DIFFUSE ENCEPHALIC CALCIFICATION

A CASE REPORT

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SUMMARY — The basal ganglia calcification is known since the last century but with the new neuroimage techniques (CT scan) its diagnosis became more frequent specially in asymptomatic patients. The (authors report a case with non-familial primary diffuse encephalic calcification with exuberant calcifications on cerebral hemispheres, cerebellum and brain stem, seen on CT scan.

KEY WORDS: Fahr's syndrome, basal ganglia calcification, diffuse encephalic calcification.

Calcificação encefálica difusa: registro de um caso

RESUMO — Os autores descrevem um caso de calcificação encefálica difusa primária, nâo-fiamilial, cuja primeira manifestação foi disartria. Posteriormente, desenvolveu movimentos coreoatetósicos em mãos e face, discalculia e déficit de memória. A TC de crânio revelou calcificações em hemisférios cerebrais, cerebelo ei tronco cerebral.

PALAVRAS-CHAVE: síndrome de Fahr, calcificação dos gânglios da base* calcificação encefálica difusa.

The first description of the histology of basal ganglia calcification (BGC) was made by Delacour (1850), Virchow and Bamberger (1855) 2,13,15,19. in 1931, Fahr reported a case with BGC not associated with atherosclerotic changes of the cerebral vessels[11]. Since then his name was linked with this condition. Fritzche, Kasanin and Crank (1935) described, by the first time, the findings of BGC on the plain X-ray 3,i7. Eaton and colleagues (1939) noted the association between BGC and hypoparathyroidism, and Sprage, later, between the former and pseudohypoparathyroidism 2. The BGC is, nowadays, a more frequent finding as the CT scan becomes widely available, and not only in the basal ganglia but also in the cerebral hemispheres, cerebellum and thalamus 1,9,10,14,21.

Intracerebral calcifications can be classified according to their distribution in median, bilateral asymmetrical and symmetrical. The latter will be considered as diffuse encephalic calcifications (DEC), being divided as primary or secondary. Their more frequent etiologies are shown in Table 1. Primary DEC probably has a familiar trace, the autosomal dominant being the most common hereditary pattern 5. There are sporadic cases reported on the literature whose etiology and familiar history can not be determined but those are rare[12]. The clinical manifestations of DEC are variable, ranging from asymptomatic pati-

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Table 1. 27 possible etiologies of BGC (modified from Harrington and col.7).

Idiopathic hypoparathyroidism	Tuberous sclerosis
Secondary hypoparathyroidism	Parkinsonism
Pseudohypoparathyroidism	Vascular disease
Pseudo-pseudohypoparathyroidism	Cerebral haemorrhage
Hyperparathyroidism	Radiation therapy
Hypothyroidism	Methotrexate therapy
Carbon monoxide intoxication	Cytomegalic inclusion disease
Birth anoxia	Encephalitis
Lead intoxication	Toxoplasmosis
Fahr's syndrome	Cysticercosis
Familial idiopathic symmetrical BGC	Congenital Chagas disease *(16)*
Hastings-James syndrome	Nephronophthisis *(18)*
Cockayne's syndrome	Systemic mitochondrial disease *(8)*
Lipoid proteinosis	

ents, whose calcifications are incidental findings, to those who experience neurological manifestations: rigidity, tremor, dystonia, blepharospasm, choreoathetosis, dementia, seizures, pyramidal syndrome, cerebellar syndrome, visual and speech complaints and rarely, cranial nerve palsies and intracranial hypertension 6,8,13,15,1 z.19,20. Pathologically, the calcified lesions are mainly located on the arterioles wall (media and adventitia lamina), venules, capillaries and perivascular region *9. Microscopic analysis discloses a proteic gel core which contains acid and neutral mucopolisacarides and other elements such as calcium, phosphorus, chlorine, iron, sulphur, potassium, magnesium, aluminium, manganese and zinc, the latter being found in high concentration According to Eaton, the process begins with a local ischemia followed by edema, anoxia, necrosis and mumification with secondary colloid ferrugination 13. The vascular change secondary to a parenchymatous «lésion» is said by Fenelon and Guillard to be the basic pathogenesis of the disease[5]. Contrariwise to the secondary type, the treatment of the DEC is purely symptomatic.

We report the case of a patient with non-familial primary diffuse encephalic calcification with exuberant calcifications on cerebral hemispheres, cerebellum and brain stem, seen on CT scan.

CASE REPORT

JBP (Reg. 194113-5), a 49-year-old right-handed white man was referred to the Neurologic Clinic of the HUCFF-UFRJ because of dysartria with an insidious onset and progressive course since the age of 46. Meanwhile his family noted some difficulty on calculation. He denied any other disease or familial history of neurological disease. The physical exam was normal and the neurologic examination disclosed choreoathetoio movements on the hands, grimacing, unsustained closing of the eyelids, dysartria, dyscalculia and he reached 22 points on the Mini-Mental Status Test. The laboratory workup was normal, including serum and urinary calcium and phosphate and serum magnesium. The skull X-ray disclosed calcifications on the bilateral basal ganglia topography. A CT scan showed an extensive calcinosis (Fig. 1).

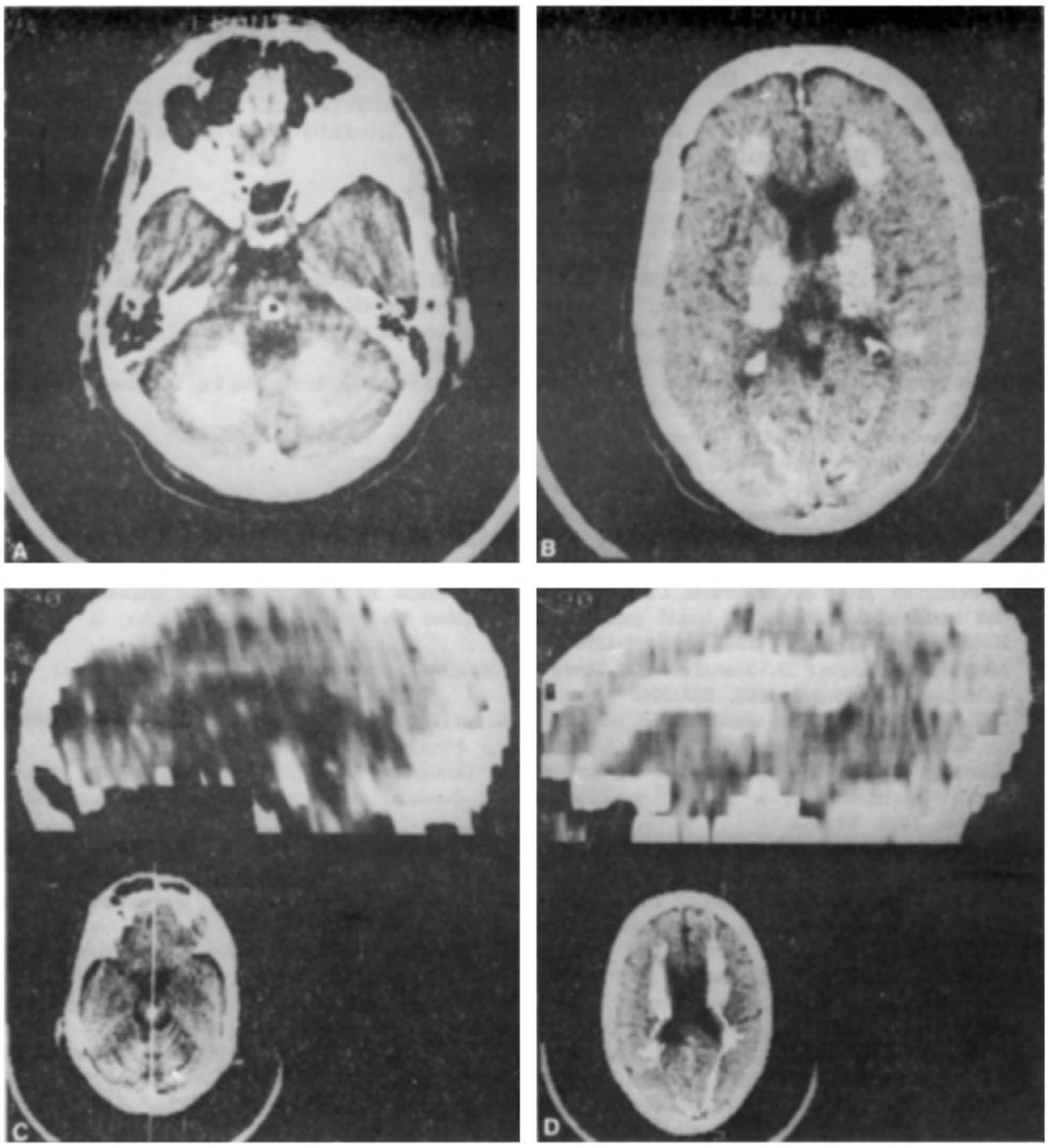

Fig. 1. Case JBF. Axial unenhanced computed tomography scans showing calcification of: A. celebellum and pons; B. frontal and occipital lobes and basal ganglia; C and D. brainstem and periventricular region in transverse reconstruction.

COMMENTS

This case calls attention because of some uncommon aspects of the disease. The more frequent kinds of initial symptoms are seizures and mental retardation, but in this patient the first manifestation was dysartria. On the review of the literature we found only one case reported with dysartria as the initial complaint 13.

Other interesting aspect is the lack of familial history of BGC or underlyng disease, allowing the inclusion of the case as a sporadic type of DEC.

The originality of this case is directly related to the tomographic features of the DEC found on the cerebral hemispheres, cerebellum, basal ganglia and brainstem. We did not find any reference on the literature review about calcification on the pons region.

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