

Rare occurrence of Hirayama disease in Brazil

Rara ocorrência da doença de Hirayama no Brasil

Mateos Bogoni¹, Bernardo Corrêa de Almeida Teixeira¹, Marianna Cioni¹

We present the case of a 17-year-old male with oblique unilateral hand and forearm amyotrophy, paresthesia and paresis. Tremor was present on finger extension. Hirayama disease was suspected, which led to dynamic cervical MRI. Hirayama disease affects predominantly young Asian males^{1,2}, with few cases reported in the Americas³.

In this condition, underdevelopment of the spinal dura mater results in posterior dural detachment on the cervicothoracic region during flexion movements, which leads to asymmetric compression and ischemia of anterior horn cells⁴. The posterior venous plexus becomes engorged⁵, which is seen on dynamic MRI as a high T2 signal crescent in the posterior epidural space during flexion, with intense enhancement (Figures 1 and 2).

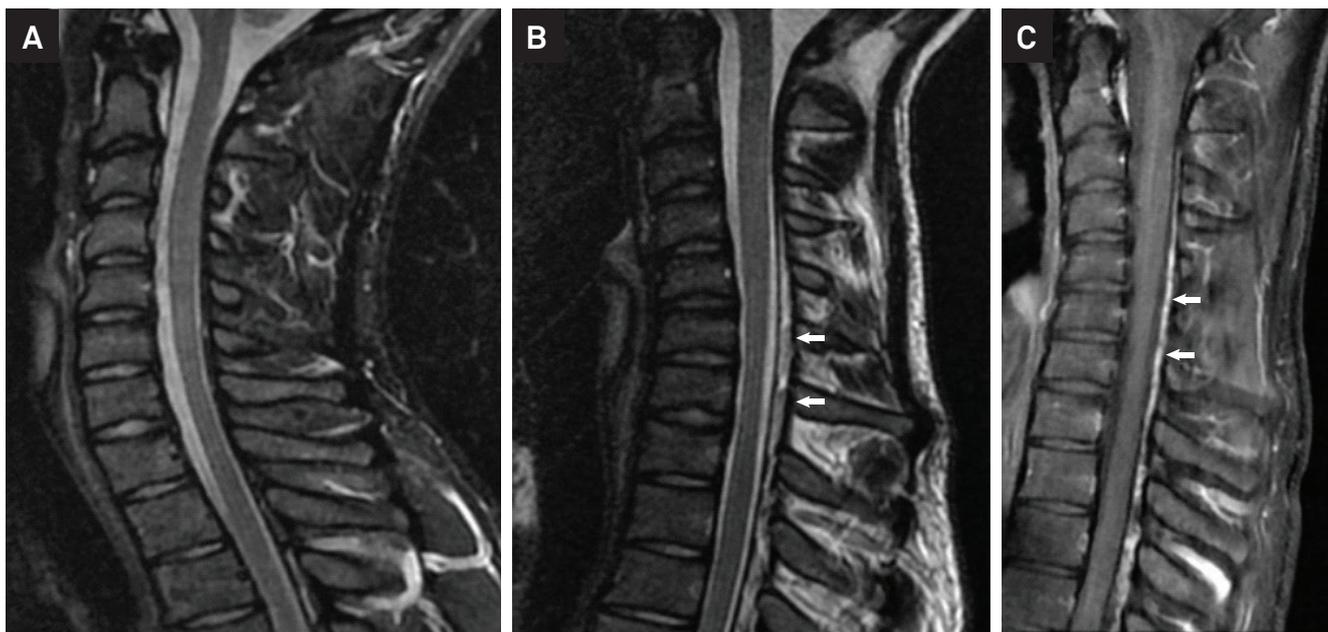


Figure 1. a) Sagittal STIR. Imaging in the neutral cervical position may appear inconspicuous at first; b) Sagittal T2 with cervical flexion. We notice detachment of the posterior dural layer, with reduction of the vertebral canal width and slight intramedullary hyperintensity. The engorged posterior vertebral venous plexus appears as a high signal crescent in the posterior epidural space (white arrows); c) Sagittal T1 Gd+ with cervical flexion. Uniform and intense enhancement of the posterior vertebral venous plexus is observed (white arrows).

¹Universidade Federal do Paraná, Hospital de Clínicas, Departamento de Clínica Médica, Setor de Radiologia e Diagnóstico por Imagem, Curitiba PR, Brasil.

Mateos Bogoni  <https://orcid.org/0000-0002-3892-3455>

Correspondence: Mateos Bogoni; Universidade Federal do Paraná, Hospital de Clínicas; Rua General Carneiro, 181; 80060-900 Curitiba PR, Brasil; E-mail: bogonimateos@gmail.com

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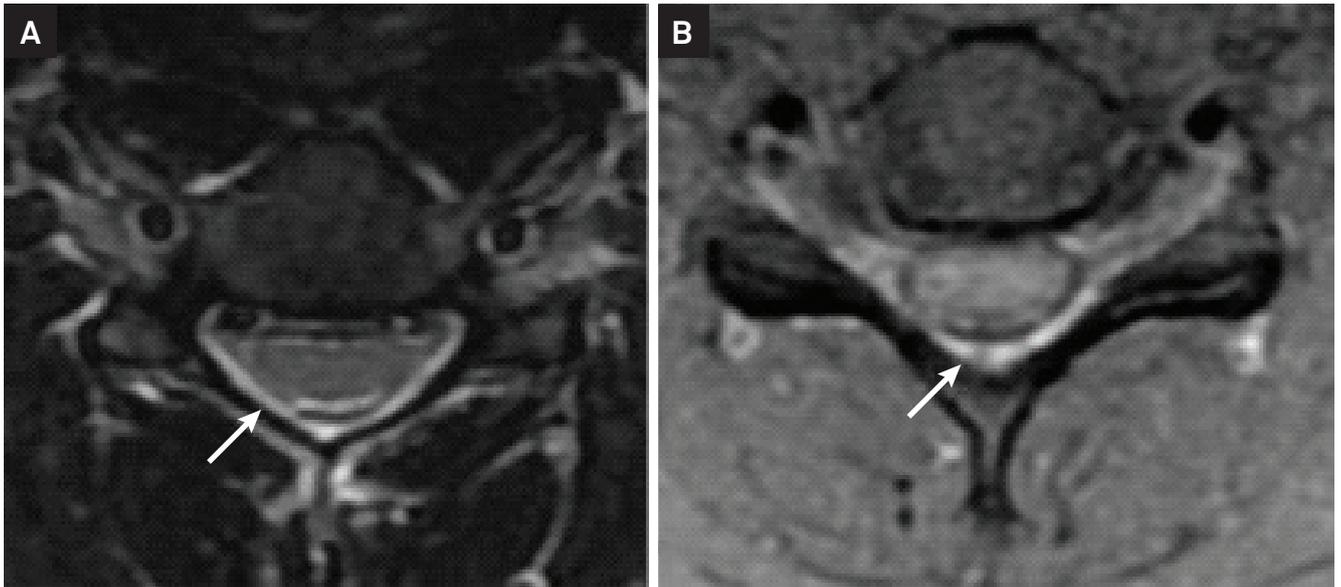


Figure 2. Axial T2 weighted (a) and T1 post-gadolinium (b) images show the posterior vertebral venous plexus (white arrows) in the epidural space during cervical flexion.

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