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"Ears of the lynx" sign in hereditary spastic paraplegias is not always the same!

Sinal da orelha do lince nas paraplegias espásticas hereditárias nem sempre é o mesmo!

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Hereditary spastic paraplegias (HSPs) are a heterogeneous group of genetic disorders featuring lower extremity spasticity and weakness. The "ears of the lynx" is a magnetic resonance imaging (MRI) sign that refers to degeneration of the forceps minor. Despite mimicking acquired conditions, like ependymitis granularis, it is considered a hallmark of some HSPs, such as types 15 and 11.[1] However, there were other types reported to have this sign,[2,3] particularly when accompanied by atrophy of the anterior portion of the corpus callosum.

A 34-year-old man with walking disturbance presented with spastic paraparesis in the lower limbs, cerebellar ataxia, and sensory–motor polyneuropathy. A brain MRI was performed (►**Figure 1**), and the exam showed a biallelic pathogenic mutation in spastic paraplegia type 7 (c.376 + 1 G > T / c.1369C > T / ENST00000645818).

We recommend brain MRIs to search for this sign in all suspected cases of HSP. However, it is important to interpret with caution when finding "ears of the lynx" in isolation, with absence of corpus callosum atrophy.

Author's Contributions

VHRM: conceptualization of the work, data acquisition, and writing of the manuscript; BDRRA: data acquisition and writing of the manuscript; FF: conceptualization of the work, data acquisition, and writing and review of the manuscript. All authors approved the final version of the manuscript and agree to be responsible for all aspects of the work.

Conflict of Interest

The authors have no conflict of interest to declare.

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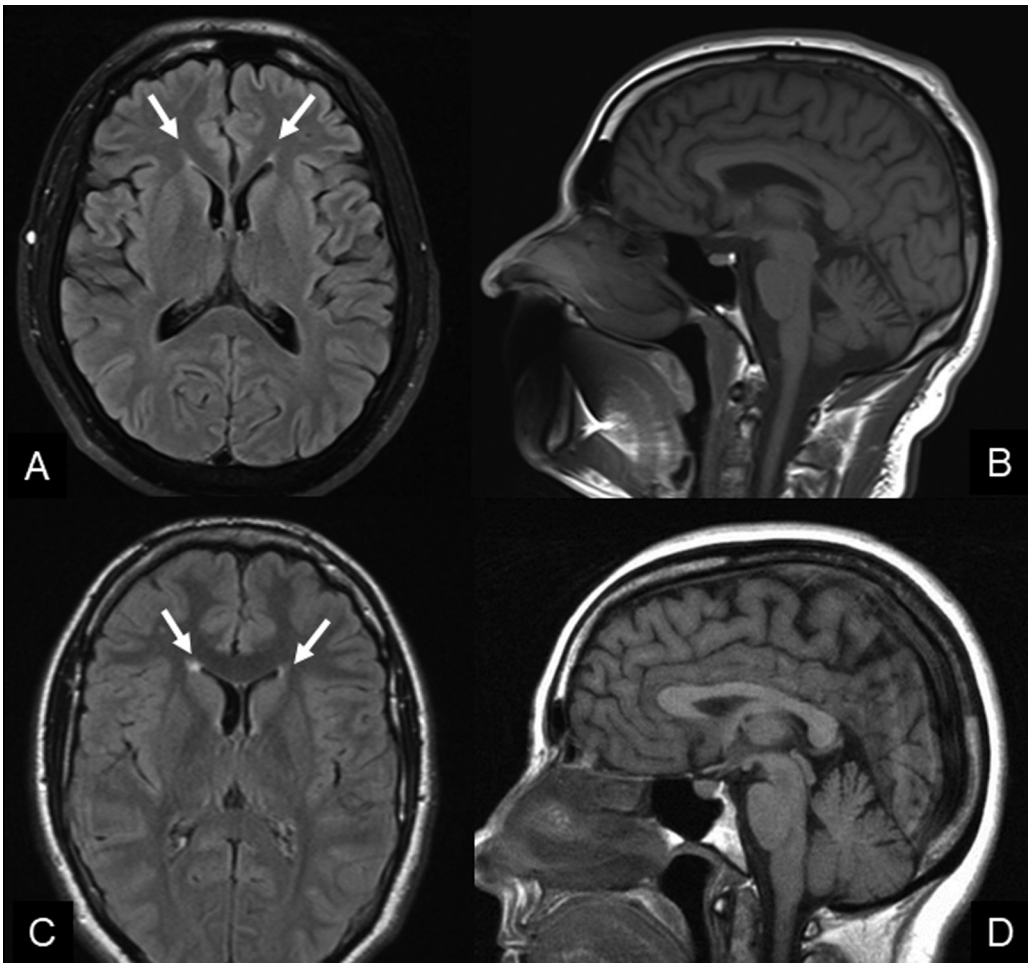

Figure 1 (A) Axial fluid attenuated inversion recovery (FLAIR), with white arrow showing the “ears of the lynx” sign and (B) Sagittal T1 showing cerebellar atrophy. Both (C) and (D) show a normal examination with an important differential diagnosis of “Ears of the lynx” signal, ependymitis granularis, which is usually more rounded and subtle.

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