



Dysplastic Cerebellar Gangliocytoma

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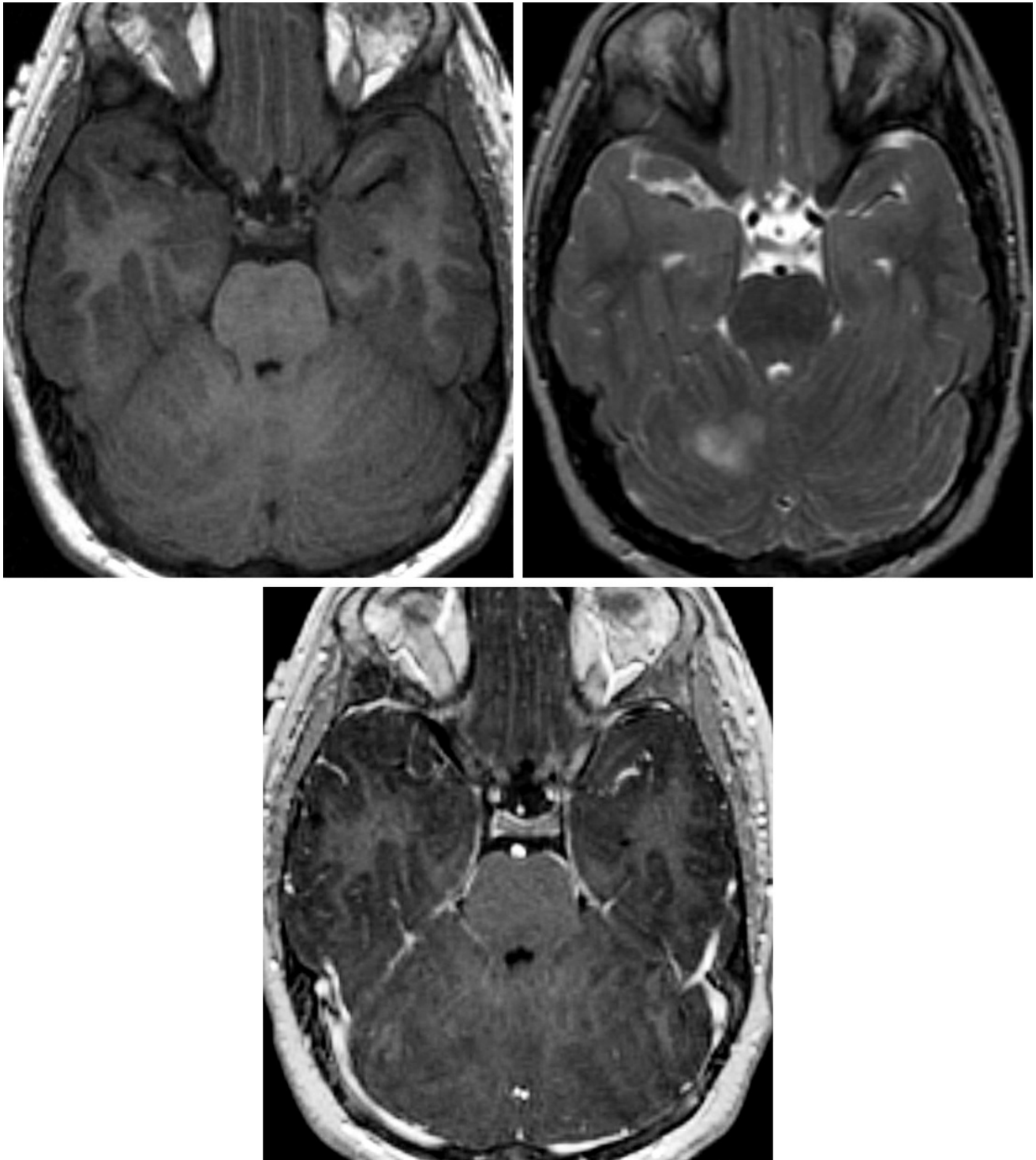


Figure 1: (Top Left) A disorganized appearance to the cerebellum is typical in this dysplastic cerebellar gangliocytoma, barely visible on T1WI. **(Top Right)** These lesions are often associated with variable degrees of

T2 hyperintensity and enhancement. (Bottom) This patient's lesion demonstrated no enhancement on T1WI postcontrast imaging.

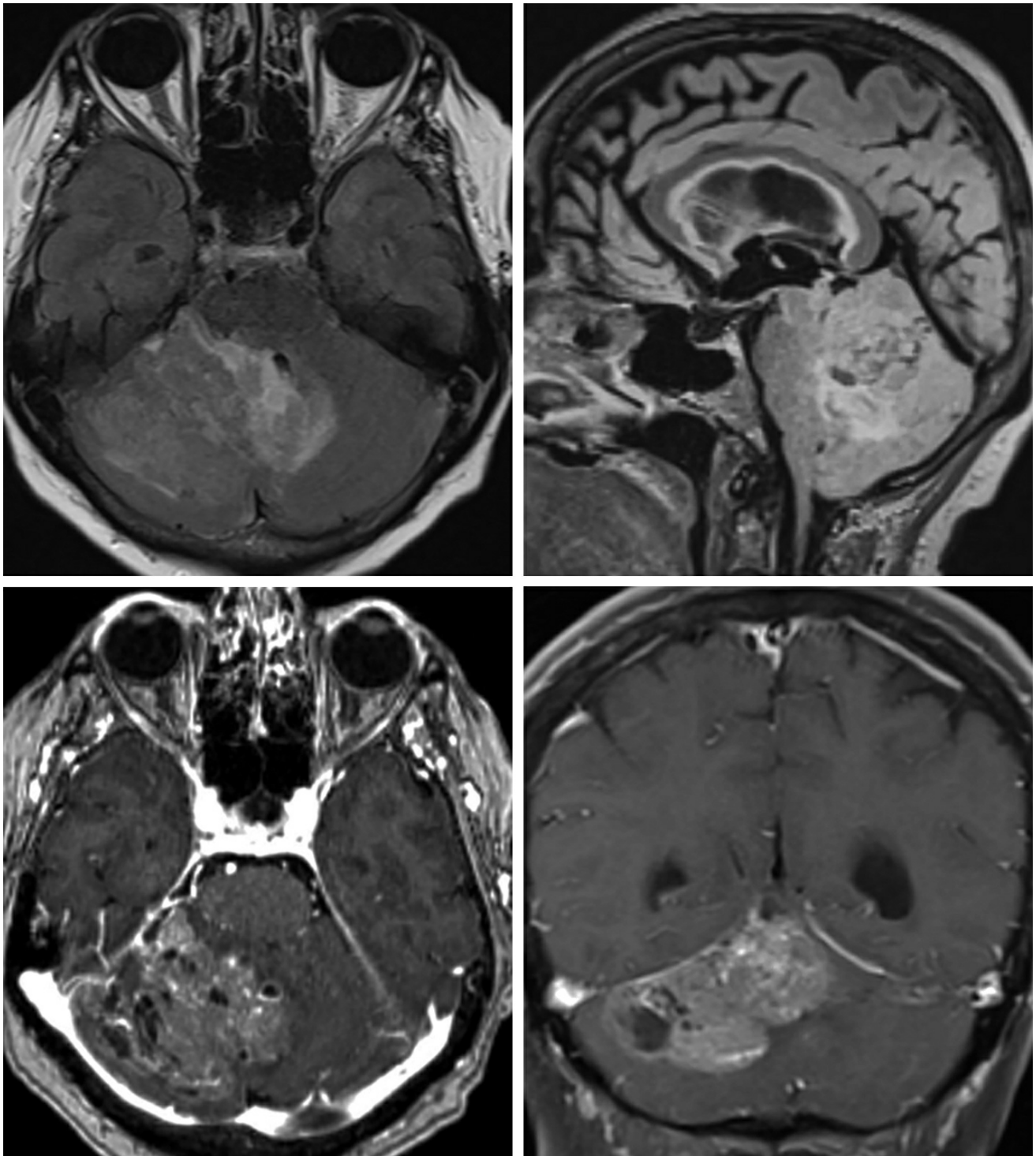


Figure 2: This heterogeneous infiltrative lesion centered in the right cerebellar hemisphere on FLAIR (top left, axial; top right, sagittal) demonstrates only mild associated edema and mild mass effect for the size of the lesion. There are small cystic changes and conspicuous enhancing vessels, and single hemispheric dominance but midline crossing that are typical of dysplastic cerebellar gangliocytoma/L'hermitte-Duclos disease on contrast-enhanced imaging

(bottom left, axial; bottom right, coronal). The degree of enhancement is moderate in this patient but highly variable in this type of tumor.

BASIC DESCRIPTION

- Also known as L'hermitte-Duclos disease (LDD), the neurologic manifestation of multiple hamartoma and neoplasia syndrome (MHAM) or Cowden syndrome (CS)
- CS + LDD = MHAM + LDD = COLD (Cowden-L'hermitte-Duclos syndrome), a neurocutaneous syndrome
- Benign cerebellar lesion of uncertain etiology

PATHOLOGY

- Benign cerebellar mass with thickened, irregular cerebellar folia
- Pathogenesis unclear: hamartomatous, neoplastic, or congenital
- WHO grade 1
- No malignant potential
- MHAM
 - Autosomal dominant inheritance of PTEN mutation
 - Hamartomas of skin, gastrointestinal and genitourinary tracts, mucosa, eye, and central nervous system
 - Increased risk of mucocutaneous tumors, thyroid adenomas, fibrocystic breast disease, and polyps
- Absence of cerebellar Purkinje cells, abnormal ganglion cells, and hypertrophic granule cell layer are microscopic features

CLINICAL FEATURES

- Any age (20–40 years old at presentation most common)
- No gender predilection
- Common presenting signs/symptoms
 - Increased intracranial pressure: headache, nausea, vomiting

- Cerebellar signs: ataxia, dysmetria, gait instability
- ±Clinical findings of MHAM/CS
- Treatment: surgical debulking ± CSF shunting

IMAGING FEATURES

- General
 - “Corduroy,” striated, or tigroid appearance of cerebellar hemisphere due to thickened, irregular folia
 - Most commonly unilateral cerebellar hemisphere involvement ± vermis
 - Variable size
 - Mass effect, tonsillar herniation, and hydrocephalus if large
 - Stable size or slow growth
- CT
 - Isodense to hyperdense lesion with thickened, irregular, and tigroid cerebellar folia
 - Variable enhancement on contrast-enhanced CT
- MRI
 - T1WI: hypointense to isointense
 - T2WI: hyperintense with hypointense to isointense cerebellar folia; hypointense vascular flow voids
 - DWI: hyperintense signal, which may represent “T2-shinethrough,” hypercellularity, or increased density of axons
 - T1WI+C: variable enhancement
 - MRS/MR perfusion: decreased NAA, Cho, and MI; increased relative cerebral blood volume (rCBV) and relative cerebral blood flow (rCBF) may be present

IMAGING RECOMMENDATIONS

- MRI without and with intravenous contrast including DWI and MR

spectroscopy; evaluate for MHAM/CS if findings of LDD are present (and vice versa) due to increased risk of other malignancy

For more information, please see the corresponding chapter in [Radiopaedia](#).

Contributor: Rachel Seltman, MD

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