Acute psychosis in polycythaemia rubra vera

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Summary

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A case of a young male patient is described who presented with polycythaemia rubra vera associated with an acute episode of schizophrenia-like psychosis. Psychiatric symptoms of the patient improved markedly following venesection and normalising of the haematocrit suggesting a causal relationship between the psychopathological and the haematological condition. The present report may emphasise the need of a careful screening for neuropsychiatric abnormalities even in younger patients with polycythaemia vera.

Keywords: polycythaemia vera; psychosis; schizophrenia

Introduction

Polycythaemia rubra vera can lead to neuropsychiatric disorders which, at least in part, may improve on venesection [1, 2]. In rare cases polycythaemia vera rubra is associated with psychotic episodes such as severe depression, mania and delirium [1, 3, 4–6]. However, there has been no report of a distinct paranoid hallucinatory psychosis in polycythaemia vera. The causal relationship between the psychiatric and the haematological condition is unclear. It has been suggested, however, that it may be explained on the basis of cerebrovascular mechanisms. We report a case of polycythaemia vera in a young male which occurred concurrently with the first onset of a schizophrenia-like psychosis.

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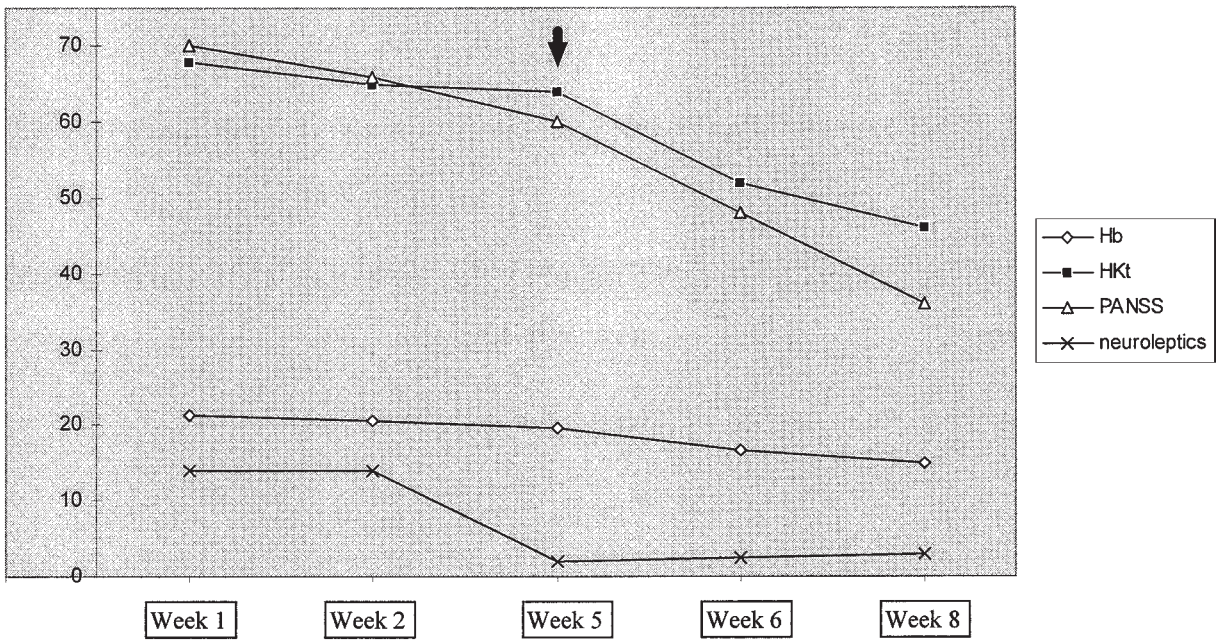
Case report

A 29-year-old university student with a tentative diagnosis of acute psychosis was referred to the psychiatric service by his general practitioner. On admission the patient complained of fatigue and irritability. He stated that this was owing to very exhausting experiences he had made during the last weeks. For example, he reported of having an intimate relationship with a room mate in the students' hostel although he actually had never spoken to this person. He was convinced of sharing a telepathic connection with this person whom he believed to be able to tune into his thoughts. He furthermore reported that other people were able to influence his thinking and his body-functions via mental power. Occasionally he felt that energy was being drawn out of his body which weakened him considerably. He had no convincing explanation for any of these occurrences but was afraid of being the victim of a strange plot. He stated that these events led him to a state of exhaustion and prevented him from continuing his studies. According to relatives and friends, although his premorbid personality was without abnormalities he had become socially withdrawn during the last months. He had no previous personal or family history of psychiatric disorders. There was no history of alcohol or drug abuse.

On admission, he was awake and fully orientated but his ability to concentrate was nonetheless impaired. Formal thoughts were long-winded and incoherent. He had delusions of reference and persecution and moreover reported of body hallucinations. His mood was slightly depressed. He scored 70 on the Negative and Positive Syndrome Scale (PANSS/7). General and neurological examinations revealed no abnormalities. An MRI scan, an EEG and results of a lumbar puncture were normal. However, his full blood count revealed considerable abnormalities (haemoglobin count: 21.4 g/dl; haematocrit: 68%; red blood cell count: 8.1×10^{12}/l). The level of lactat dehydrogenase was elevated to 568 U/l (normal range: 120–240 U/l).

Figure 1 Follow-up of haematological and psychopathological parameters. The arrow indicates initiation of venesection. Hb: haemoglobin (g/dl); HKt: haematokrit (%); PANSS (positive and negative syndrome scale, Kay et al. 1987, total score), neuroleptics (chlorpromazine equivalents/100).



Initially the diagnosis of acute paranoid-hallucinatory psychosis was made and a first episode of schizophrenia was suspected. The patient was treated with 16 mg benperidol a day. However, one week after admission the patient's psychopathological state only showed a slight improvement. Owing to the above mentioned laboratory abnormalities the patient was subsequently referred to the haematological department where he was diagnosed as suffering from polycythaemia vera and where a series of repeated venesections was initiated. At the same time, treatment with benperidol was stopped and since the patient complained of severe extrapyramidal side effects the neuroleptic medication was changed to 200 mg perazine a day. The drop in haematocrit level was accompanied by a rapid and marked improvement of the psychopathological state of the patient (fig. 1). A HMPAO-SPET scan which was performed when the haematocrit had dropped to 52% revealed no abnormalities. Subsequently, the patient's condition improved further and he was discharged in a stable physical and very good remitted psychopathological state.

Discussion

Polycythaemia vera is commonly accompanied by neurological disorders such as headache, dizziness, erythromyalgia, transient ischaemic attacks and strokes [8, 9]. Tinney et al. [10] reviewed 163 cases of polycythaemia vera and found 127 cases (78%) in which symptoms were referable to the nervous system. However, association of polycythaemia vera with psychiatric disorders has rarely been reported. Review of the literature revealed 12 cases of distinct psychiatric syndromes co-occurring with polycythaemia vera. These previous reports include patients suffering from depression with and without psychotic symptoms [3–6, 11–13], delirium and mania [1], confusion [6], obsessive compulsory disorder [14], olfactory, auditory and complex visual hallucinations [5, 14, 15], amnestic deficits [15–17] and twilight states [18, 19].

The clinical presentation of the patient described here suggested the presence of first-episode schizophrenia. Diagnosis of polycythaemia vera, however, led us to consider a possible link between the two disorders. The relationship between the haematological and the psychiatric condition may be explained in three ways: (1) the association may be coincidental; (2) the physical illness may have served as a non-specific stressor which may lead to an exacerbation of genuine schizophrenia in an individual at risk; (3) the haematological disorder may have altered the cerebral blood flow and subsequently the brain metabolism, resulting in an organic psychosis. The latter explanation is supported by the observation that the psychiatric condition improved markedly when venesection was introduced, although previous administration of a highly potent neuro-

leptic (benperidol) had not shown to be effective. Furthermore, the patient did not have any personal or family history of psychiatric disorders. It is well-known that an increase of haematocrit and blood viscosity in polycythaemia vera may lead to an alteration of cerebral blood flow resulting in impaired psychological functioning [2]. Moreover, the common observation of post-stroke depression has served as a heuristic model which illustrates the potential interaction between pathological changes in regional cerebral blood flow and psychiatric disorders [1, 5]. In order to support the hypothesis that blood flow changes are present in our patient, we performed a SPET scan which showed no significant regional cerebral blood flow changes. This negative result may, however, be explained by the fact that at the time of the scan the haematocrit was already in the normal range.

The described case is remarkable in two respects: (1) it is the first case of a distinct paranoid-hallucinatory syndrome associated with polycythaemia vera and (2) the affected patient is much younger than the previously described patients. Previous reports support the hypothesis that subclinical alterations of psychological functioning may be present in a high proportion of polycythaemia vera patients and may precede the development of haematological and neurological complications for many years [15, 18]. These patients may be individuals at risk in developing psychotic disorders and may even have a higher predisposition towards cerebrovascular accidents such as strokes and transient ischaemic attacks. This hypothesis should be addressed by systematic research since early recognition of subtle neuropsychiatric symptoms in these patients may allow early intervention to prevent potentially serious complications. The present report may therefore emphasise the need of a careful screening for psychiatric abnormalities even in younger patients with polycythaemia vera. Reversely, presence of polycythaemia vera should be considered in patients presenting with acute psychotic disorders, particularly since venesection may lead to a marked improvement of psychiatric symptoms.

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