Bilateral striopallidodentate calcinosis (Fahr’s disease)

Madam, Bilateral striopallidodentate calcinosis (Fahr's disease), is a rare syndrome characterized by symmetrical calcification over the basal ganglion and dentate nucleus. We report the case of a young female who met with a road traffic accident and CT scan showed calcification in basal ganglia. A 40 year old female presented with the history of a fall from an auto-rickshaw. Following the fall she had 2-3 episodes of vomiting and right ear bleed. She was complaining of persistent headache. Her general and systemic examination was unremarkable except high blood pressure (140/100 mmHg). There were no focal neurological deficits. In view of multiple episodes of vomiting and persistent headache she was investigated with computerized tomography (CT scan). CT scan head showed calcification in both basal ganglia (Figure). There were no clinical features suggestive of hypoparathyroidism. Blood and urine investigations including serum calcium and phosphate levels were normal. A diagnosis of idiopathic bilateral basal ganglionic calcification (Fahr's disease) was made. With the increasing use of CT scan it has been possible to detect subtle calcifications in basal ganglia in many patients without any previous neurological symptoms. As in the present case, imaging findings of symmetric and extensive calcification are usually typical and conspicuous. Neurological and psychiatric symptoms, if present at all, are highly variable and include progressive mental deterioration, convulsive seizures, parkinsonism, dysarthria and ataxia, psychosis and affective disorders. In suspected cases of Fahr's disease, other causes of intracranial calcification must be ruled out, such as parathyroid disorders, vascular lesions, infectious diseases like toxoplasmosis, syphilis and inflammatory illnesses such as systemic lupus erythematosus. As in our case usually the patients with idiopathic disease need only symptomatic support.

Amit Agrawal
Division of Neurosurgery, Datta Meghe Institute of Medical Sciences, Sawangi (Meghe), Wardha- 442004, Maharashtra, India.

References

Retraction of Original Article in whole

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Author: Nighat Nisar, Majid Hafeez Qadri, Kiran Fatima, Shakeela Parveen
Institution: Community Medicine Department, Sindh Medical College DUHS, Baqai Medical University, Parasitology Department, Karachi University
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