CASE REPORTS

Primary polycythaemia presenting with chorea

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Summary

A case of primary polycythaemia presenting with severe chorea is reported and compared with previous cases. The aetiological factors of the chorea are discussed and the importance of early diagnosis stressed.

Introduction

Primary polycythaemia (polycythaemia rubra vera) commonly presents with neurological symptoms (Calabresi and Meyer, 1959; Lucas, 1912) but is only rarely associated with chorea. Edwards, Prosser and Wells (1975) found 23 previous cases of chorea associated with primary polycythaemia and one associated with secondary polycythaemia, they reported a further 2 of their own, one with primary and the other with secondary polycythaemia. Chorea was the presenting symptom in 11 of the patients with primary polycythaemia (4 males and 7 females) and there were 14 other reports of chorea developing in patients who were already known to have primary polycythaemia (3 males, 10 females, one sex not reported). These cases are listed in Table 1.

The preponderance of females was pointed out by Ashenhurst (1972); the reason for this is not clear. The mechanisms of chorea as a secondary phenomenon in polycythaemia and in some other system disorders such as hyperthyroidism, systemic lupus erythematosus and chorea associated with the contraceptive pill remain obscure. A case is now reported of primary polycythaemia presenting with chorea in a male patient.

Case report

A 59-year-old man gave a 9-week history of pro-

gressive slurring of speech and involuntary movements of his face and limbs. He stated that he had never felt well since he had stopped donating blood 5 years previously. There was no history of rheumatic fever and no family history of chorea. He was not on any medication before the onset of his symptoms.

Examination confirmed severe choreiform movements of the face and limbs, his speech was severely affected and he could only walk with support. His blood pressure was 170/110 mmHg. There were no other neurological or general abnormalities, in particular the spleen was not palpable.

Investigations. Hb 21.8 g/dl; PCV ratio 0.686; RBC $9.21 \times 10^{12}/l$; MCV 73 fl; MCH 23.7 pg; MCHC 31.8 g/dl; total WBC $19.1 \times 10^9/l$; neutrophils $16.43 \times 10^9/l$; lymphocytes $1.91 \times 10^9/l$; monocytes $0.76 \times 10^9/l$; platelets $183 \times 10^9/l$; ESR (Westergren) 0 mm/hr; red cell volume at 10 min 45 ml/kg at 40 min 44 ml/kg (normal 26-33); plasma volume 41.6 ml/kg (normal 40-50); blood volume 86 ml/kg (normal 60-80); LAP score 214 (normal 15-100); arterial blood gases, pH 7.402, P_{CO_2} 5.39 kPa (normal 4.5-6.1), base excess 1.3 mmol/l (normal ± 2.3), standard bicarbonate 24.2 mmol/l (normal 22-26), P_{O_2} 11.5 kPa (normal 12-15). A bone marrow examination showed pan-hypercellularity with absent iron stores. Urine microscopy, a chest X-ray, an intravenous urogram, thyroid function tests, antinuclear factor, a Wassermann reaction, and a CT brain scan were normal. An electroencephalogram showed a moderate non-specific abnormality with an excess of irregular theta activity.

Treatment and progress. The patient was treated initially with 3 venesections of 500 ml of blood each with volume replacement but there was only minimal

TABLE 1. Table of patients with chorea associated with polycythaemia as listed in the literature

Author	Date	Sex	Age	Primary (P) Secondary (S)	Presentation
Bardachzi	1909	F	59	P	chorea
Umney	1909	F	34	P	other
Naville & Brüttsch	1919	F	33	P	chorea
Pollock	1922	F	38	P	other
Schiff, Trelles & Ajuriaguerra	1936	F	78	P	other
Dameshek & Henstell	1940	M	39	P	chorea
	1940	F	49	P	other
Kotner & Tritt	1942	F	64	P	other
Reinhart <i>et al.</i>	1946	F	56	P	other
Harvier <i>et al.</i>	1950	M	49	P	other
Alajouanine <i>et al.</i>	1952	F	56	P	other
Trotsenburg & Koster	1954	F	63	P	chorea
Paliard, Croizat & Schott	1958	M	73	P	chorea
Calabresi & Meyer	1959	?	?	P	?
Bogolepov	1962	M	70	P	chorea
Bakke & Stavem	1963	F	74	P	chorea
Friedemann <i>et al.</i>	1965	F	62	P	other
Gauthier-Smith & Prankerd	1967	F	57	P	other
	1967	F	74	P	chorea
Bietti, Pompili & Sinibaldi	1968	F	75	P	other
Heathfield	1968	F	77	S	chorea
Nehilil <i>et al.</i>	1969	F	70	P	chorea
Sangster	1970	F	71	P	chorea
Ashenhurst	1972	M	68	P	chorea
Edwards, Prosser & Wells	1975	M	5	S	other
	1975	M	45	P	other
D. J. Thomas (personal communication)	1980	F	?	P	chorea
Present report	1980	M	59	P	chorea

improvement of his chorea (Fig. 1). He was then started on tetrabenazine (75 mg daily) and the abnormal movements diminished, but when the drug was discontinued the chorea returned. After a further 4 venesections the chorea had improved but

was still clinically obvious so tetrabenazine was restarted and the chorea abated. Two weeks later the haemoglobin was 12.1 g/dl, the tetrabenazine was again withdrawn but his chorea increased. He is now well maintained on tetrabenazine, 25 mg daily.

FIG. 1. The effect of venesection and tetrabenazine on the patient's clinical and haematological state. Clinical state is classified as follows: grade 1, mild chorea not noticed by patient; grade 2, abnormal movements noticed by patient; grade 3, moderate chorea with slurring of speech; grade 4, patient unable to walk unaided, and speech unintelligible.

Discussion

Chorea is a rare presenting feature or late complication of primary polycythaemia. The exact mechanism is unknown but it is assumed that one factor is reduced cerebral blood flow which is directly related to the haematocrit (Thomas *et al.*, 1977). In this and previous cases (Pollock, 1922; Ashenhurst, 1972) venesection alone was insufficient to stop the chorea, implying that permanent damage had occurred in the central nervous system. A detailed neuropathological report in such a case (Kotner and Tritt, 1942) showed widespread venous congestion with perivenous demyelination, infarction and haemorrhages in the cortex, subcortical white matter, basal ganglia and choroid plexus, but

their findings were insufficient to account for the chorea as similar changes have been reported in cases of polycythaemia without chorea by Hutchinson and Miller (1906). The role of platelet function has been discussed by Rosenthal (1949), Shield and Pearn (1969), and Edwards *et al.* (1974). Aminoff *et al.* (1974) have found abnormal dopamine metabolism in platelets from patients with Huntington's chorea. There is no clear correlation between the platelet count and the chorea in the present case.

The present authors have not been able to find any single factor which would explain why some patients with polycythaemia develop chorea, it may be that some individuals have an underlying predisposition

to chorea which can be unmasked by polycythaemia or other conditions such as hyperthyroidism or systemic lupus erythematosus.

The diagnosis of polycythaemia should always be considered in patients with obscure neurological symptoms including chorea as delay in diagnosis can result in serious neurological deficit and intellectual impairment which persists even when the polycythaemia is corrected.

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