Teaching NeuroImages: Limbic encephalitis associated with relapsing polychondritis

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Figure Follow-up brain MRI and systemic manifestations

Axial fluid-attenuated inversion recovery images. (A) Initial brain MRI shows bilateral temporal lobe hyperintensities, more prominent in mesial areas, especially right (arrow). (B) Follow-up MRI performed 9 months later discloses progressive temporal lobe atrophy and persistent brain hyperintensities. (C, D) Auricular chondritis and episcleritis.

A 43-year-old man presented with subacute fever, costochondritis, headache, seizures, and amnesia. Brain MRI (figure) showed bilateral temporal lobe hyperintensities. CSF revealed 105 white cells/mm³ (77% lympho-monocytes), elevated protein, and normal glucose. Infection and paraneoplastic/autoimmune workup was negative. He improved after corticosteroid treatment.

Nine months later, he experienced a neurologic relapse, associated with bilateral auricular chondritis and episcleritis (figure), fulfilling the McAdam criteria for relapsing polychondritis.¹ Despite aggressive immunotherapy, severe cognitive deficits persisted.

CNS involvement is uncommon in relapsing polychondritis¹ and may present as limbic encephalitis. Autopsy findings have implicated vasculitis as the underlying pathogenesis.²

AUTHOR CONTRIBUTIONS

Mateus Mistieri Simabukuro had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. He contributed to study concept and design, acquisition of data, analysis and interpretation of data, drafting of the manuscript, and critical revision of the manuscript for important intellectual content. Leandro Tavares Lucato had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. He contributed to study concept and design, acquisition of data, analysis and interpretation of data, drafting of the manuscript, and critical revision of the manuscript for important intellectual content. Samuel Katsuyuki Shinjo contributed to study concept and design, acquisition of data, analysis and interpretation of data, drafting of the manuscript, and critical revision of the manuscript for important intellectual content. Wellington Lima Flores contributed to study concept and design, acquisition of data, and analysis and interpretation of data. Luiz Henrique Martins Castro had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. He contributed to study concept and design, acquisition of data, analysis and interpretation of data, drafting of the manuscript, and critical revision of the manuscript for important intellectual content.

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