IDIOPATHIC BASAL GANGLIA CALCIFICATION WITH ALTERED IOFLUPANE SPECT AND GOOD RESPONSE TO L-DOPA TREATMENT: A CASE REPORT

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Background

Basal ganglia calcification is histopathologically characterized by massive calcification, that most commonly affects the basal ganglia (BG), but also cerebellum, thalamus, and brainstem.[1] It may be a secondary manifestation of any of more than 50 medical conditions, such as metabolic, environmental, autoimmune, mitochondrial, sporadic, or inherited genetic disorders. However, similar patterns of brain calcifications may often be found in idiopathic cases with normal biochemical and endocrinology examination findings (calcium, phosphorus, and parathyroid hormone levels). Therefore an eponym idiopathic basal ganglia calcifications (IBGC), also known as Fahr's disease (FD), has been coined for this clinically heterogeneous disorder. CT Scan reveals cerebral calcifications, mostly localized in lenticular nucleus, putamen, thalami, caudate and dentate nuclei.[2] Single photon emission CT (SPECT) with Ioflupane plays a crucial role in detecting presynaptic dopaminergic abnormalities in parkinsonian syndromes. In IBGC, Generally, in the absence of nigrostriatal presynaptic neurodegeneration, Ioflupane SPECT is expected to be normal. Response of parkinsonian features to L-Dopa therapy is poor, indeed.

Methods

- 60 year old man, with unremarkable family history, presented motor awkwardness on both legs associated with "freezing" of gait, rigidity and mild body bradykinesia.

- CT scan documented calcifications of basal ganglia and cerebellar hemispheres (Figure 1); DAT SPECT showed marked impairment in the bilateral caudate and putamen nuclei (Figure 2).

- Normal Blood and urinary calcium levels, as well as blood PTH, ESR and CBC levels.

- Normal Cerebrospinal fluid analysis.

- Neuropsycological assessment: MMSE 26/30, CDT 9.5/10, FAB 11/18, mild cognitive dysfunction most of all involving frontal- and temporal-lobe abnormalities.

Results

- A diagnosis of "Idiopathic Basal Ganglia Calcifications" was formulated.

- Dopaminergic therapy with L-Dopa up to 1000 mg per day was attempted, with little subjective benefit.

- Positive Acute L-Dopa challenge (UPRDS change 28% from overnight OFF state to ON state).

- One year later he developed dyskinesias and stereotyped movements to hand mainly on the left.

Discussion

We describe a man with sporadic basal ganglia calcifications presenting with:
- symptoms resembling vascular parkinsonsim;
- positive Iofluopane Spect like in primary (degenerative) parkinsonism;
- a positive response to L-Dopa treatment.

As magnetic resonance imaging has poor sensitivity for brain calcifications, in the absence of a CT scan this clinical picture could have been misunderstood for a long time.

Nigrostriatal function has been described in only a few of these cases, providing conflicting results:
- normal in two subjects with prominent cognitive impairment;
- Abnormal in two patients with predominant extrapyramidal symptoms[3];
- Abnormal in one patient with prevalent cerebellar signs[4];

Considering the lack of nigral neuronal loss in cerebral calcifications, we can explain the abnormal DAT binding as secondary to a local damage by the abnormal calcifications[5]. Still, the mechanism underlying the responsiveness to L-Dopa in our patient remains unknown.

Conclusions

Idiopathic basal ganglia calcifications can be associated with a wide range of clinical and radiological findings and must always be considered in differential diagnosis of extrapyramidal syndromes.

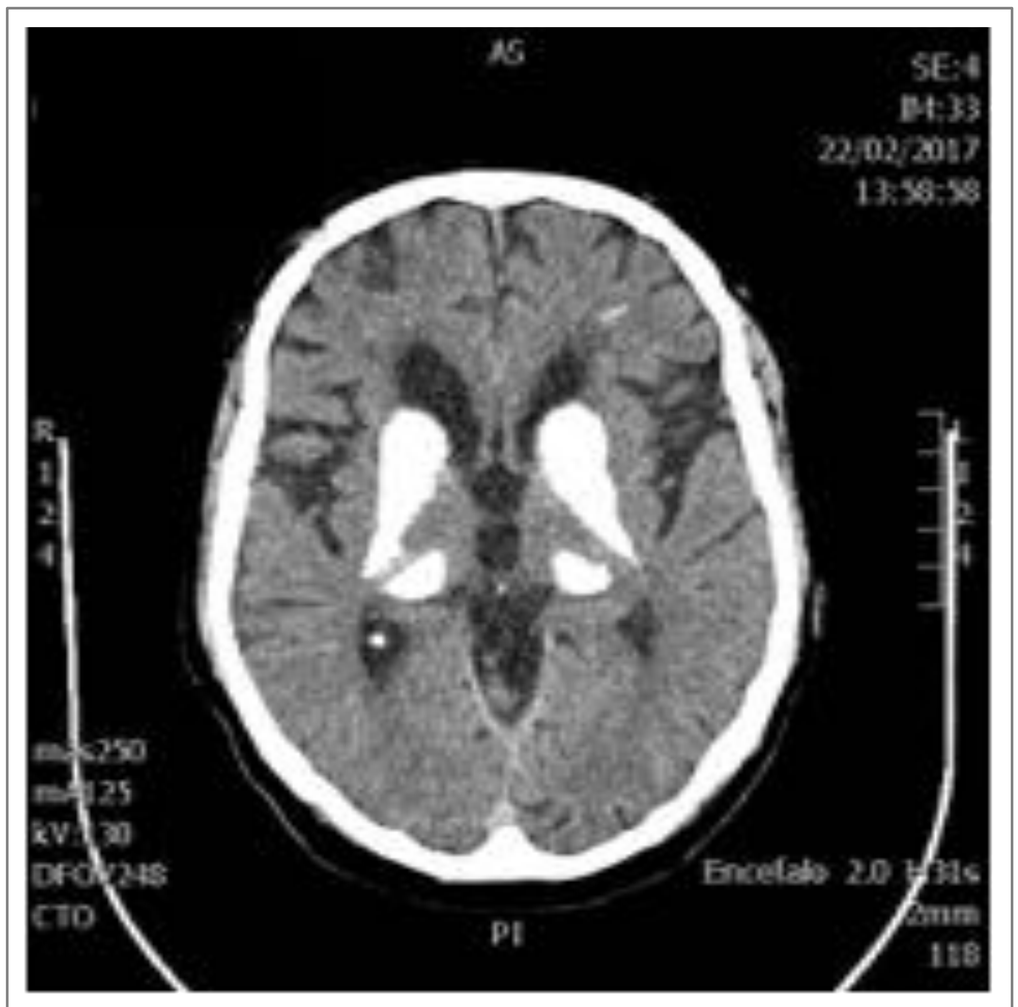

Figure 1: Patient's CT scan showing extensive calcifications involving both cerebellar hemispheres

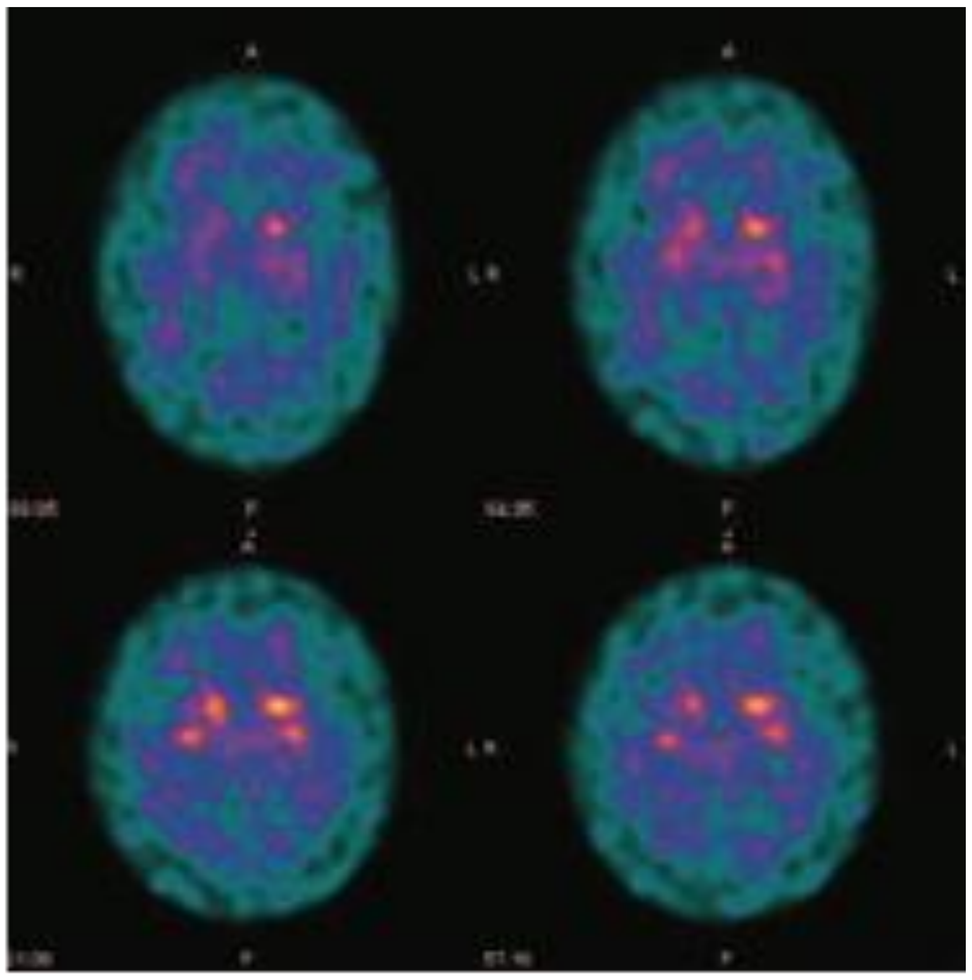

Figure 2: Ioflupane SPECT showing bilateral marked compromise of dopaminergic nigrostriatal pathway at caudate and putamen level

References

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