

Progressive ataxia and palatal tremor: T1-weighted with magnetization transfer pulse hyperintensity in the inferior olivary nucleus

Ataxia progressiva com tremor palatal: hiperintensidade nos núcleos olivares inferiores na sequencia T1 com pulso de transferência de magnetização

James Henrique Yared^{1,3}, Bruno Siqueira Campos Lopes², Ricardo Mendes Rogério², Lázaro Luis Faria do Amaral², Nelson Fortes Paes Diniz Ferreira¹

¹Neuroradiology Department, ALTA medicina diagnóstica, São Paulo SP, Brazil;

²Neuroradiology Department, Medimagem, São Paulo SP, Brazil;

³Neurology Department, Hospital Beneficência Portuguesa, São Paulo SP, Brazil.

Correspondence: James Henrique Yared; Rua Teixeira da Silva 363; 04002-031 São Paulo SP - Brasil; Email jamesyared@hotmail.com

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Progressive ataxia and palatal tremor (PAPT) is a rare distinct clinical entity characterized by symptomatic palatal tremor associated with progressive ataxia^{1,2}. Few cases have been described in literature, and MRI findings show, in most cases, cerebellar atrophy and bilateral olivary hypertrophy with signal change¹. We report a case where MRI study at 3T demonstrated marked bilateral olivary complex hyperintensity at T1, with magnetization transfer contrast pulse (MTC) sequence, which was subtle at conventional T2 and FLAIR sequences at 1,5 and 3 T.

CASE

A 54-year-old male patient presented with a 10-year history of predominantly truncal ataxia manifested mainly by difficulty in climbing stairs with associated oscillopsia and spontaneous nystagmus with closed eyes. He showed significant gait instability on heel-to-toe test and also a 2Hz palatal tremor. Familial history of ataxia was negative, as well as genetic tests for spinocerebellar ataxia 1, 2, 3 and 7. Routine blood analysis, thyroid-stimulating hormone, vitamin B12 and antinuclear antibodies were normal.

MRI examinations at 1,5 and 3,0 T showed only moderate cerebellar atrophy and subtle signal change in olivary nuclei on conventional FSE T1, T2 and FLAIR sequences, which were overlooked at the first report. Additional images on 3 T using FSE T1-weighted with magnetization transfer pulse (TR1000, TE 20) showed marked signal alteration of olivary complexes (Figure).

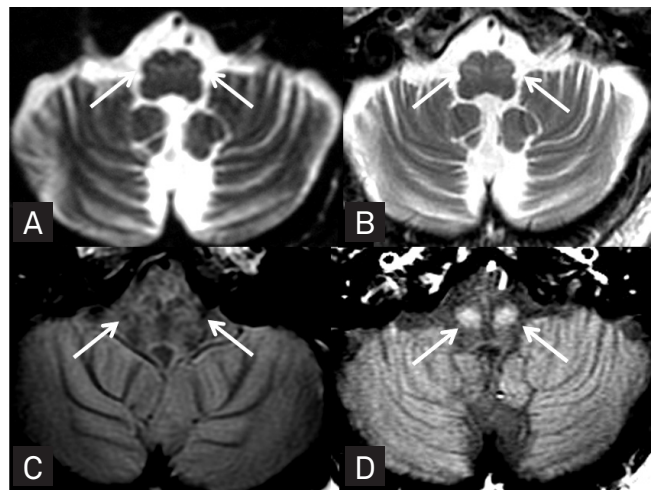


Figure. Patient: Axial T2 FSE at 1,5 T (A) and 3 T (B) and T1 with MTC (TE/TR = 20/1000 ms) (C) showing cerebellar atrophy and evident hyperintensity in olivary nuclei, mainly on image (D), Normal control: T1 with MTC (TE/TR = 20/1000 ms).

DISCUSSION

Progressive ataxia and palatal tremor (PAPT) is a rare disorder more commonly found as a sporadic condition with no identifiable cause. Most cases manifest as slowly progressing ataxia, that can be associated with visual changes, dysarthria and dysphagia^{1,2}. Typical MRI abnormalities are cerebellar atrophy, hypertrophy and hyperintensity at T2-weighted and proton density sequences at olivary complexes. The main differential diagnosis of sporadic PAPT is hypertrophic olivary degeneration, a

condition that is believed to be the result of a monophasic illness, such as vascular injury, trauma, demyelinating lesion and posterior fossa tumor with detectable lesion at Guillain Mollaret triangle^{3,4}.

Magnetization Transfer Contrast is a well-known MRI technique based on the application of off-resonance radiofrequency pulses that can be used to obtain an alternative contrast method and tissue specificity⁵. The prominent hyperintensity detected at T1-weighted MTC images could be explained, as in HOD, by myelin loss or even transynaptic degeneration. We found no previous

cases of PAPT using T1 with MTC sequence in the medical literature.

Not all cases of PAPT reported in literature have MRI showing abnormalities in the olivary complexes. This could be explained by the radiologist experience or even by the fact that some believe the alterations can be present for only a fraction of time during the course of the disease. Our case suggests that using higher magnetic fields and unconventional techniques such as T1-weighted MTC can bring additional information and raise sensitivity for the detection of abnormalities of the olivary complexes in PAPT.

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