THE NEUROLOGIC SPECTRUM OF POLYCYTHEMIA VERA: A COMPREHENSIVE LITERATURE REVIEW

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Introduction

- Polycythemia vera (PV), a myeloproliferative neoplasm driven mainly by the JAK2 V617F mutation, is associated with thrombotic and hyperviscosity-related complications.
- Neurological manifestations, including chorea, cognitive disturbances, and peripheral neuropathy, although uncommon, are clinically significant and often under-recognized.
- Literature suggests chorea in PV may result from hyperviscosity-induced basal ganglia dysfunction, altered dopaminergic signaling, or venous congestion.
- While isolated cases report symptom improvement with phlebotomy or cytoreductive therapy, comprehensive analysis remains limited.
- This review summarizes neurological manifestations of PV to improve recognition, inform management, and identify research gaps.

Aim

- Systematically review existing literature on the neurological manifestations of PV, focusing on movement disorders such as chorea, with the aim to elucidate underlying pathophysiological mechanisms, clinical presentations, and therapeutic outcomes.

Methods

- A comprehensive narrative review of peer-reviewed case reports and clinical studies detailing PV neurological manifestations, emphasizing movement disorders such as chorea.
- We conducted a literature review across international medical databases, including PubMed and NEJM, for previously reported cases and studies.
- This review synthesizes data from adult patients with PV presenting neurological manifestations, particularly chorea, reported in case studies and clinical series.
- Included patients span a wide age range, with a predominance of elderly females harboring the JAK2 V617F mutation.
- Interventions reported in reviewed cases include serial phlebotomies, cytoreductive therapy, and symptomatic treatment with neuroleptics or benzodiazepines.
- Improvement or resolution of choreiform movements correlated with normalization of hematological parameters.
- Secondary outcomes include prevention of thromboembolic complications.

Results

- Neurological manifestations of PV are diverse, including chorea, headaches, dizziness, thrombotic events, and cognitive changes.
- Chorea is rare (~1–2.5%), predominantly in elderly females with JAK2 mutation.
- Most literature reports improvement in chorea following hematocrit normalization by phlebotomy and cytoreductive therapy.
- Early diagnosis and treatment reduce morbidity and mortality.

Discussion

- Neurological manifestations of PV, including chorea, are rare but clinically significant.
- Chorea typically affects elderly females with JAK2 mutations and often improves with hematocrit control.
- Magnetic resonance imaging is usually unremarkable; Magnetic resonance venography may help exclude cerebral venous thrombosis.
- Early recognition and treatment are crucial to prevent thrombotic and neurological complications, highlighting the need for greater awareness and research.

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