Indian Journal of Surgical Oncology (June 2025) 16(3):807–812
https://doi.org/10.1007/s13193-024-02137-z

CASE REPORT

Solitary Metastasis Mimicking Glioblastoma in a Patient with Fahr's Disease

Onur Taydas[1] · Busra Has[2] · Zakir Sakci[3] · Hayri Ogul[4] · Mecit Kantarci[5]

Received: 7 January 2024 / Accepted: 20 November 2024 / Published online: 30 November 2024

Abstract

Fahr's disease is defined as the accumulation of bilateral idiopathic calcification in the basal ganglia, dentate nucleus, and centrum semiovale. The coexistence of Fahr's disease and intracranial tumors is extremely rare, with only five cases documented in the literature. This study aimed to present a rare case, discussing both similarities and differences with previously reported cases. A 70-year-old female patient presented to our hospital with a progressive headache over the past month. Brain magnetic resonance imaging (MRI) with intravenous contrast was performed for further evaluation. Bilateral basal ganglia, thalamus, and dentate nuclei showed calcifications consistent with Fahr's disease. After excluding toxic, infectious, and endocrine causes of calcification, the patient was diagnosed with Fahr's disease. Magnetic resonance imaging also revealed a midline localized, irregularly contoured, and heterogeneously intense mass with contrast enhancement at the centrum semiovale level, accompanied by peripheral edema. Radiologically, the mass resembled a high-grade glioma or metastasis, and the presence of a lipid peak further suggested metastasis. Based on radiological findings, the patient was diagnosed with brain metastasis of breast carcinoma. The patient received chemotherapy for metastasis, which resulted in substantial regression of the mass. In patients with known Fahr's disease, new-onset headaches and related symptoms should warrant evaluation for possible brain tumors. In these cases, imaging findings should be assessed along with physical examination findings and the patient's medical history to support the diagnostic process.

Keywords Fahr's disease · Glioblastoma · Metastasis · Magnetic resonance imaging

Introduction

Fahr's disease is defined as the accumulation of bilateral idiopathic calcification in the basal ganglia, dentate nucleus, and centrum semiovale [1]. Symptoms typically appear in the fourth and fifth decades, although cases in childhood have also been documented, albeit rarely [2]. Three main criteria are typically sought in diagnosing Fahr's disease: bilateral, non-atherosclerotic idiopathic calcification of the basal ganglia, psychiatric symptoms, and choreoathetosis or extrapyramidal movement disorders. Many patients with basal ganglion calcifications are asymptomatic, which limits understanding of the origin and pathology of the disease [3].

The coexistence of Fahr's disease and intracranial tumors is exceptionally rare, with only five cases having been reported in the literature. This study aimed to present a rare case, discussing both similarities and differences with previously documented cases.

Case Report

A 70-year-old female patient presented to our hospital with a progressive headache over the past month. She also reported nausea and frequent vomiting over the last few months. The patient had a history of lumpectomy and sentinel lymph node biopsy for luminal-A type T2N0M0 stage ductal breast

✉ Hayri Ogul
drhogul@gmail.com

1 Department of Radiology, Medical Faculty, Sakarya University, Sakarya, Turkey

2 Department of Radiology, Medical Faculty, Duzce University, Duzce, Turkey

3 Department of Radiology, Health Sciences University Umraniye Training, and Research Hospital, Istanbul, Turkey

4 Department of Radiology, Medical Faculty, Istanbul Medipol University, Istanbul, Turkey

5 Department of Radiology, Medical Faculty, Ataturk University, Erzurum, Turkey

cancer 10 years earlier, followed by ten courses of radiotherapy and 5 years of tamoxifen therapy after surgery. She had been followed up in remission for 10 years. Her physical examination and laboratory findings were unremarkable. Brain magnetic resonance imaging (MRI) with intravenous contrast was performed for further evaluation and revealed T1 and T2 hypointense calcifications in the bilateral basal ganglia, thalamus, and dentate nuclei, consistent with Fahr's disease. Blooming artifacts in these areas in susceptibility-weighted imaging (SWI) (Fig. 1A–C) and hyperdense calcifications in the pathologic signal areas in computed tomography (CT) (Fig. 2A–C) further supported Fahr's disease..

To exclude possible causes, the patient's serum calcium, parathyroid hormone, phosphorus, vitamin D, and alkaline phosphatase levels were examined, but they were all found within normal limits. After excluding toxic, infectious, and endocrine causes, the patient was diagnosed with Fahr's disease. MRI also revealed a midline, irregularly contoured, heterogeneously intense mass with contrast enhancement at the centrum semiovale level, accompanied by peripheral edema (Fig. 3A–C), suggestive of a high-grade glioma or metastasis. Diffusion-weighted imaging ($b = 1000$ s/mm^2) showed the lesion as hypointense, while the apparent diffusion coefficient (ADC) map demonstrated hypointensity accompanied by restricted diffusion (Fig. 4A, B). Diffusion tensor tractography color mapping indicated invasion and disruption of callosal commissural fibers (Fig. 5A). Dynamic susceptibility contrast (DSC) magnetic resonance perfusion showed a hyperperfused, transcallosal heterogeneous lesion (Fig. 5B). Magnetic resonance spectroscopy revealed a marked elevation of lipids at 1.3 and 0.9 parts per million and an increased Cho/NAA ratio (Fig. 6A–C), further suggesting metastasis.

Based on radiological findings, the patient was diagnosed with brain metastasis of breast carcinoma. The patient underwent chemotherapy with five cycles of 150 mg/m^2

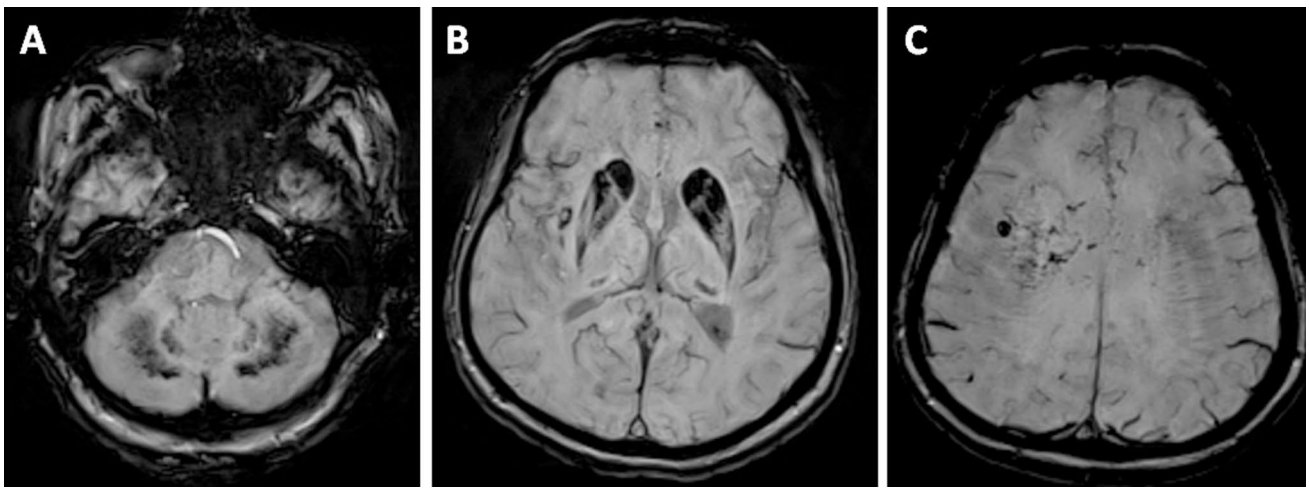

Fig. 1 Consecutive SWI scans (**A**, **B**, and **C**) showing symmetric blooming artifacts in the bilateral basal ganglia, thalamus, and dentate nuclei

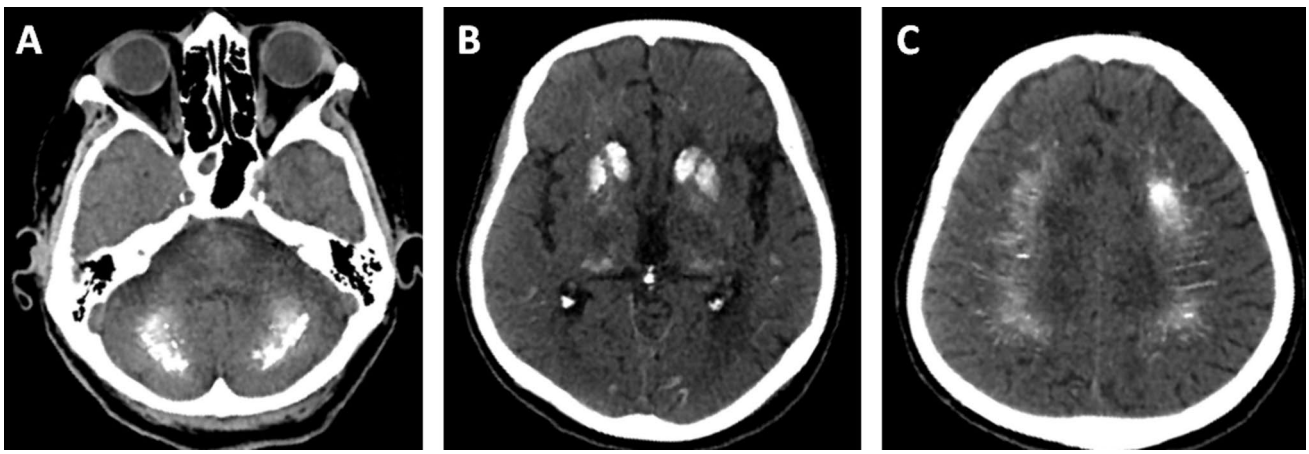

Fig. 2 Consecutive CT scans (**A**, **B**, and **C**) revealing hyperdense calcifications in the pathologic signal areas observed on SWI

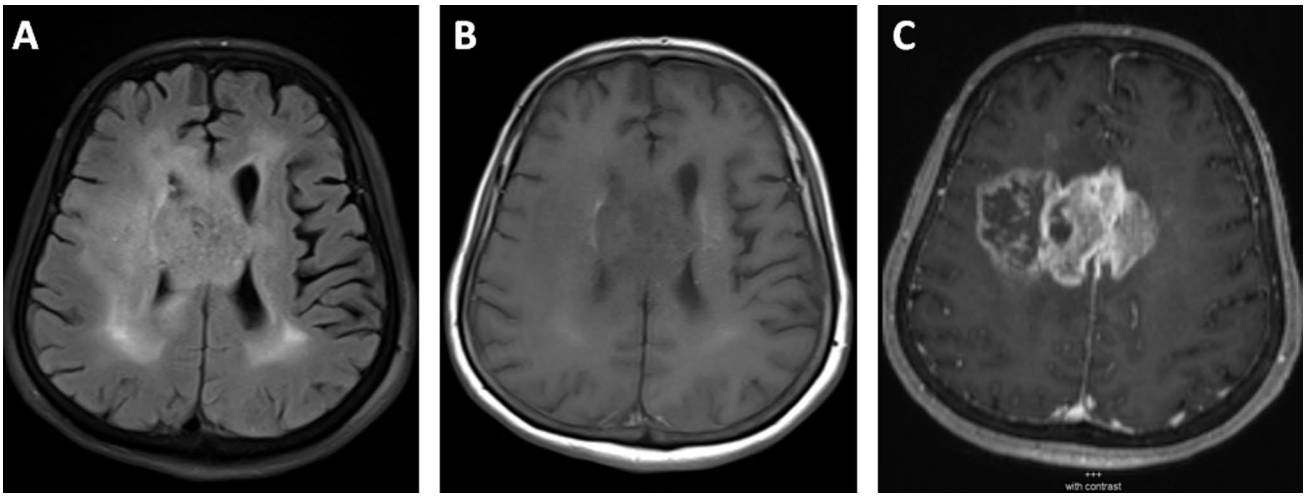

Fig. 3 Fluid attenuation inversion recovery (FLAIR) weighted (**A**) and per- (**B**), and post-contrast (**C**) T1-weighted MRI sequences showing a midline, irregularly contoured, heterogeneously intense mass with contrast enhancement at the centrum semiovale level, accompanied by peripheral edema

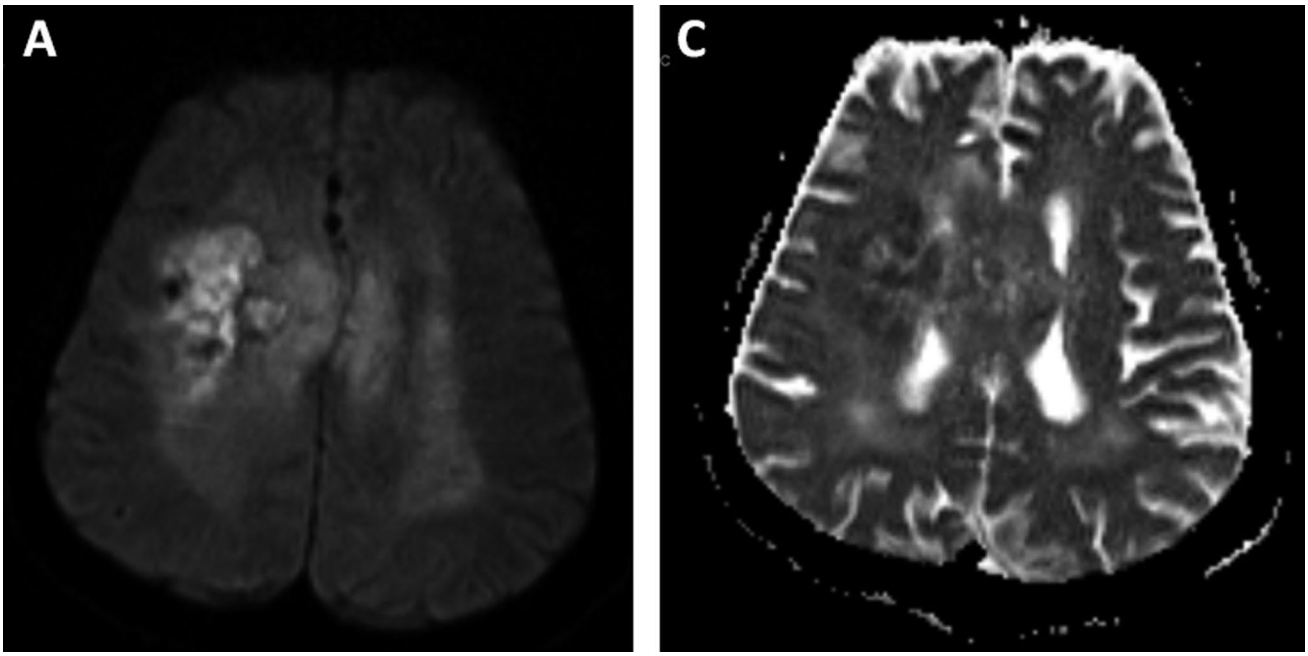

Fig. 4 *DWI* ($b = 1000$ s/mm^2); **A** image showing a hyperintense signal within the lesion and the ADC map **B** showing hypointensity accompanied by restricted diffusion

temozolomide combined with 75 mg/m^2 cisplatin every 28 days. Chemotherapy response was evaluated using the response assessment in neuro-oncology brain metastases (RANO-BM) criteria [4]. A follow-up gadolinium-enhanced MRI, obtained 1 month after the completion of chemotherapy (6 months after the metastasis diagnosis), showed a 60% reduction in the target lesion size, with the patient clinically stable and not requiring additional corticosteroids. In light of these findings, the patient was evaluated as having a partial response according to RANO-BM criteria. The clinical and radiological diagnoses of the mass were confirmed as brain metastasis of breast cancer.

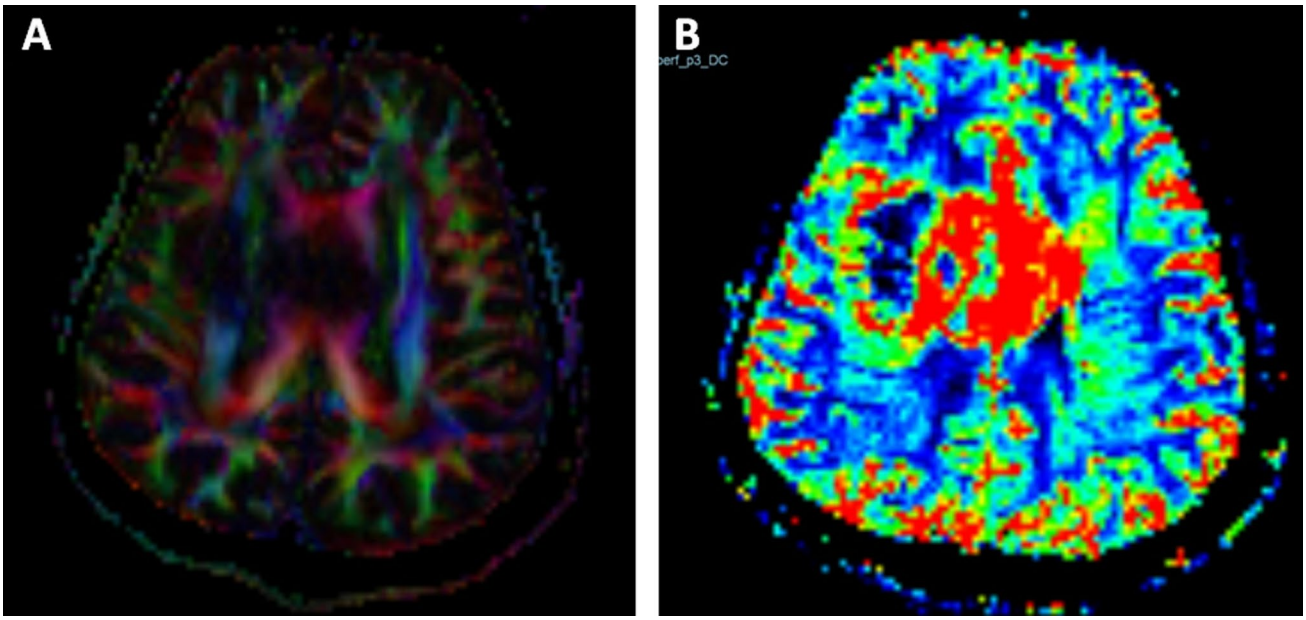

Fig. 5 Diffusion tensor tractography color map (**A**) revealing invasion and disruption of callosal commissural fibers and DSC MR perfusion showing a hyperperfused transcallosal lesion

Discussion

To date, only five cases of brain tumors associated with Fahr's disease have been reported in the literature [5–8]. Patient ages ranged from 8 to 36 years, with a median age of 22 years. Our patient, at age 70, was older than those previously reported. Three of the documented patients were female, and two were male, indicating a slight female predominance, and our patient was also female. Unlike the previous cases of low-grade gliomas, our case involved a diagnosis of brain metastasis from breast cancer, making it unique in the literature. Tumor locations in previous cases were infratentorial in four patients and supratentorial in one. In our patient, the tumor was supratentorial. While all infratentorial tumors were reported to be cystic lesions without contrast enhancement located in the cerebellar hemisphere, the tumor in our case had intense contrast enhancement and no cystic component.

Due to advances in imaging methods, diagnosing calcium accumulation in the brain has become easier. Calcified areas can be easily identified by their hyperdense appearance on CT scans. On MRI, calcium shows different imaging properties. Because calcium is a diamagnetic substance, hyperintensity may be seen on T1-weighted images. However, as calcium concentration increases, the signal intensity typically decreases. On T2-weighted series, calcium may appear either hypointense or hyperintense, depending on the specific minerals present in the deposits. Hyperintensity may also result from inflammatory processes that can accompany Fahr's disease. In recent years, SWI, a 3D gradient echo sequence, has enabled more accurate detection of calcium compared to conventional MRI sequences [9].

In Fahr's disease, lesions are typically bilateral and symmetrically located. Calcifications tend to be diffuse, extensive, and conglomerated, most commonly found in the basal ganglia, especially the putamen. However, they may also occur in the dentate nuclei, thalamus, brain stem, centrum semiovale, and subcortical white matter [10].

In conclusion, in patients with known Fahr's disease, brain tumors should be investigated in cases of new-onset headaches and similar symptoms. In these patients, imaging findings, along with physical examination findings and the patient's medical history, play a critical role in the diagnostic process.

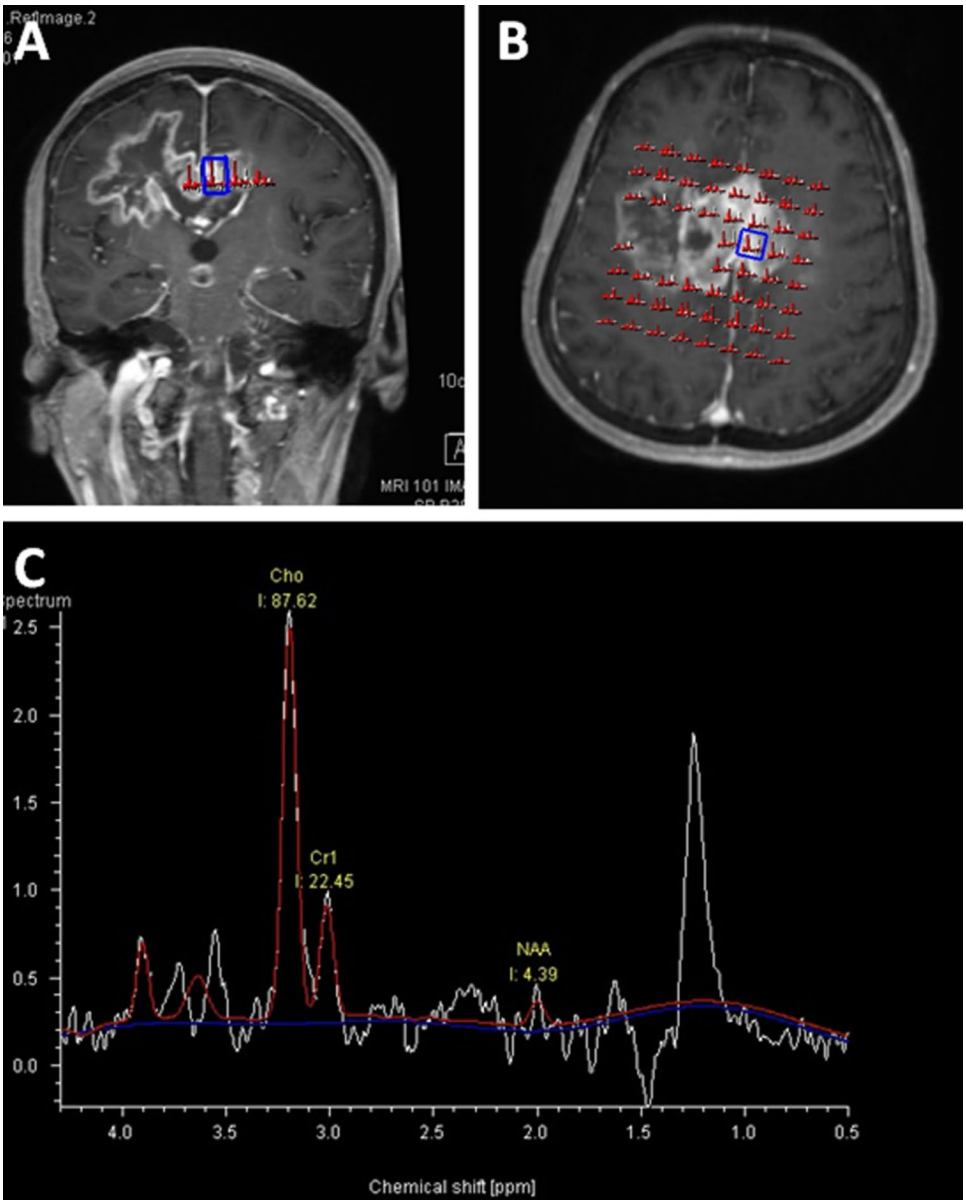

Fig. 6 Magnetic resonance spectroscopy images (**A**, **B**, and **C**) showing a marked elevation of lipids at 1.3 and 0.9 parts per million and an increased Cho/NAA ratio

Declarations

Conflict of Interest The authors declare no competing interests.

References

1. Donzuso G, Mostile G, Nicoletti A, Zappia M (2019) Basal ganglia calcifications (Fahr's syndrome): related conditions and clinical features. Neurol Sci 40:2251–2263
2. Billard C, Dulac O, Bouloche J, Echenne B, Lebon P, Motte J et al (1989) Encephalopathy with calcifications of the basal ganglia in children. A reappraisal of Fahr's syndrome with respect to 14 new cases. Neuropediatrics 20:12–9
3. Forstl H, Krumm B, Eden S, Kohlmeyer K (1992) Neurological disorders in 166 patients with basal ganglia calcification: a statistical evaluation. J Neurol 239:36–38
4. Lin NU, Lee EQ, Aoyama H, Barani IJ, Barboriak DP, Baumert BG, Bendszus M, Brown PD, Camidge DR, Chang SM, Dancey J, de Vries EG, Gaspar LE, Harris GJ, Hodi FS, Kalkanis SN, Linskey ME, Macdonald DR, Margolin K, Mehta MP, Schiff D, Soffietti R, Suh JH, van den Bent MJ, Vogelbaum MA, Wen PY (2015) Response assessment in neuro-oncology (RANO) group. Response assessment criteria for brain metastases: proposal from the RANO group. Lancet Oncol 16(6):e270-8
5. Ang LC, Rozdilsky B, Alport EC, Tchang S (1993) Fahr's disease associated with astrocytic proliferation and astrocytoma. Surg Neurol 39:365–369
6. Morimoto K, Mogami H, Ozaki K (1984) Fahr's disease associated with cystic astrocytoma. Eur Neurol 23:265–268
7. Ozerov SS, Semenova Zh B, Zubairaev MS, Pitskhelauri DI, Ozerova VI, Strebkova NA (2004) Concurrence of Fahr's disease with

cerebellar tumors. Zh Vopr Neirokhir Im N N Burdenko 34–5. discussion 35–6
8. Lin Q, Piao Y, Ling F, Xu G (2015) Concurrence of Fahr's disease and brain tumor: a case report and review of the literature. Exp Ther Med 10:1915–1917
9. Sahin N, Solak A, Genc B, Kulu U (2015) Fahr disease: use of susceptibility-weighted imaging for diagnostic dilemma with magnetic resonance imaging. Quant Imaging Med Surg 5:628–632
10. Snijders BMG, Peters MJL, van den Brink S, van Trijp MJCA, de Jong PA, Vissers LATM, Verduyn Lunel FM, Emmelot-Vonk MH, Koek HL (2024) Infectious diseases and basal ganglia calcifications: a cross-sectional study in patients with Fahr's disease and systematic review. J Clin Med 13(8):2365