# 真性紅血球增多症所致舞蹈症
# Polycythemia chorea: a case report

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***Background*** Patients with polycythemia vera (PV) may manifest with neurological disorders such as chorea (0.5-5%). We investigated the brain metabolism and dopamine system of a genetically confirmed PV patient using 2-[$^{18}$F]fluoro-2-deoxy-D-glucose positron emission tomography (FDG PET) scan and Tc-99m-labeled tropanes dopamine transporter (Tc-99m-TRODAT-1) single photon emission computed tomography (SPECT) imaging.

***Case Report*** A 70-year-old woman developed acute onset of choreiform movement of left limbs and face 6 months prior to admission. Along with the quench of the left side symptoms, the involuntary movement of the right limbs appeared 4 months later. There was no family history of chorea. Physical examination showed erythromelalgia of the hands with mild clubbing of fingers (Fig. 1). Neurological examination disclosed choreiform movements of the orofaciolingual regions and the limbs, with the right side predominantly involved. All four limbs showed hypotonia with diminution of tendon reflexes and flexor plantar responses. She had no bradykinesia, rigidity or rest tremor.

A complete blood count revealed elevated values of red cell count (5.91 million/ul; range 4–4.5), hemoglobin concentration (16.8 gm/dl; range 12–16), hematocrit (51.7%; range 35–48%) and platelet count (769×10$^3$/ul; range 130–400×10$^3$). Genetic analysis showed heterozygous mutation of JAK2 (codon 617, from V to F). The laboratory investigations confirmed the diagnosis of PV according to the World Health Organization (WHO) criteria. Magnetic resonance imaging of the brain was unremarkable.

In Tc-99m-TRODAT-1-SPECT, reduction of TRODAT-1 uptake was found in bilateral basal ganglia, especially in the left side (Fig. 2a). A significant increase in FDG metabolism on the PET image was noted over the right dorsolateral prefrontal cortex and left insular cortex (Fig. 3).

The patient received consecutive phlebotomies (250ml each) with one-month interval. Normalization of hemoglobin and hematocrit values along with the amelioration of chorea and erythromelalgia appeared after the fifth phlebotomy.

A follow-up TRODAT-1 SPECT of the brain 10 months after the initial image showed an increase in dopamine transporter uptake compared to the previous images and the images were more symmetric at bilateral striatum (Fig. 2b). A follow-up FDG PET brain examination at the same interval also showed normalization of brain metabolism. The beneficial effect of scheduled phlebotomy to her choreiform movements was maintained in her regular follow-up.

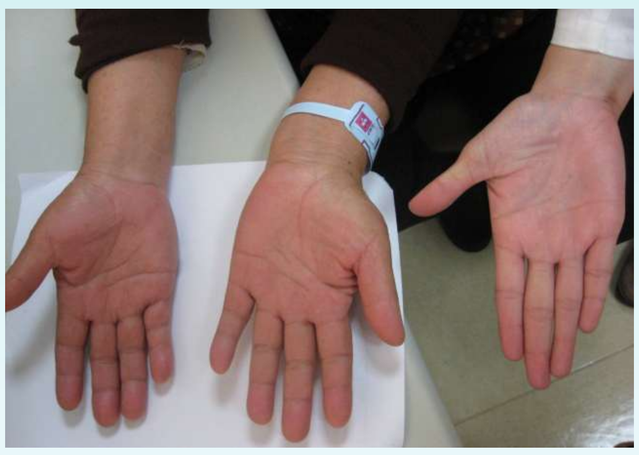

**Figure 1. Erythromelalgia and mild clubbing of patient's hand (left).**
Left: the patient's hands
Right: the normal person's right hand

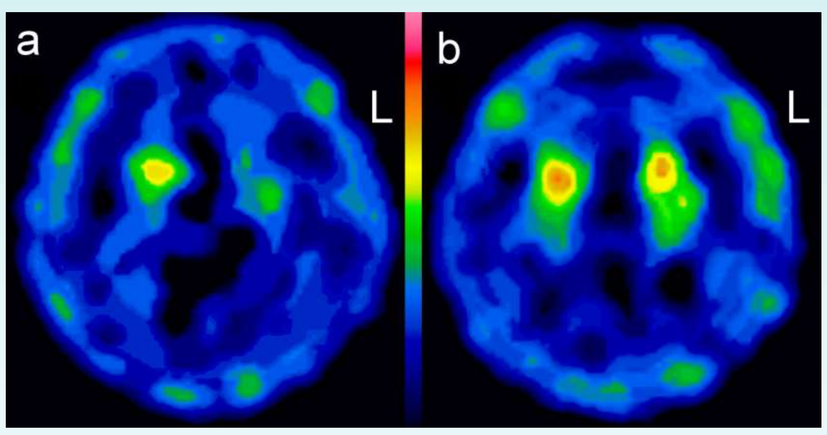


**Figure 2. Tc-99m-TRODAT-1 SPECT of the brain.**
**a.** Before treatment, reduction of TRODAT-1 uptake is found in bilateral basal ganglia, especially in the left side.
**b.** After treatment, increase in dopamine transporter uptake is observed and there is more symmetric uptake of dopamine transporter in bilateral striatum.

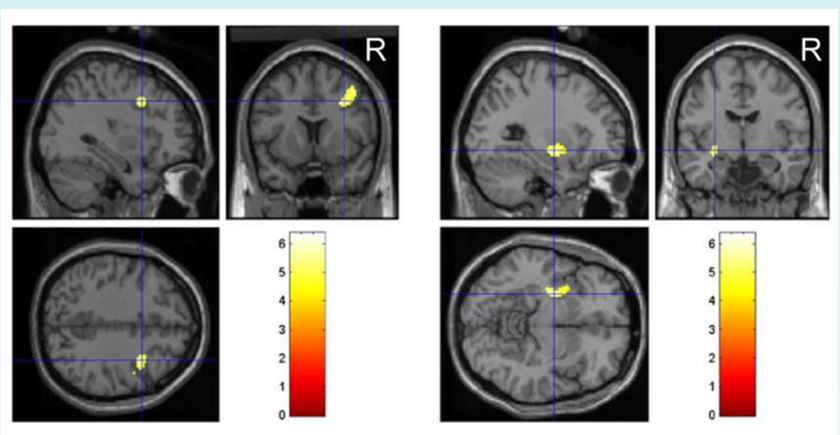


**Figure 3. FDG PET images transferred to statistical parametric mapping.**
One hypermetabolic cluster is shown in right dorsolateral prefrontal cortex with 194 contiguous suprathreshold voxels and the other one is in left insular cortex with 169 contiguous suprathreshold voxels.

***Conclusion*** The generation of polycythemia chorea could be relevant to the reversible alteration of the basal ganglia-cortical metabolism and dopaminergic functional perturbation.