Journal of Neurology

Open access

Case Report

Fahr's Syndrome with Hypoparathyroidism Presenting with Acute Ischemic Stroke

Krishnendu Choudhury*, Biswajit Paul, Koushik Ray, and Shibendu Ghosh

Department of Medicine and Neurology, Jagannath Gupta Institute of Medical Science, Budge Budge, Kolkata

***Correspondence**: Department of Medicine and Neurology, Jagannath Gupta Institute of Medical Science, Budge Budge, Kolkata

Abstract

Introduction: This study is aimed at an institutional review of Vestibular Schwannomas (VS) treatment with the first Gamma Knife machine in the Kingdom in order to describe expected goals of outcomes.
Methods. The clinical information of VS patients from the prospective database of Gamma knife Radiosurgery (perfexion) were reviewed and analyzed through SPSS v22 for outcomes. A total of 47 consecutive patients were reviewed and 35 got selected for final review considering at least one or more year of post radiosurgery follow up. All the patients were treated between the year 2013 and 2019 completing machine's first Cobalt sources half-life
Results. A total of 35 patients were reviewed. The cohort consisted of 13 males and 22 females with a median age of53 years (23-80 years). 22 patients had no comorbidity and the rest had at least one including DM, HTN or both. 7 patients had post-operative Radiosurgery to their residual disease and 28 were treated with Radiosurgery alone. 34 patients presented with symptomatic disease. Mean follow up duration for < 2yrs group: 13.8 months, while > 2 yrs: months. Hearing deficiency; Sever SN hearing loss: 15 patients, Mild to moderate: 15 and normal hearing: 5 patients. Tinnitus: 20 patients and Vestibular dysfunction: 23 patients. Duration of follow up time (more than 2 years) isthe single predetermining factor associated with improved outcomes (P<0.001
Conclusion. Gamma knife Radiosurgery is an effective treatment modality with expected treatment goals to achieve requires at least a period of 2 years to become clinically appreciable

Keywords: Fahr's syndrome, Hypoparathyroidism, Acute Ischemic Stroke

Received date: Feb 01, 2024; **Accepted date:** Feb 11, 2024; **Published date:** Feb 28, 2024

Citation: Choudhury K, Paul B, Roy K, Ghosh S."Fahr's Syndrome with Hypoparathyroidism Presenting with Acute Ische mic Stroke. J Neu 2,1.

INTRODUCTION

Idiopathic basal ganglia calcification, also known as Fahr's disease, is a rare, genetically transmitted autosomal dominant, neurological disorder characterized by abnormal deposits of calcium in areas of the brain that control movement. It can also occur independently as dentato-thalamo- lenticular calcification as a sporadic disorder in elderly people. Manifestations such as movement disorders like chorea, dystonia or spasticity, paratonic rigidity are present in minority of patients and most of the patients are asymptomatic. Some of them are reported to have associated hypo- parathyroidism with decreased vitamin D and parathormone level in serum.

CT scan and MRI brain can reveal calcifications primarily in the basal ganglia, lenticular nucleus and dentate nuclei of cerebellum and in other areas such as the thalamus and cerebral cortex. There is no reliable correlation between age, extent of calcium deposits in the brain, and development of neurological features.

Dystonia and paratonia also manifest as abnormalities of muscle tone, but arise more due to the network dysfunction between the basal ganglia and the thalamo-cerebello-cortical connections.

Ischemic cerebrovascular disease may be associated, and incidence of ischemic or hemorrhagic stroke in Fahr's syndrome has

been rarely reported. We report here an elderly woman who had presented with acute ischemic stroke and found to have Fahr's syndrome in brain imaging. She was also detected to have primary hypoparathyroidism with decreased serum vitamin D3 and parathormone deficiency.

CASE REPORT

A 65 year old female admitted with sudden onset slurring of speech and weakness of left side of face and left half of her body. There was no fever, diarrhea, convulsion or loss of consciousness. She did not have any history of chronic hypertension, heart disease, kidney disease or diabetes mellitus. There was no history of cognitive impairment or abnormal movement in her or any family members. There was no history of neck radiation or any thyroid disease or operation of neck or thyroid gland.

On examination she was conscious, orientated, her pulse, temperature and respiration were within normal range. Blood pressure was 170/100 mm Hg. Her mental status did not show any significant impairment; Speech was dysarthric with normal comprehension and speech output. Cranial nerves examination showed visual acuity 6/18 with immature cataract in both eyes and left facial palsy (lower face) upper motor neuron type; Motor system examination revealed decreased power (grade1/5) in left upper and lower limbs with hypertonia, hyperreflexia and Babinski sign. Her right upper and lower limbs power was slightly reduced with paratonic rigidity in right upper limb. There was sensory extinction in her left side. Cerebellar signs were absent on right side. There was no carotid bruit. Investigations: Routine blood examinations CBC ESR CRP were normal. CBG and RBS were normal, PPBS and FBS, HbA1C were normal. CT scan did not show any hypodensity. There was calcification in the putamen, globus pallidus, thalamus and dentate nucleus of cerebellum bilaterally (Figures 1 and 2). MRI brain revealed diffusion restricted lesion (Figure 3) in right parietal, frontal and external capsule regions (Middle Cerebral Artery territory infarcts) with corresponding ADC dark mapping (Figure 4). There was Gradient Echo blooming in bilateral thalamus, lenticular nucleus and dentate nucleus of cerebellum indicating calcification (Figures 5 and 6).

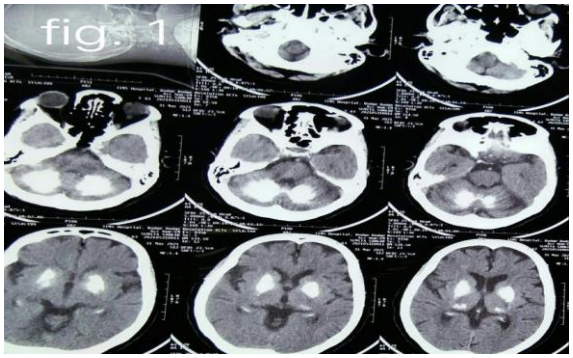

Figure 1. Calcification in the putamen, globus pallidus, thalamus.

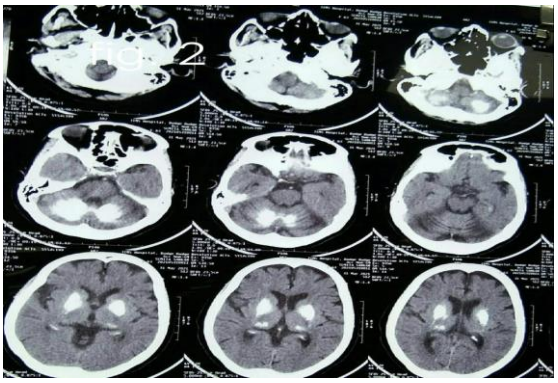

Figure 2. Dentate nucleus of cerebellum bilaterally.

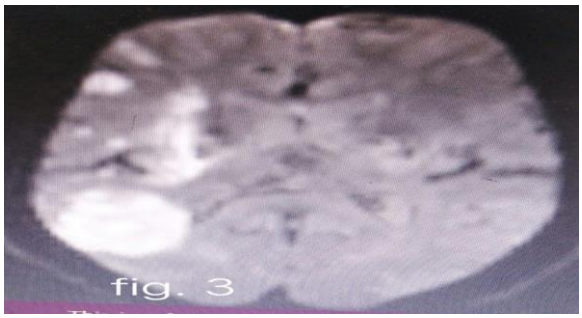

Figure 3. MRI brain revealed diffusion restricted lesion.

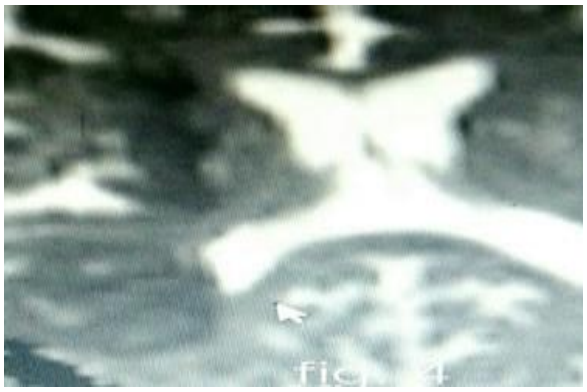

Figure 4. Frontal and external capsule regions (Middle Cerebral Artery territory infarcts) with corresponding ADC dark mapping.

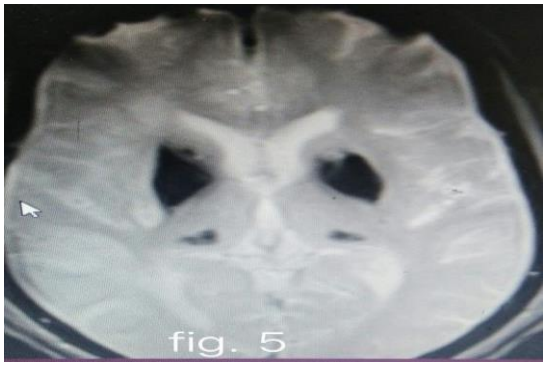

Figure 5. There was Gradient Echo blooming in bilateral thalamus, lenticular nucleus.

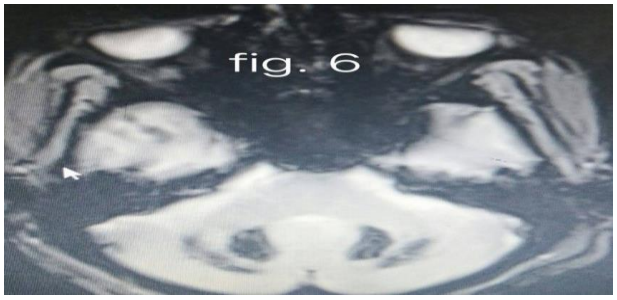

Figure 6. Dentate nucleus of cerebellum indicating calcification.

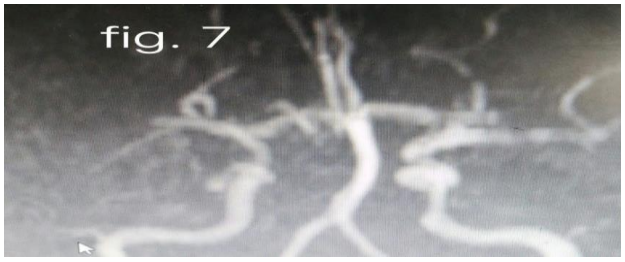

Figure 7. MR angio showed occlusion in distal right MCA with decreased flow along M2, M3 branches.

Figure 8. MR angio showed occlusion in distal right MCA with decreased flow along M2, M3 branches

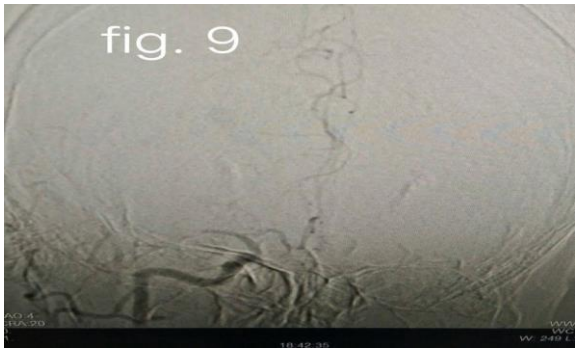

Figure 9. DSA showed distal MCA occlusion with collaterals from ACA-PCA.

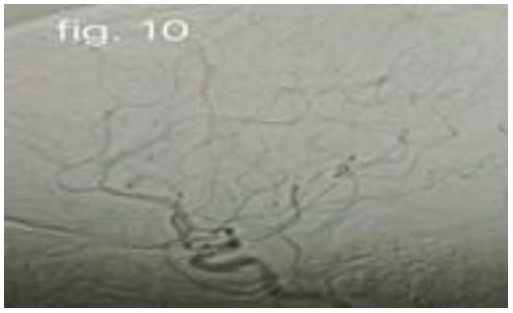

Figure 10. Calcified plaque in right ICA origin with 50% luminal narrowing.

DISCUSSION

This elderly woman had a recent acute ischemic stroke with left hemiparesis and without any identifiable history of neurological illness. She also had hypertension and hyperlipidemia which were not detected or treated earlier. No other risk factors of stroke were present. CT scan of brain showed hyperdense calcifications in putamen, globus pallidus, thalamus and cerebellar dentate nucleus and white matter consistent with Fahr's syndrome. MRI brain revealed acute ischemic infarct in right parietal, external capsular, and frontal lobe regions consistent with right MCA occlusion.

In our patient, there was no obvert manifestation of hypothyroidism but she had significantly reduced serum parathormone (iPTH) and vitamin D3 consistent with primary hypoparathyroidism and treatment was started with aspirin, atorvastatin, vitamin D3, calcium and calcitriol. Physiotherapy was initiated within 24 hrs and continued daily.

Movement disorder like dystonia and paratonia with Parkinsonism mimics are sometimes associated with cerebellar thalamic lenticular network dysfunction, and may be presenting symptoms of Fahr's syndrome [1,2]. Fahr's syndrome is often associated with other diseases like parathyroid dysfunction, usually hypoparathyroidism.

In our patient, there was no symptomatic movement disorder, but she had paratonic rigidity in her non-paretic upper limb which may be partially related to the basal ganglia degeneration.There has been one case report demonstrating acute ischemic stroke in Fahr's disease with positive MRI findings of lacunar infarct in right thalamus [3]. Another case of lacunar infarct in left posterior limb of internal capsule has been reported [4]. Only one case of a young child with hypoparathyroidism has been reported having large artery thrombotic stroke [5].

Our patient was an elderly woman with Fahr's syndrome and hypoparathyroidism who had a large artery athero-thrombotic stroke, and probably first of its kind reported in literature.
Researchers have observed a distinct association between reduced circulating 25(OH)D and risk of ischemic stroke in hypertensives [6].

CONCLUSION

The risk factor association between ischemic stroke and Fahr's syndrome has yet to be established, but cannot be overlooked especially in patients with hypoparathyroidism and hypovitaminosis D, considering its potentially treatable nature, with control of calcium and phosphate homeostasis.

REFERENCES

1) Amir A, Ghanem AA. "Familial Idiopathic Basal Ganglia Calcification (Fahr`s disease)"
2) .Neurosciences (Riyadh) 19 (2014): 171-7.
3) Sanjay BK, Bhansali A, Upreti V, Subbiah S, et al. "Spectrum of Neurological Manifestations of Idiopathic Hypoparathyroidism and Pseudohypoparathyroidism". Neurol India 59 (2011): 586-9.
4) Rocha E, Rouanet C, Reges DS, V, Gagliardi V, et al. "Fahr's Syndrome and Acute Ischemic Stroke: A Case Report". Stroke and Neurocritical Care Division, Department of Neurology and Neurosurgery, Federal University of Sao Paulo- UNIFESP, Sao Paulo, Brazil.

5) Chih Y, Lo CP, Wu MC. “Ischemic Stroke in a Young Patient With Fahr's Disease:
6) A Case Report . BMC Neurol 16 (2016): 33.
7) Dhawan RS, Jondhale SN, Sahu JK, Vyas S, et al. “Large Artery Stroke in a Child With Hypoparathyroidism”. J Child Neurol 30 (2015): 1084-5.
8) Christopher R, Majumdar V, Nagaraja D. “Vitamin D and Cerebrovascular Disease”. Ann Indian Acad Neurol 24 (2021): 130-1.