A CASE OF ACUTE CHOREO-ATHETOSIS IN A PATIENT WITH POLYCYTHEMIA VERAE

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INTRODUCTION

Polycythemia vera is a sporadic myeloproliferative disorder characterised by increased red blood cell mass. Associated thrombosis, haemorrhaging, and hyperviscosity commonly result in neurological manifestations.

NEUROLOGICAL COMPLICATIONS IN PATIENTS WITH POLYCYTHEMIA

- Headache/migraine: 41%
- Dizziness or vertigo: 30%
- Paresthesias: 13%
- Visual disturbances: 11%
- Stroke: 9%
- Tinnitus: 3%

Mechanisms for basal ganglia (and their thalamo-cortical connections) dysfunction have not been clearly elucidated yet.

PROPOSED MECHANISMS FOR BASAL GANGLIA DYSFUNCTION

- Hypoperfusion due to venous stasis
- Receptor hypersensitivity in a setting of reduced catecholamine levels
- Altered dopamine metabolism

SPECT images of the brain. (a) before treatment: there is reduced TRODAT-1 uptake in the bilateral basal ganglia, especially on the left side. (b) after treatment: there is increased and more symmetrical dopamine transporter in the bilateral striatum compared to 'before treatment' image.

Values of binding potentials for striatum, putamen and caudate before and after treatment:

<table>
<thead>
<tr>
<th>Region</th>
<th>Before Treatment</th>
<th>After Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right striatum</td>
<td>0.45</td>
<td>0.66</td>
</tr>
<tr>
<td>Left striatum</td>
<td>0.21</td>
<td>0.71</td>
</tr>
<tr>
<td>Right putamen</td>
<td>0.36</td>
<td>0.60</td>
</tr>
<tr>
<td>Left putamen</td>
<td>0.23</td>
<td>0.66</td>
</tr>
<tr>
<td>Right caudate</td>
<td>0.78</td>
<td>0.93</td>
</tr>
<tr>
<td>Left caudate</td>
<td>0.20</td>
<td>0.85</td>
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</tbody>
</table>

Administration of neuroleptics may suppress abnormal choreiform movement; however, effective management of polycythemia vera requires serial venesections in conjunction with chemotherapy.

AIM OF THIS CASE-REPORT

We present a case of a new onset chorea in a patient with a diagnosis of polycythemia vera, to confirm the link between hematological pathology and movement disorders.

CASE-REPORT

A 77 year old woman was admitted to our department for investigation about sudden onset involuntary movements involving face, trunk, and limbs.

- No family history of chorea or dementia and no use of chorea-inducing drugs.
- Arterial hypertension, paroxysmal atrial fibrillation, aortic valve transplant in treatment with oral anticoagulants, polycythemia vera in cyclic treatment with hydroxyurea.
- In full-being, a sensation of swollen tongue, dry mouth, and sore throat for about a month. Unintentional movements of extension and retraction of the tongue. Involuntary movements in the lower limbs first, then involving the trunk and the upper limbs.
- At the admission, a hematocrit value of 54.8% (reference values 34.8-45) was found.
- Prompt treatment with Haloperidol resulted in a reduction of involuntary movements, and the restoration of hydroxyurea and a single phlebotomy (on haematological advice) gave further improvement in symptoms.
- At discharge, her hematocrit value was 46.6%, and involuntary movements ceased.

LEARNING POINTS

- The onset of a choreic syndrome in patients with polycythemia can alert us about deterioration in the hemoglobin values
- Polycythemia must be considered because this diagnosis leads to effective treatment and prevention of serious complications – deep vein thrombosis, pulmonary embolism, stroke

CONCLUSIONS

The clinician should always consider the possibility of a hematological etiology in a patient with sudden-onset involuntary movements and a history of hematological diseases.

REFERENCES