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Teaching NeuroImages: Diffuse Midline Glioma Mimicking Edema in Cerebral Venous Thrombosis

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Diffuse Midline Glioma Mimicking Edema in Cerebral Venous Thrombosis

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A 57-year-old man presented with two weeks of headache, binocular diplopia, and progressive confusion. CT venogram showed extensive cerebral venous thrombosis (CVT) (Figure 1A). He was started on a heparin drip before conversion to therapeutic enoxaparin. Initial (Figure 1B) and follow-up (Figure 1C) MRI brain imaging showed left thalamic FLAIR hyperintensity without diffusion restriction (Figure 1D-E). Thalamic biopsy revealed a diffuse midline glioma (DMG), H3 K27-altered, CNS WHO grade 4 (Figure 2). Whole exome sequencing confirmed the presence of *H3-3A* (previously *H3F3A*) K27M and absence of mutations in IDH1/2.[1] Initial differential diagnosis for the FLAIR hyperintensity favored edema due to venous congestion as the result of thrombosis of the left vein of Trolard (Figure 1F). Given a negative hypercoagulability panel, after biopsy confirmed diagnosis, the CVT was thought to be precipitated by the DMG, which likely carries an increased risk of CVT similar to glioblastoma.[2] By the follow-up Neuro-Oncology visit his mental status had returned to normal. His anticoagulation was changed to apixaban and he began a course of temozolamide and radiation.

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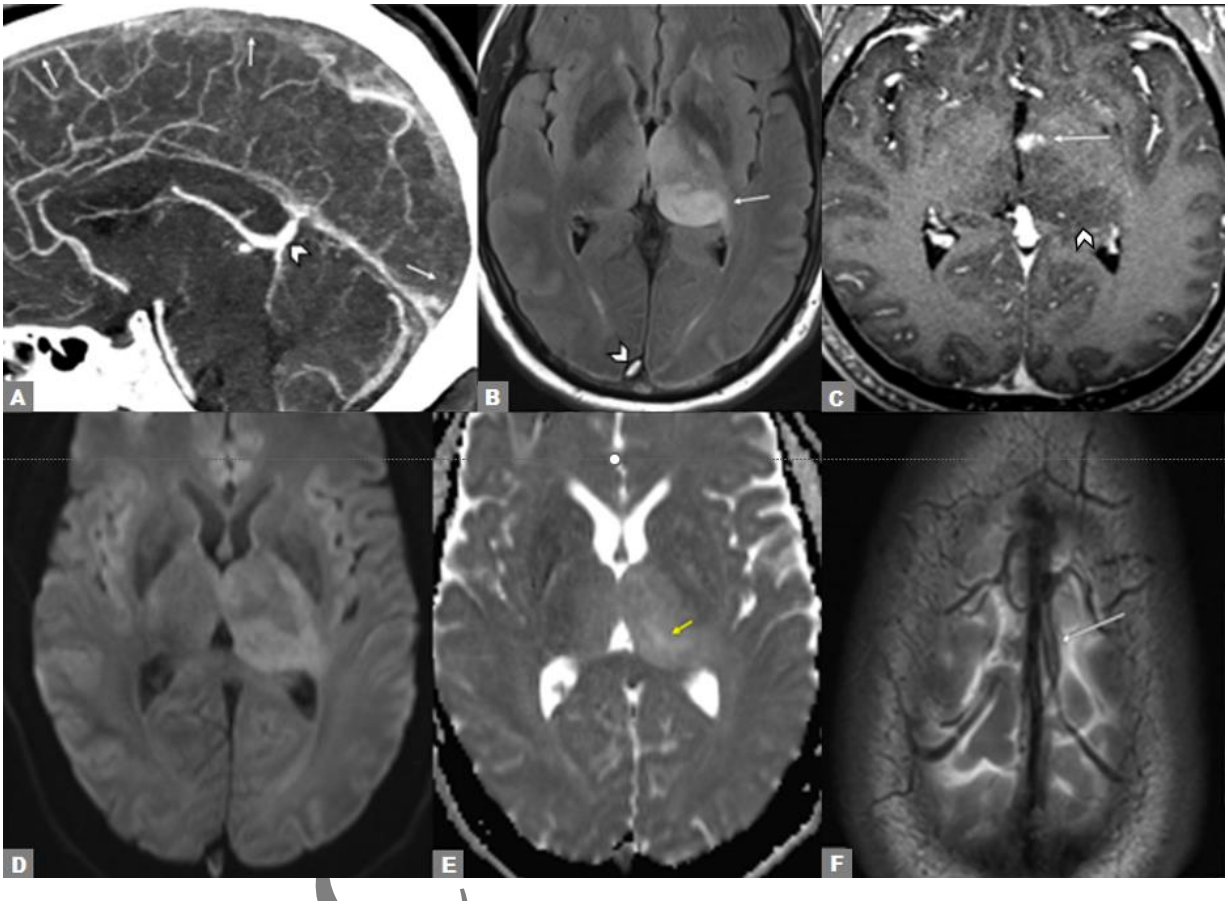

Figure 1. CT Venogram and MRI of the Brain. (A) Initial CT venogram demonstrates filling defects in the superior sagittal sinus (SSS, arrows). Deep cerebral veins (arrowhead) enhanced normally. (B) Axial T2 FLAIR image demonstrates expansile left thalamic T2 hyperintensity (arrow) and SSS thrombosis (arrowhead). (C) Subsequent postcontrast T1 image shows focal enhancement (arrow) within the thalamic lesion (arrowhead). (D) DWI and (E) ADC demonstrate facilitated diffusion (yellow arrow) which did not help distinguish between venous congestion and glioma. (F) Axial T2 image demonstrates left vein of Trolard thrombosis (arrow).

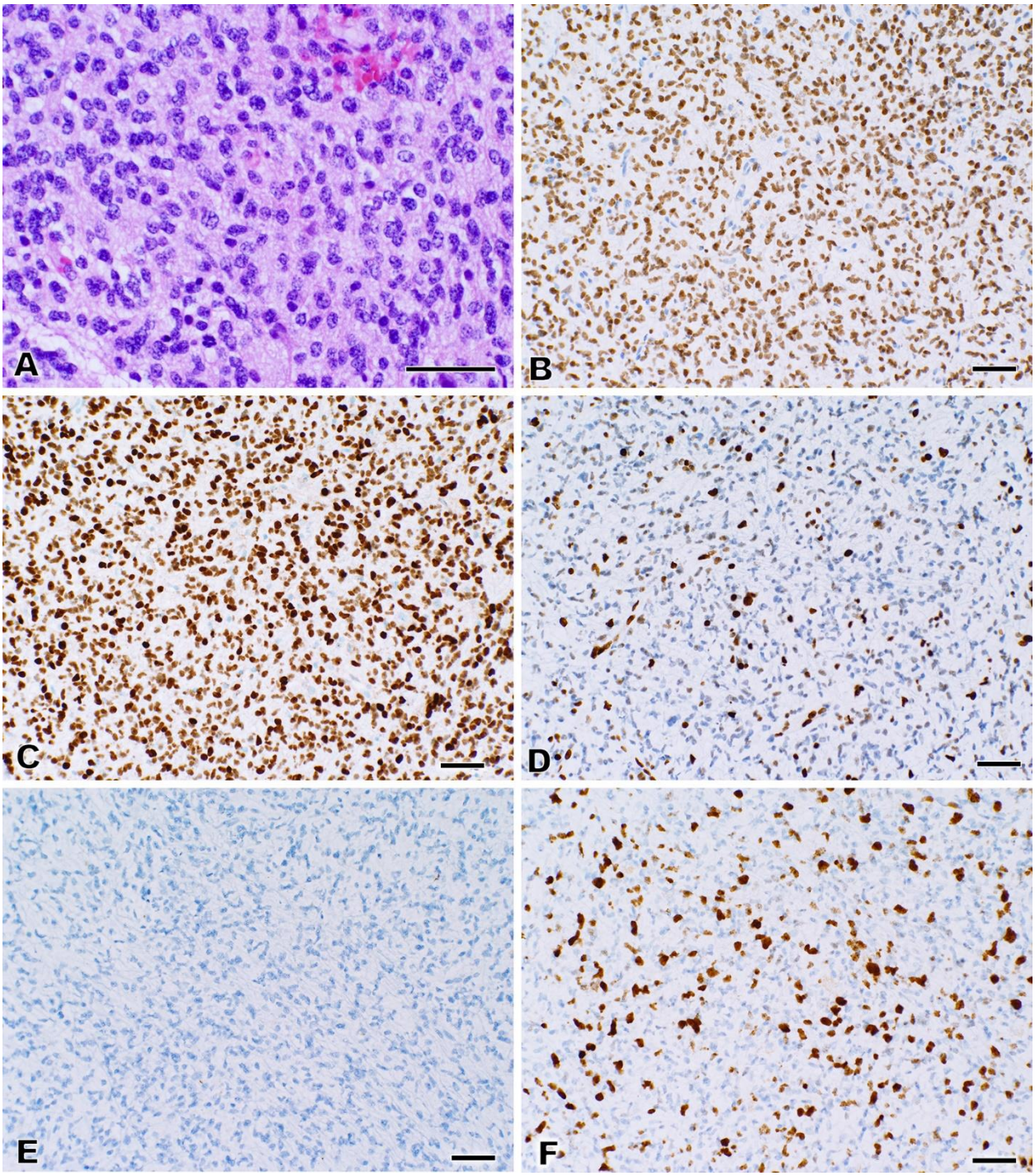

Figure 2. Diffuse Midline Glioma Pathology (A) H&E section demonstrating a glial neoplasm with high cellularity and mitoses, consistent with malignant glioma. Immunohistochemistry was positive for (B) Olig2 and (C) H3 K27M, a mutation-specific immunohistochemical stain characteristically positive in DMG. (D) Immunohistochemistry for H3K27me3, assessing for native trimethylation at the K27 position, was lost in tumor cells but retained in entrapped normal brain cells, including endothelial cells. (E) Tissue was negative for IDH1 R132H mutation. (F) Ki-67 proliferation index was elevated. Bar = 50 microns.

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