of the reported cases treating the psychiatric symptoms using psychotropic medications only showed slight improvements. Both physical and psychiatric symptoms reduced considerably following venesection alone or in combination with cytoreduction.

The final diagnosis according to ICD 10 is most likely an Organic Delusional (Schizophrenia-like) Disorder (F06.2). This diagnosis is supported by the following:

- ▶ Persistent persecutory delusions dominating the clinical picture;
- ▶ Evidence of systemic physical disease, that is, PV;
- ▶ A temporal relationship between the development of the underlying disease and the onset of the mental syndrome;
- ▶ Recovery from the mental disorder following venesections;
- ▶ Absence of evidence to suggest an alternative cause of this mental syndrome.

His response to antipsychotic medication was difficult to assess mainly due to non-concordance because of side effects. The improvement to a series of venesections suggests that treatment with antipsychotic medication would not have been as effective as treatment for the physical disease.

Learning points

- ▶ Initial psychiatric assessments must be comprehensive and include routine blood investigations and a physical examination to rule out organic aetiology.
- ▶ Increasing awareness among healthcare professionals of the different presentations of polycythemia vera (PV) may prevent the risk of iatrogenic harm.
- ▶ Collaborative care through close liaison between mental healthcare professionals and haematologists will aid a symptomatic recovery, crucial in patients who are acutely psychotic and not responding to conventional treatments.
- ▶ Patients diagnosed with PV should be regularly monitored for neuropsychiatric symptoms.

Contributors RR is the main author who was involved in the clinical care of the patient while TP is the responsible clinician for the patient described in the case report. She scrutinised the contents of the report before submission.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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