

Bilateral Posterior Subcapsular Cataract Revealing Fahr Syndrome: An Ophthalmologic Case Report

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ABSTRACT

Background: Fahr syndrome is a rare neurological disorder characterized by bilateral intracranial calcifications, particularly involving the basal ganglia. Ocular manifestations are uncommon but may significantly affect visual function.

Case presentation: We report the case of a 45-year-old patient presenting with progressive bilateral visual impairment associated with neurological symptoms including memory disturbances and extrapyramidal manifestations. Ophthalmologic examination revealed decreased visual acuity and bilateral posterior subcapsular cataract. Brain computed tomography demonstrated bilateral calcifications involving the caudate and lenticular nuclei. Laboratory investigations revealed severe hypocalcemia, hyperphosphatemia, and decreased parathyroid hormone levels. The patient underwent sequential cataract surgery by phacoemulsification with intraocular lens implantation, resulting in significant improvement in visual acuity.

Conclusion: Cataract represents a rare ocular manifestation of Fahr syndrome. Early ophthalmologic evaluation and surgical management can significantly improve visual outcomes and quality of life.

Keywords : Fahr syndrome; cataract; posterior subcapsular cataract; basal ganglia calcification; hypocalcemia.

1. INTRODUCTION

Fahr syndrome, also known as idiopathic basal ganglia calcification, is a rare neurological disorder characterized by abnormal bilateral intracranial calcifications involving the basal ganglia and other brain regions responsible for motor control.

The clinical presentation is highly variable and may include neurological, psychiatric, and cognitive manifestations. The condition may occur in familial or sporadic forms and is often associated with metabolic disturbances affecting calcium and phosphate metabolism [1].

Although neurological symptoms are the most frequently reported manifestations, ocular complications remain rare. Cataract formation has occasionally been described in association with chronic disturbances in calcium metabolism [2].

In this report, we present a case of bilateral posterior subcapsular cataract occurring in a patient with Fahr syndrome and discuss the possible pathophysiological mechanisms and ophthalmologic management.

2. CASE REPORT

A 45-year-old patient presented to the ophthalmology department with progressive bilateral visual impairment evolving over approximately 18 months. Neurological symptoms including memory disturbances and movement abnormalities had been present for nearly one year.

Ophthalmologic examination revealed a marked decrease in visual acuity in both eyes. Slit-lamp biomicroscopy demonstrated dense bilateral posterior subcapsular cataracts, while fundus examination was unremarkable.

Because of the neurological symptoms, brain imaging was performed. Computed tomography revealed bilateral symmetrical calcifications involving the basal ganglia, particularly the caudate and lenticular nuclei, consistent with Fahr syndrome.

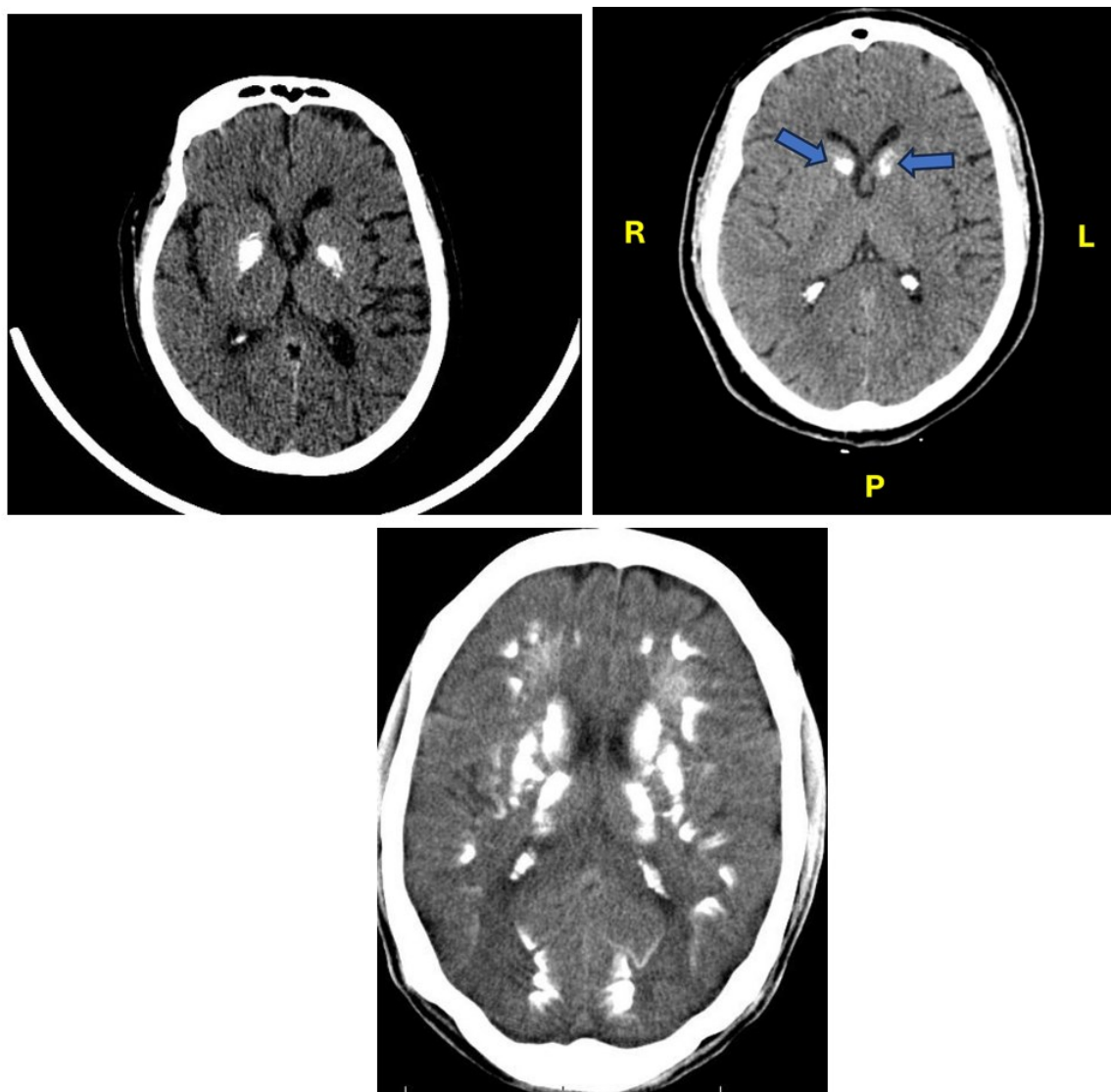


Figure 1: Brain CT scan showing bilateral calcifications involving the caudate and lenticular nuclei consistent with Fahr syndrome

Laboratory investigations revealed severe hypocalcemia ($\text{Ca}^{++} = 65.3 \text{ mg/L}$) and hyperphosphatemia (59.4 mg/L). Parathyroid hormone levels were decreased ($\text{PTH} = 4.2 \text{ pg/mL}$), and vitamin D levels were also reduced.

Based on clinical, radiological, and biological findings, the diagnosis of Fahr syndrome secondary to calcium-phosphate metabolism disturbance was established. Slit-lamp examination confirmed the presence of a dense posterior subcapsular cataract in both eyes.



Figure 2: Slit-lamp photograph showing dense posterior subcapsular cataract.

Because of the progressive decline in visual acuity, the patient underwent cataract extraction by phacoemulsification with implantation of an intraocular lens in both eyes. The procedures were performed sequentially with a two-month interval.

Postoperative evolution was favorable, with significant improvement in visual acuity.

3. DISCUSSION

Cataract is defined as a progressive opacification of the crystalline lens leading to decreased visual acuity. Although age-related cataract represents the most common etiology, metabolic disorders affecting calcium and phosphate balance may also contribute to cataract formation.

Fahr syndrome is a rare disorder characterized by abnormal deposition of calcium salts in various brain regions, particularly the basal ganglia. The estimated prevalence is less than one case per 100,000 individuals [2-3].

Ocular manifestations are uncommon but may occur due to metabolic disturbances associated with the disease. Chronic hypocalcemia has been proposed as a possible mechanism leading to cataract formation through alterations in lens metabolism and mineral deposition [4].

Posterior subcapsular cataract is frequently associated with metabolic disorders and may lead to significant visual impairment even in relatively young patients.

Computed tomography plays a crucial role in establishing the diagnosis of Fahr syndrome by revealing characteristic intracranial calcifications.

Management of cataract in these patients is similar to that in other forms of cataract. Phacoemulsification with intraocular lens implantation remains the gold-standard treatment and generally provides favorable visual outcomes.

A multidisciplinary approach involving ophthalmologists, neurologists, and endocrinologists is essential for optimal management.

4. CONCLUSION

Cataract represents a rare but potentially disabling ocular manifestation of Fahr syndrome. Recognition of this association is important, particularly in patients presenting with visual impairment associated with neurological symptoms.

Early diagnosis and appropriate surgical management can significantly improve visual outcomes and quality of life.

ETHICAL CONSIDERATION

This study was conducted in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and the Helsinki Declaration of 1975, as revised in 2000. The authors declare that they have obtained the necessary authorizations for publication.

CONFLICTS OF INTEREST

All authors declare that they have no conflicts of interest.

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AI DECLARATION

The authors declare that generative AI tools were used solely for language correction, editing, and formatting.

All outputs generated by automated tools were carefully reviewed and validated by the authors, who take full responsibility for the manuscript's accuracy and integrity.

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