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Fahr's Disease with Secondary Hyperparathyroidism

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Abstract

Fahr's syndrome refers to a rare syndrome which is characterized by symmetrical and bilateral intracranial calcification.[1,2]

Here we present a case of a 48 year old male patient , who presented to our outpatient clinic with history of tenderness in the right hip since 3 days following accidental fall at home and history of recurrent falls, walking like a drunken person ,inappropriate laughter and crying, difficulty in speaking and dystonic posturing , dificulty in eating and deglutition since 02 years .

NCCT Head revealed bilateral, symmetrical, large areas of calcification over the basal ganglia, the thalamus, the cerebellum , the pons and the occipital lobe and the diagnosis of ***Fahr's disease*** was done and other secondary causes of calcification were ruled out .

Laboratory investigations revealed vitamin D insufficiency and ***secondary hyperparathyroidism*** leading to susceptibility for bone fractures.

Thus**,** ***Fahr's disease*** patient is a highly susceptible patient, which requires a multidisciplinary approach in order to prevent postural instability and nutritional insufficiencies, thus reducing the risk of complications like fractures.

Keywords: Fahr's disease, secondary hyperparathyroidism .

Introduction

It was first described by a German Neurologist Karl Theodor Fahr in 1930 [1,2]

Fahr 's Disease is a rare , genetically dominant , inherited neurological disorder characterized by abnormal deposition of calcium in areas of the brain that control movement , including the basal ganglia and cerebral cortex . Symptoms of the disorder may include deterioration of motor function , dementia , seizures , headache , dysarthria , spasticity and spastic paralysis , eye impairment and athetosis . Fahr disease can also include symptoms characteristic of Parkinsons disease . Age of onset is typically in the 40s or 50s , although it can occur at any time in childhood or adolescence. Histologically, these deposits which contain proteins and polysaccharides, are found in the perivascular space and in the media layer of the small vessels. The pathogenesis is not known, but it may be secondary to the impairment of the blood brain barrier or to a neuronal calcium phosphoric metabolism disorder [2-5].

We wish to highlight that , as there is no specific treatment for Fahr disease , it is very important for the treating clinician to take care of the nutritional insufficiencies which occurs along the disease course to prevent the complications like fractures and improve the quality of life.

Case Discussion

A 43 year old male patient, presented to our outpatient clinic with history of severe tenderness in the right hip following accidental fall at home since 3 days difficulty in walking in the form of walking like a drunken person ,inappropriate laughter and crying, difficulty in speaking and dystonic posturing , dificulty in eating and deglutition and aggressive behavior since 03 years .

On examination, BP: 150/90 mmHg , PR: 100 / min , RR: 16 cycles / min and local tenderness over the right hip present .

He was moderately built and nourished . He was a non smoker, non alcoholic and attenders denied of any high risk behaviour.

On CNS examination, patient was not oriented to time, place and person. Obvious extrapyramidal movement like dystonia, oral dyskinesia present, rigidity, violent movement of hands and legs.

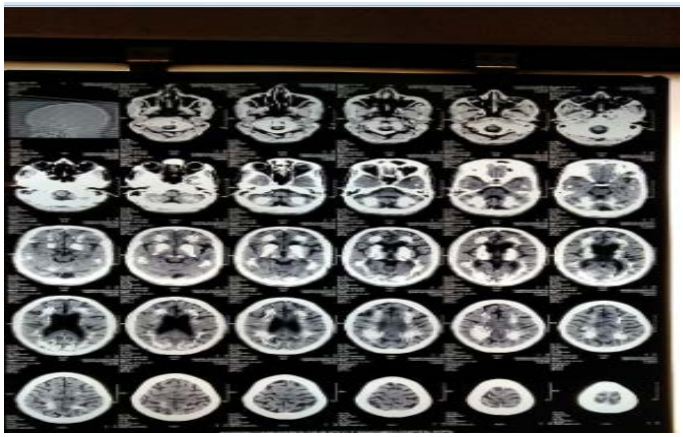

Figure 1: CT scan of the brain revealed bilateral, symmetrical, large areas of calcification over the basal ganglia, the thalamus, the cerebellum and the Pons and the occipital lobe, which were suggestive of Fahr's disease (fig 1).

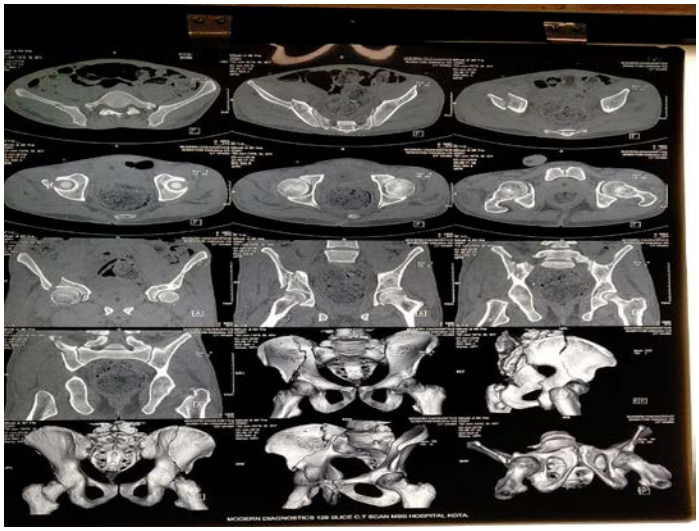

Figure 2: CT scan of the pelvis showed multiple displaced fracture of the right iliac bone with no other fracture demonstrated in rest of the visualized bone(fig 2).

Serum lactate was negative and hence ruled out mitochondrial disease. The TORCH analysis by ELISA , VDRL and HIV tests gave negative results. His laboratory investigations revealed a normal haemogram and his renal parameters and liver function test were normal .To rule out calcium phosphorus metabolism abnormalities investigations were done and are as follows(table 2) and was consistent with secondary hyperparathyroidism due to vitamin D insufficiency .

Table 1.

Laboratory Values	
1.Serum calcium 1. Ionised calcium	7.2 mg/dl(8.7-10.2) 4.55.3
2. Serum phosphorus	4.8 mg/dl(2.5-4.5)
3. Serum PTH	87.90 pg/Ml(08-51 pg/ml)
4. Serum 25-OH-Vitamin D3	22.50 ng/mL(30-100 sufficiency)
5. Serum magnesium	1.96 mg/dl(1.5-2.3)
6. Urinary calcium and phosphorus and USG parathyroid normal	

Fracture was managed symptomatically with traction , complete bed rest , analgesics and intravenous fluids.Management was difficult due to severe extrapyramidal symptoms like rigidity, dystonia and postural instability . Our patient was treated with mega doses of vitamin D3 and calcium supplements and was discharged on levodopa , carbimazapine and oral calcium and vitamin D3 and is under regular follow up.

There is no cure for Fahr's syndrome nor is there a standard course of treatment. In this context, at present time management and treatment strategies mainly focus on symptomatic relief and to cure the reversible factors.

Conclusion

Fahr disease with secondary hyperparathyroidism and vitamin D insufficiency is a rare manifestation. Here we like to highlight the importance of calcium and vitamin D3 supplements to be given to a patient of Fahr disease as they are highly susceptible to fractures due to recurrent falls and abnormal mineral metabolism. Fahr's syndrome patient is a "weak" patient, which requires a multidisciplinary approach in order to prevent the mobility reduction, to improve the condition of postural instability, thus reducing the risk of fractures using preventive measures.

References

1. Murat Gülsün , Ali FuatBaykız , SerdarKabata , Hasan Belli. Fahr's Syndrome - Three cases presenting with psychiatric signs. *Eur J Gen Med.* 2006;3(1):35-40.

2. Kotan D, Aygul R. Familial Fahr's disease in a Turkish family. *South Med J*. 2009;102(1):85-86.

3.Billard C, Dulac O, Bouloche J, Echenne B, Lebon P, Motte J. Encephalopathy with calcifications of the basal ganglia in children. A reappraisal of Fahr's syndrome with respect to 14 new cases. *Neuropediatrics*.1989;20(1):12-19.

4. Malik R, Pandya VK, Naik D. Fahr's disease: A rare neurodegenerative disorder. *Indian J Radiol Imaging.* 2004;14:383-84.

5. Kotan D, Aygul R. Familial Fahr's disease in a Turkish family. South Med J. 2009;102(1):85-86.