

Degos Disease with Multiple Infarcts Involving the Central Nervous System

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Case Summary

A previously healthy 4-year-old presented with fever and headaches. The child's clinical course included infarctions in the brain and spinal cord, subdural hematomas, and perforated gastric ulcers.

Brain biopsy was nonspecific, showing chronic vasculopathy with intraluminal fibrin thrombi in microvasculature. Skin lesions were recognized, and the diagnosis was confirmed with skin biopsy.

Imaging Findings

Brain MRI at initial presentation revealed a small, right-sided subdural fluid collection. An acute infarct with hemorrhagic components in the right frontal lobe was also visualized. MRI with a gadolinium-based contrast agent (GBCA) revealed abnormal leptomeningeal enhancement involving the right cerebral hemisphere, concerning for an infectious process (Figure 1).

Follow-up brain MRI two weeks later showed increased leptomeningeal enhancement with new infarctions in the right frontal lobe and the right-sided subdural collection (Figure 2). MRI also demonstrated new hemorrhagic infarctions in the right cerebral hemisphere, an increase in the diffuse leptomeningeal enhancement in the right cerebral hemisphere, enlarging bilateral subdural collections, and dural enhancement. GBCA-enhanced imaging revealed vessel wall enhancement involving a distal branch of the left middle cerebral artery, but no MRI abnormalities of the left cerebral hemisphere.

Diffusion weighted imaging of the spinal cord demonstrated multiple areas of reduced diffusion, representing acute infarcts.

A few months after initial presentation, MRI showed numerous chronic infarcts in both cerebral hemispheres with volume loss and bilateral subdural collections (Figure 3). The patient died from complications of the disease.

Diagnosis

Malignant atrophic papulosis (Degos disease)

Discussion

Malignant atrophic papulosis (Degos disease), is a rare dermatological disorder with organ involvement of the gastrointestinal (GI) tract and central nervous system (CNS).¹ Pathologically, it is a small-vessel vasculopathy targeting the skin, GI tract, and CNS. GI or CNS involvement portends a poor prognosis and high mortality.²

Neuroradiologic reports of this condition are rare. In addition to CNS and GI involvement, this child presented with progressive clinical deterioration from numerous infarcts over several months despite multiple treatments. The literature does suggest Degos disease may have a familial predisposition.³

Neuroimaging plays an important role in the work-up of Degos disease. Acute infarcts and subdural hematomas can develop over the course of the disease and are best visualized with MRI. GBCAs allow for visualization of leptomeningeal enhancement, which typically

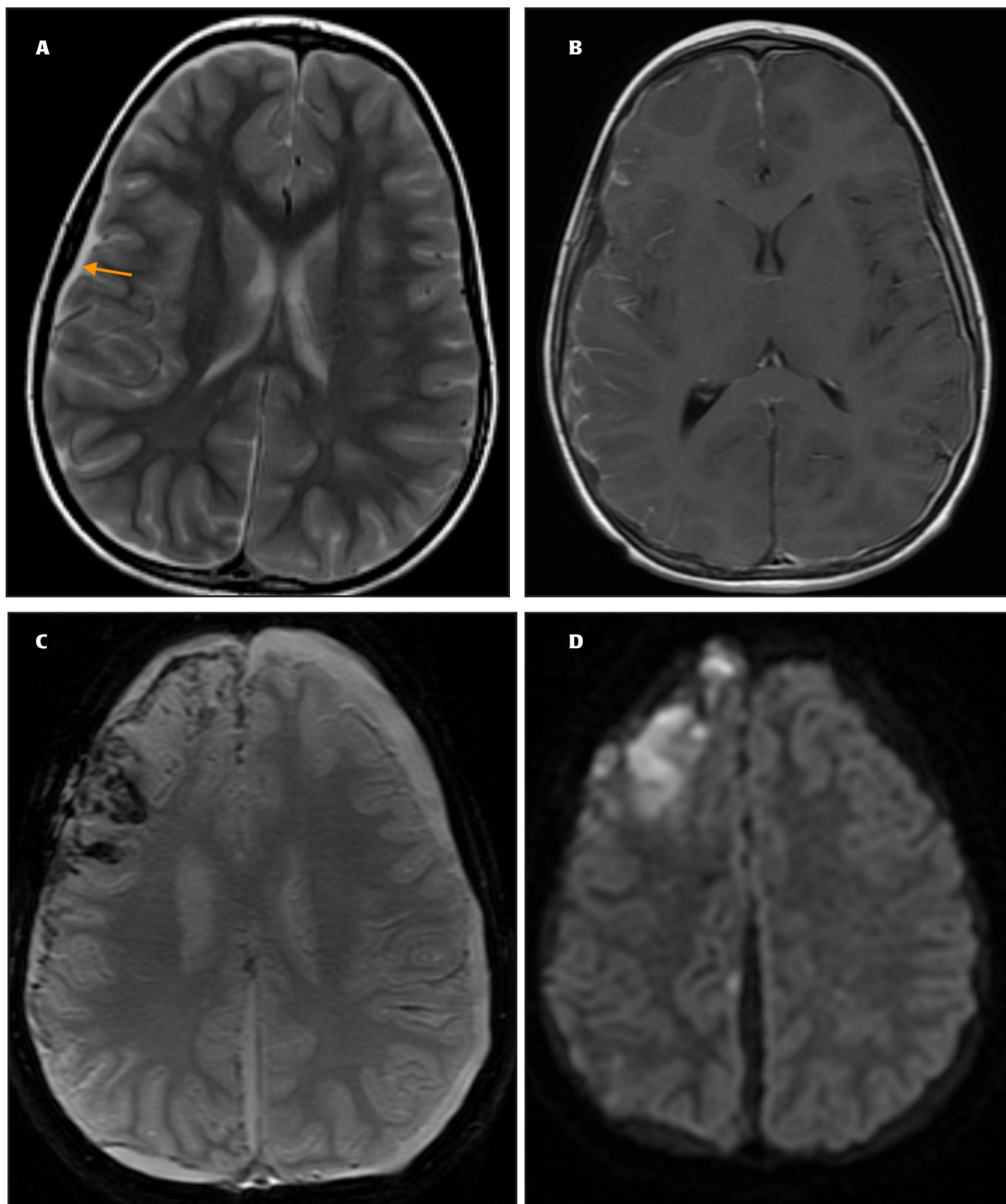


FIGURE 1. Initial presentation. (A) Axial T2 demonstrates a small right hyperintense subdural collection (arrow). (B) Postcontrast T1 imaging a few weeks later demonstrates leptomeningeal enhancement of the right cerebral hemisphere. (C) Susceptibility weighted imaging shows multiple areas of susceptibility representing hemorrhagic transformation of acute infarcts seen with (D) diffusion weighted imaging.

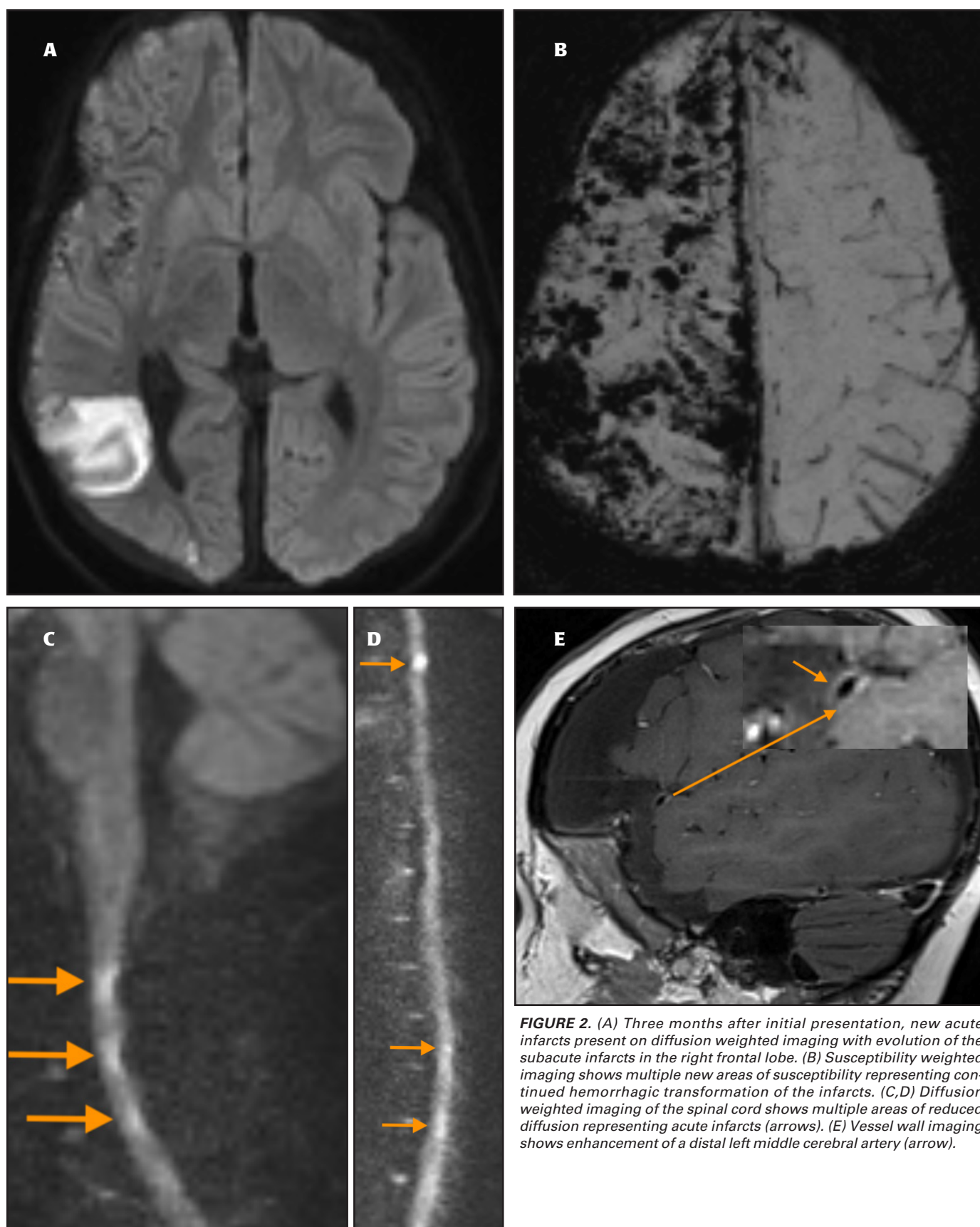


FIGURE 2. (A) Three months after initial presentation, new acute infarcts present on diffusion weighted imaging with evolution of the subacute infarcts in the right frontal lobe. (B) Susceptibility weighted imaging shows multiple new areas of susceptibility representing continued hemorrhagic transformation of the infarcts. (C,D) Diffusion weighted imaging of the spinal cord shows multiple areas of reduced diffusion representing acute infarcts (arrows). (E) Vessel wall imaging shows enhancement of a distal left middle cerebral artery (arrow).

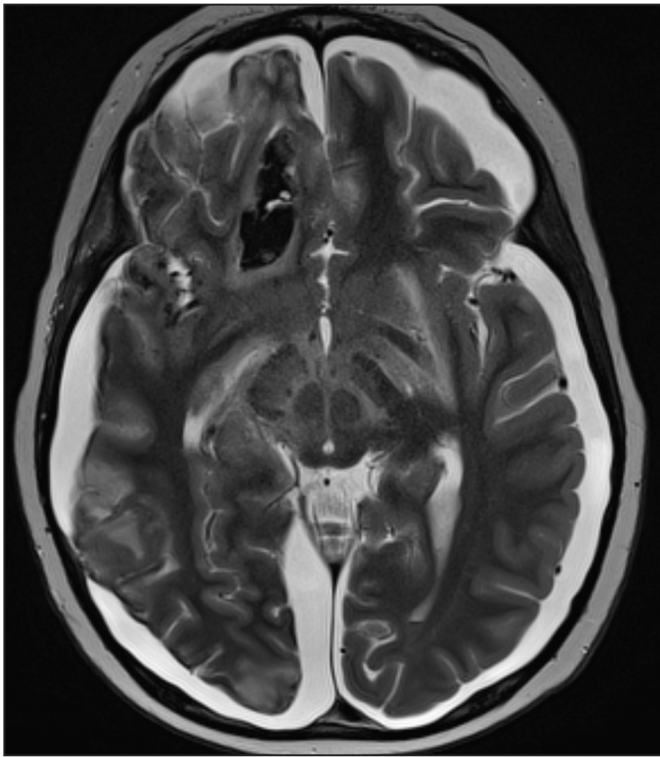


FIGURE 3. Axial T2 shows chronic infarcts in both cerebral hemispheres with volume loss and bilateral subdural collections appear 4 months after presentation.

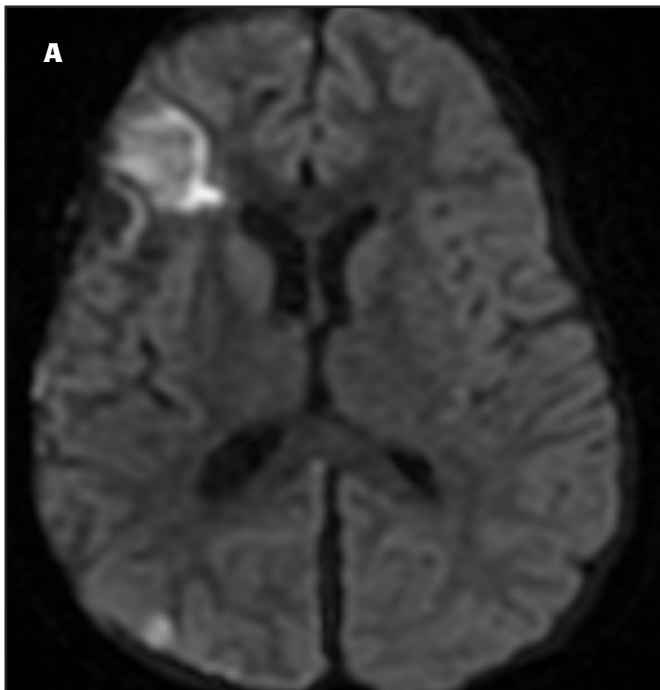


FIGURE 4. Summary of the imaging findings in Degos disease. (A) Diffusion weighted imaging shows reduced diffusion in the right frontal lobe representing an acute infarct (B) CT of the abdomen demonstrates free air (arrow) from gastrointestinal perforation. (C) A skin lesion of this patient with Degos disease, which is an important finding that can be biopsied for diagnosis.

correlates with the regions of CNS involvement. Vessel wall imaging is an advanced imaging technique, which allows for visualization of abnormalities along the vessel wall, with suppression of the luminal blood and cerebrospinal fluid signal.⁴ GBCA is required for determining vessel wall enhancement, which may represent inflammation or endothelial permeability in plaques.

In the setting of Degos disease, vessel wall enhancement may represent wall inflammation or possibly be related to small-vessel vasculopathy.⁵

Conclusion

Multifocal intestinal, cerebral, and cutaneous ischemic lesions in the setting of vessel wall enhancement should raise concern for Degos disease (Figure 4).

References

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