Dysplastic Cerebellar Gangliocytoma (DCG)

Last Updated: October 1, 2018

Figure 1: A disorganized appearance to the cerebellum is typical in this dysplastic cerebellar gangliocytoma, barely visible on
T1WI (top row left). These lesions are often associated with variable degrees of T2 hyperintensity (top row right) and enhancement. This patient’s lesion demonstrated no enhancement on T1WI post contrast imaging (bottom).

Figure 2: This heterogeneous infiltrative lesion centered in the right cerebellar hemisphere on FLAIR (top row left – axial, top row 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 on contrast-enhanced imaging (bottom row left – axial, bottom row right – coronal). The degree of enhancement is moderate in this patient, but is highly variable in this type of tumor.

**Basic Description**

- Also known as Lhermitte-Duclos disease (LDD), the neurologic manifestation of multiple hamartoma and neoplasia syndrome (MHAM) or Cowden syndrome (CS)
- CS + LDD = MHAM + LDD = COLD (Cowden-Lhermitte-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, GI and GU tracts, mucosa, eye, and CNS
  - 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 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**
  - Iso- to hyperdense lesion with thickened, irregular, and tigroid cerebellar folia
  - Variable enhancement on contrast-enhanced CT

- **MRI**
  - T1WI: hypo- to isointense
  - T2WI: hyperintense with hypo- 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 IV 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.

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**DOI:** [https://doi.org/10.18791/nsatlas.v1.03.01.12](https://doi.org/10.18791/nsatlas.v1.03.01.12)

**References**


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