Progressive Multifocal Leukoencephalopathy

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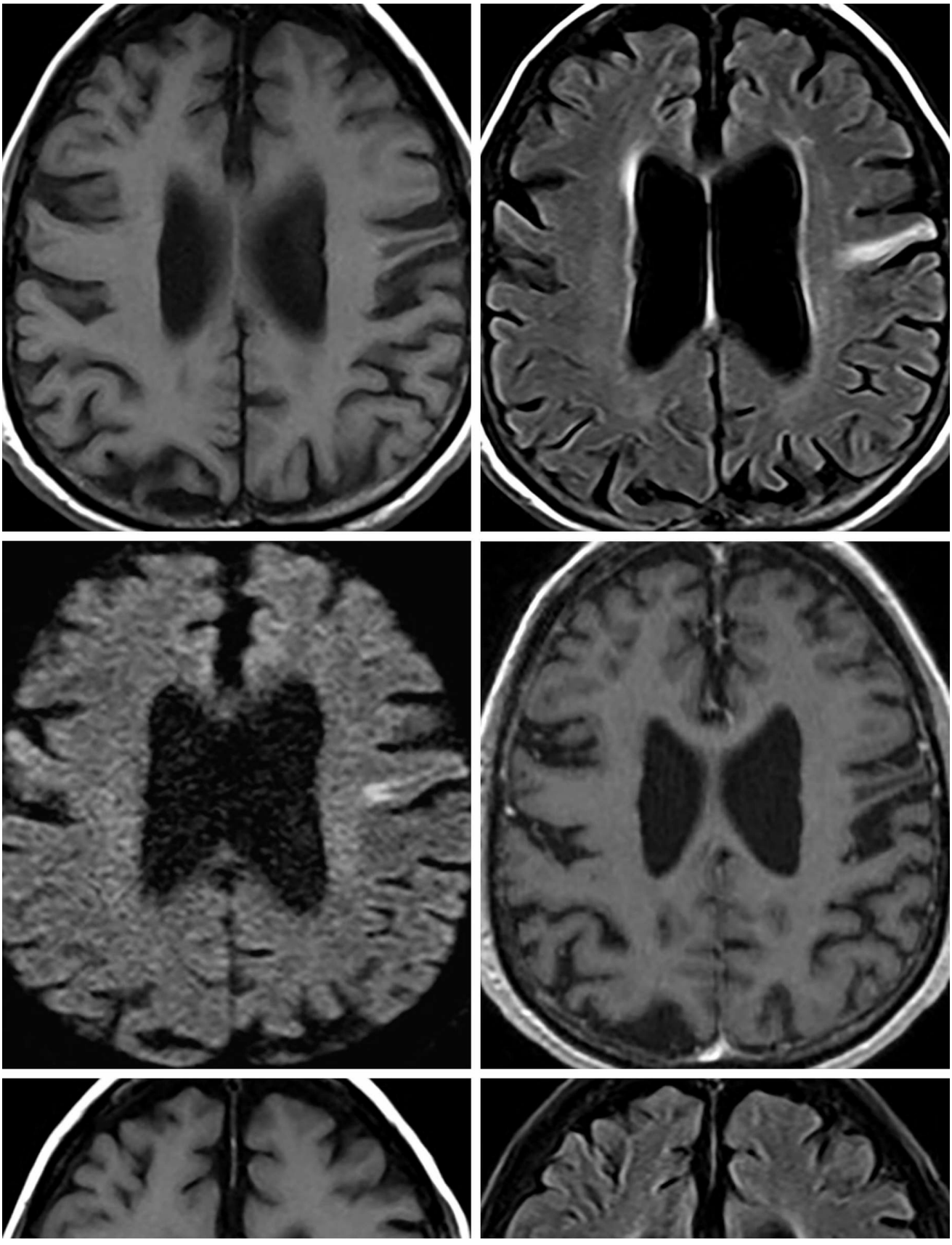

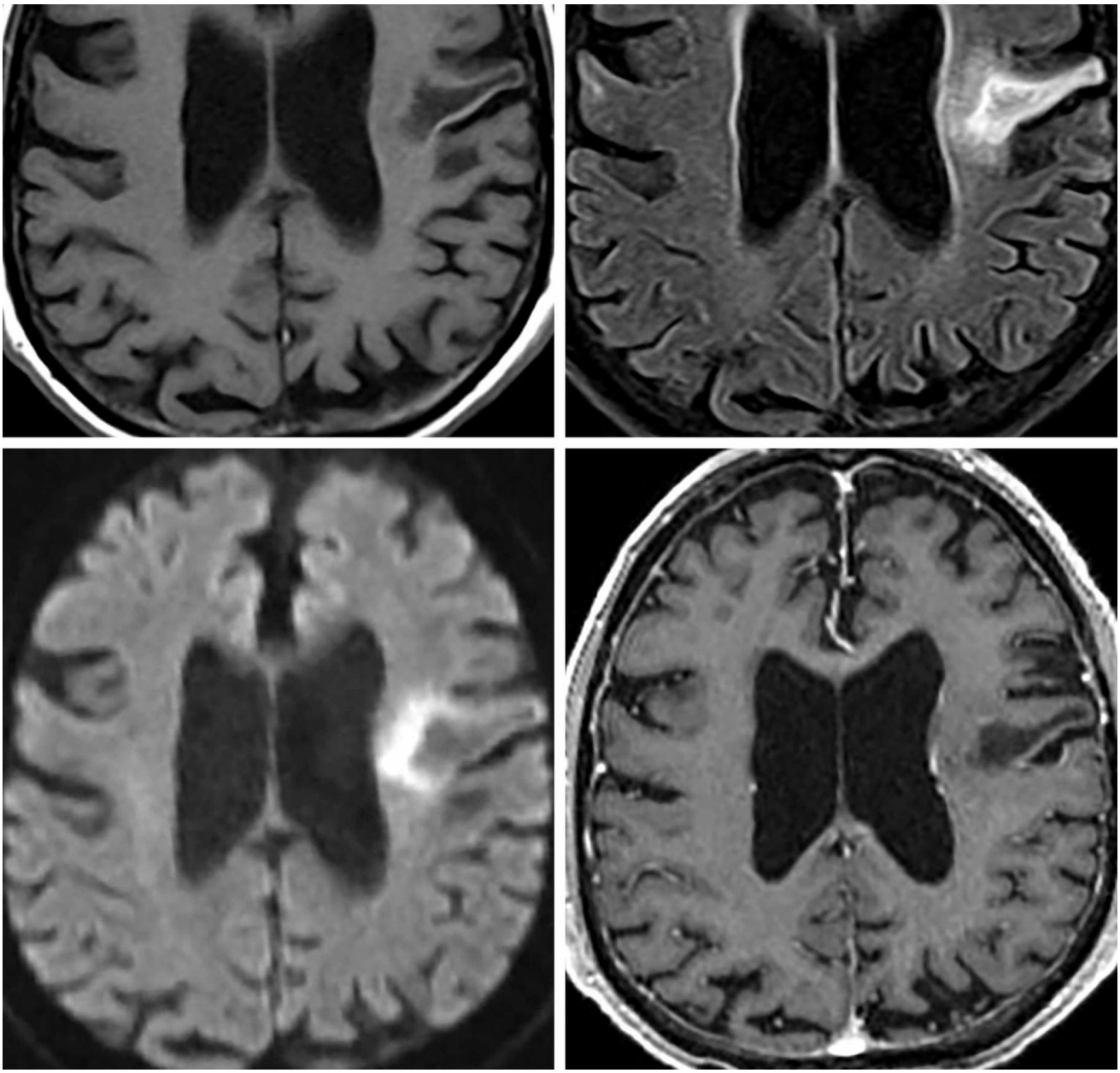

Figure 1: Initial imaging of a patient with AIDS demonstrates a linear, T1-hypointense (top left), FLAIR-hyperintense (top right) region within the subcortical white matter of the left dorsolateral frontal lobe with associated volume loss. There was minimal associated peripheral reduced diffusivity (second row left) and no significant contrast enhancement (second row right). Little was known about the patient at presentation to the emergency department, and given the patient's recent onset of symptoms, a diagnosis of subacute infarction was made. Follow-up examinations over the next several months demonstrated a significant interval increase in T1-hypointense (third row left), FLAIR-hyperintense (third row right) signal abnormality within the subcortical white matter extending to the lateral ventricle. The degree of adjacent reduced diffusivity continued to increase in size. (Bottom Left) Postcontrast imaging does not show abnormal enhancement. The continued increase

in size and degree of signal abnormality on multiple follow-up examinations raised concern of a glioma. Ultimately, the lesion was resected, and findings were compatible with progressive multifocal leukoencephalopathy.

Description

- Opportunistic infection caused by JC polyomavirus

Pathology

- Hematogenous spread of JC virus or reactivation of latent brain lesion
- Progressive demyelinating disorder that results from JC virus infection of the myelin-producing oligodendrocytes

Clinical Features

- Symptoms
    - Altered mental status, progressive neurological symptoms, headache, lethargy
- Demographics
    - Associated with immune suppression
        - AIDS, postorgan transplant, on chemotherapy or biologic agents (most commonly natalizumab, a common treatment for refractory multiple sclerosis)
- Prognosis
    - 30% to 50% mortality rate in the first few months

Imaging

- General
    - Asymmetric, multifocal, demyelinating plaques involving the subcortical and deep white matter
    - Mass effect is unusual
    - Enhancement may be associated with improved survival rates

    - Evaluate pattern of enhancement. If increasing, this is suggestive of immune reconstitution syndrome.
- Modality specific
    - CT
        - Asymmetric focal zone(s) of low attenuation that involve the periventricular and subcortical white matter
    - MRI
        - T1WI
            - Hypointense
        - T2WI/FLAIR
            - Hyperintense
            - Involves the juxtacortical white matter
        - DWI
            - May demonstrate peripheral diffusion restriction in acute setting
        - Contrast
            - Faint peripheral enhancement may be present
        - MRS
            - Decreased *N*-acetyl-aspartate (NAA), increased lactate, choline, and lipids
- Imaging recommendations
    - Standard-protocol MRI (including DWI) with intravenous contrast
- Mimic
    - The appearance may be very similar to an infiltrative glioma and often has a similar time course and growth pattern; striking T1 hypointensity and juxtacortical (without cortical) involvement often help to distinguish

For more information, please see the corresponding chapter in Radiopaedia and the Progressive Multifocal Leukoencephalopathy chapter within the Cerebral Infectious Diseases subvolume in *The Neurosurgical Atlas*.

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