Fahr's syndrome presenting with neuropsychiatric symptoms: a case report

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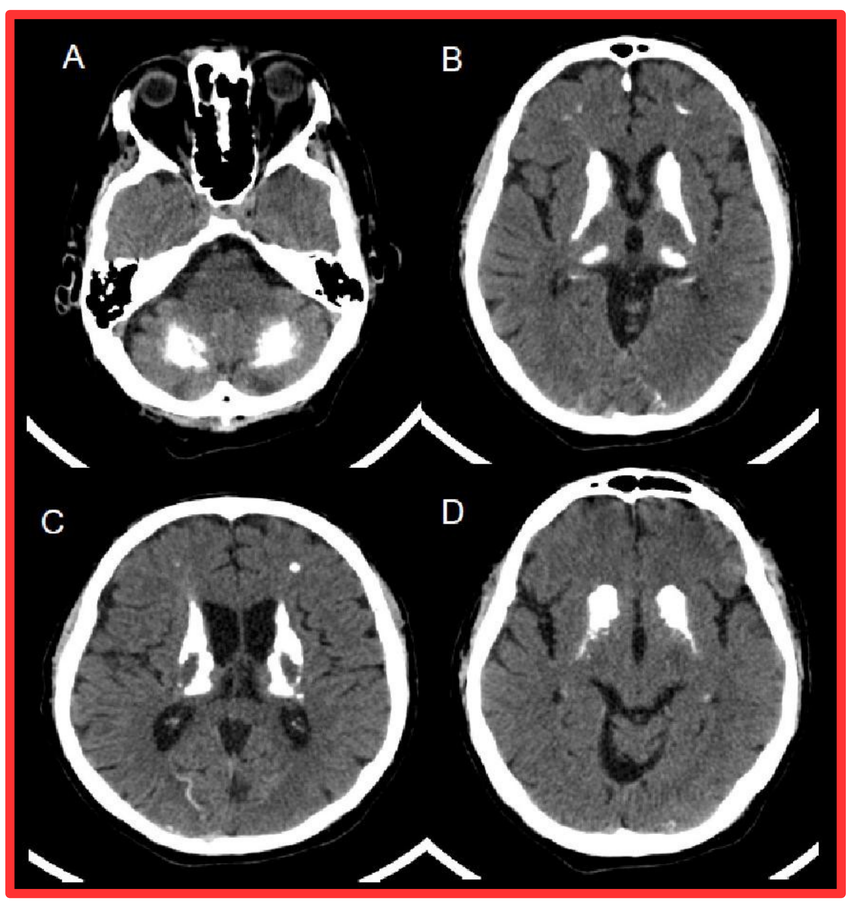
Background

Fahr's disease is characterised by idiopathic calcification of the basal ganglia, nuclei dentate of the cerebellum and centrum semiovale. The disease usually manifests itself in the 3 to 5 decade of life, but may appear in childhood or later in life. Neuropsychiatric symptoms can be the first or the most prominent manifestations ranging from mild difficulty in concentration, memory changes in personality, behavior, frank psychosis and dementia

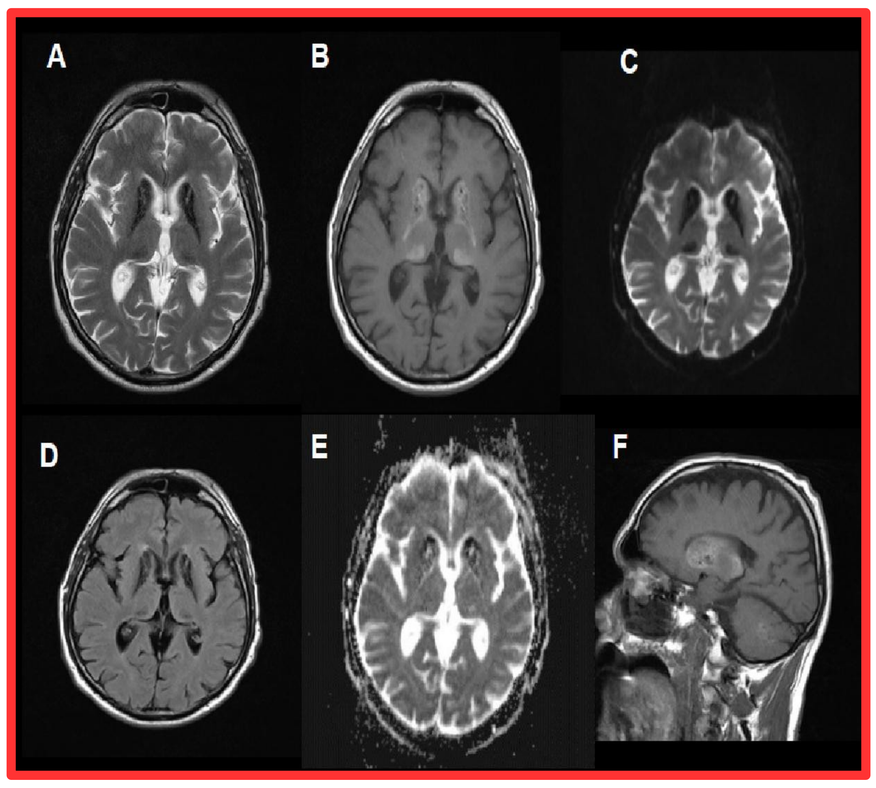
Case Report

We have recently seen a 55-year-old man who was referred with progressive mental deterioration for the previous year.
The first symptoms were those of a dysexecutive syndrome with alterations in abstract reasoning, calculation, sequential tasks and apathy.



Three months later, memory loss and depressive mood began. Patient did not report any depressive features, delusions, or hallucinations.
Finally she was unable to perform daily living activities, with decreased verbal fluency, apathy and inability to make decisions.
His family history was not significant.
On neurological examination we did not observe signs of parkinsonism or abnormal movements.
The score of the Mini Mental State Examination was 15/35 (Italian version); it was not possible to proceed to the administration of other neuropsychological tests or MRI for the lack of cooperation of the patient despite the ongoing therapy.
In view of low parathyroid level, her ionic calcium levels were done which came out to be within normal range (1.12 mmol/l).
Standard blood tests were normal including calcium and phosphorus, and so was the hormonal profile including thyroid hormones and parathormone.
Serologic tests for syphilis and HIV were negative.
Computed tomography showed extensive bilateral calcifications in the dentate nuclei of the cerebellum, basal ganglia and centrum semiovale.
Based on above history and investigations, diagnosis of Fahr's syndrome (bilateral BGC, dysarthria and neuropsychiatric symptoms) was kept.

Patient showed partial improvement in behavioral symptoms; hence, risperidone was added in a dose of 3 mg/daily. Patient responded (with risperidone, lorazepam, memantine and treatment for hypo-parathyroidism) in next 40 days he improved in psychiatric symptoms: depression, hallucinations, apathy.



Conclusion

This case further emphasizes the importance of the role of neuroimaging and the search for disrupted phosphocalcic metabolism and abnormal parathyroid levels in patients with atypical psychotic symptoms.
Judicial use of neuroleptic drugs can definitely help in controlling the behavioral symptoms in Fahr's syndrome.

References

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