Case Studies

Granulomatous Angiitis of the Central Nervous System Associated with Hodgkin's Lymphoma: Case Report and Literature Review

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Granulomatous angiitis of the central nervous system (GACNS) is a rare cerebrovascular disorder. It usually presents with multifocal neurologic symptoms symptoms including stroke, encephalopathy, and headache. A limited number of case reports describe neurological deficits resulting from GACNS as the manifesting symptoms of Hodgkin's lymphoma (HL). We describe the case of a patient with neurological symptoms from GACNS that led to the diagnosis of HL, as well as a literature review focusing on the association between GACNS and HL.
Key Words: Granulomatous angiitis—Hodgkin's lymphoma—CNS vasculitis—paraneoplastic—angiitis—stroke—cancer.

Introduction

Granulomatous angiitis of the central nervous system (GACNS) is a rare cerebrovascular disease.[1] The disease can present with neurological symptoms including headache, stroke, confusion, and obtundation.[1] GACNS pathology is characterized by lymphocytic, epithelioid, and histiocytic transmural inflammation of the blood vessel wall with the presence of multinucleated giant cells.

A limited number of case reports describe neurological deficits resulting from GACNS as the first presentation of Hodgkin's lymphoma (HL), making the clinical recognition of the disease particularly important. We report a patient presenting with neurological symptoms from GACNS that led to the diagnosis of classic HL, followed by a literature review describing common features to help medical practitioners diagnose this condition accurately.

Case Presentation

A 25-year-old woman reported a 1-year history of intermittent headaches. She then developed speech difficulty with word substitution and dysnomia. A neurological examination showed mixed aphasia but was otherwise nonfocal.

Brain magnetic resonance imaging with gadolinium demonstrated large confluent white matter hyperintensities with multifocal, punctate perivascular enhancement (Fig 1, A-C). Cerebrospinal fluid revealed elevated proteins at 57 mg/dL (normal range, 15-45 mg/dL), glucose of 42 mg/dL, red blood cell count of 2/μL, and white blood cell count of 26/μL (normal range, 0-5 leukocytes/μL); opening pressure was 25 cmH_2O, with normal cytology.

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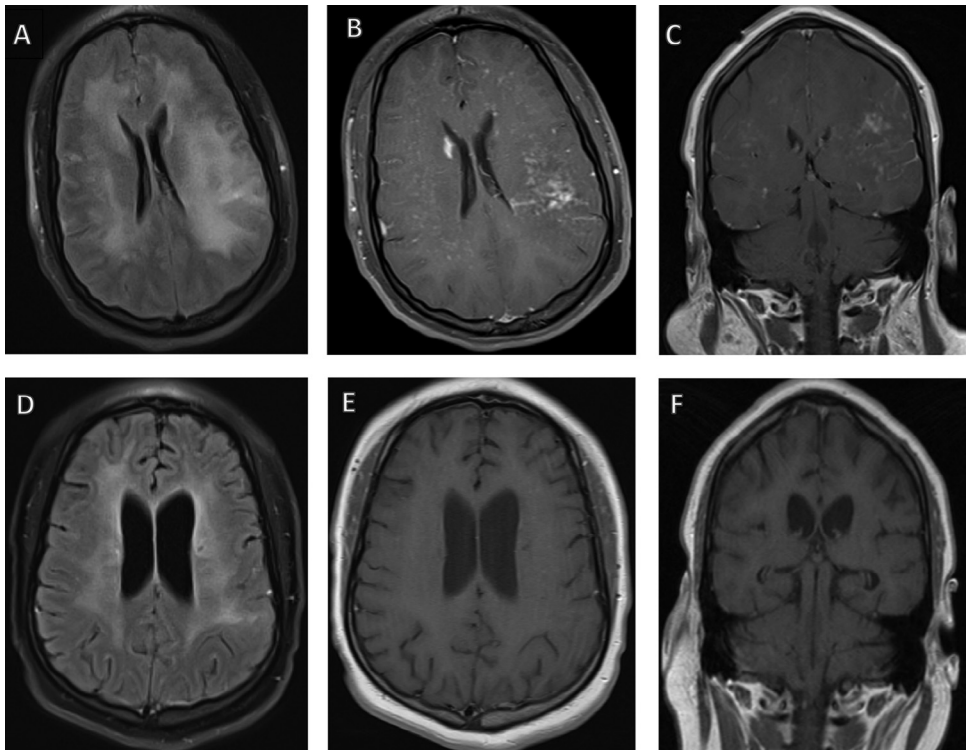

Figure 1. *MRI brain before and after treatment. (A) T2 FLAIR sequence demonstrates extensive white matter T2 hyperintensities in the frontal, temporal, and parietal lobes. (B) Axial and (C) Coronal T1 postgadolinium show innumerable punctate and linear enhancing foci bilaterally within both cerebral hemispheres. Many enhancing lesions are perivascular in distribution, and there is leptomeningeal enhancement within several convexity sulci. (D) T2 FLAIR sequence improvement in the previously noted hyperintensities and (E) axial and (F) coronal T1 postgadolinium images shows no evidence of enhancement 20 months after cyclophosphamide and ABVD therapy.*

Cerebrospinal fluid bacterial culture, Lyme screen, venereal disease research laboratory test, varicella-zoster virus, herpes simplex virus, and Epstein–Barr virus polymerase chain reaction were negative. Paraneoplastic autoantibodies, antinuclear and antineutrophil cytoplasmic antibodies, rapid plasma reagin, and urine toxicology screen were negative.

A large painless, lymph node was palpated in the left supraclavicular area. PET CT of the chest, abdomen, and pelvis revealed mediastinal hypermetabolic lymphadenopathy including paratracheal and subcarinal lymph nodes (Fig 2). A right axillary lymph node was biopsied, and pathological analysis was consistent with classic HL and nodular sclerosis subtype (Fig 3, A).

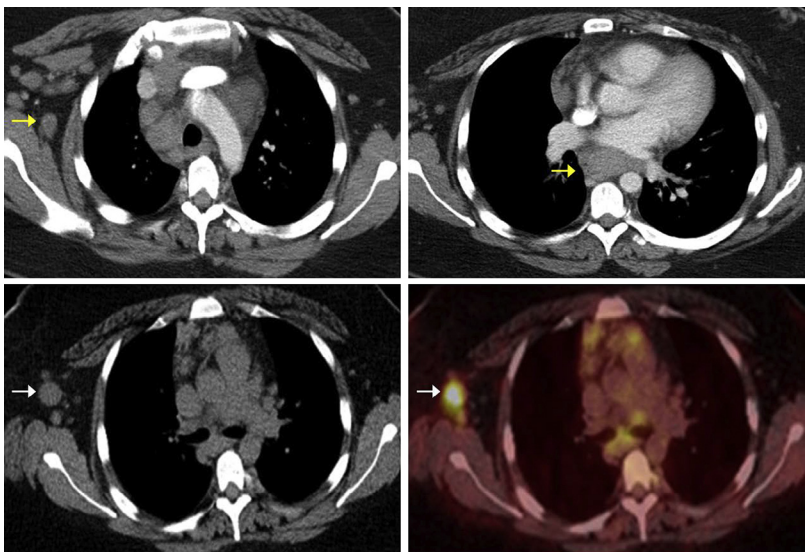

Figure 2. *PET computed tomography scan of the chest/abdomen/pelvis demonstrates bulky mediastinal and axillary lymphadenopathy (arrows).*