Medulloblastoma Masquerading as Bilateral Lhermitte-Duclos Disease

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Learning Objectives

By the conclusion of this session, participants should be able to:
1) Understand Lhermitte-Duclos disease and its association with Cowden syndrome
2) Describe the characteristic MRI/MR spectroscopy appearance of Lhermitte-Duclos
3) Consider the differential diagnosis of medulloblastoma in this context

Introduction

Lhermitte-Duclos disease (LDD; dysplastic cerebellar gangliocytoma) is a rare beingn cerebellar tumour with thickened cerebellar folia. A striated ('tiger-striped') appearance on T2W MRI is considered to be pathognomonic of LDD. Approximately 25% of cases are associated with autosomal dominant Cowden syndrome of multiple hamartoma-neoplasia. In a few published cases, characteristic MRI findings have been mimicked by a more agressive tumour, medulloblastoma (WHO grade IV).

Methods

We present a case where a 53 year old patient with a bilateral cerebellar tiger striped pattern on MRI refused biopsy and was kept under imaging surveillance for 2 years (with interval VP shunt insertion). Genetic screening for Cowden syndrome was negative. Reimaging at the time of clinical deterioration demonstrated severe diffuse posterior fossa swelling and a new area of focal enhancement. Biopsy of this enhancing lesion delivered a discrete tumour nodule which was found to be medulloblastoma on histology. Subsequent imaging 30 days post-op showed markedly reduced swelling in the entire posterior fossa (Figure 1). We performed a review of the literature identifying similar cases and their clinical/imaging profile.

Results

Six patients (Table 1) were identified in the literature (age 18 months to 28 yrs; 3 cases were bilateral) with imaging findings consistent with LDD but subsequent histology confirming WHO grade IV medulloblastoma. MR Spectroscopy was identified as critical in excluding LDD in 4 cases.

Conclusions

In the context of patients with MRI features consistent with LDD, a low threshold for biopsy or surgical resection should be adopted include bilateral disease, negative genetic screen for Cowden syndrome or non-diagnostic/equivocal MR spectroscopy result.

References

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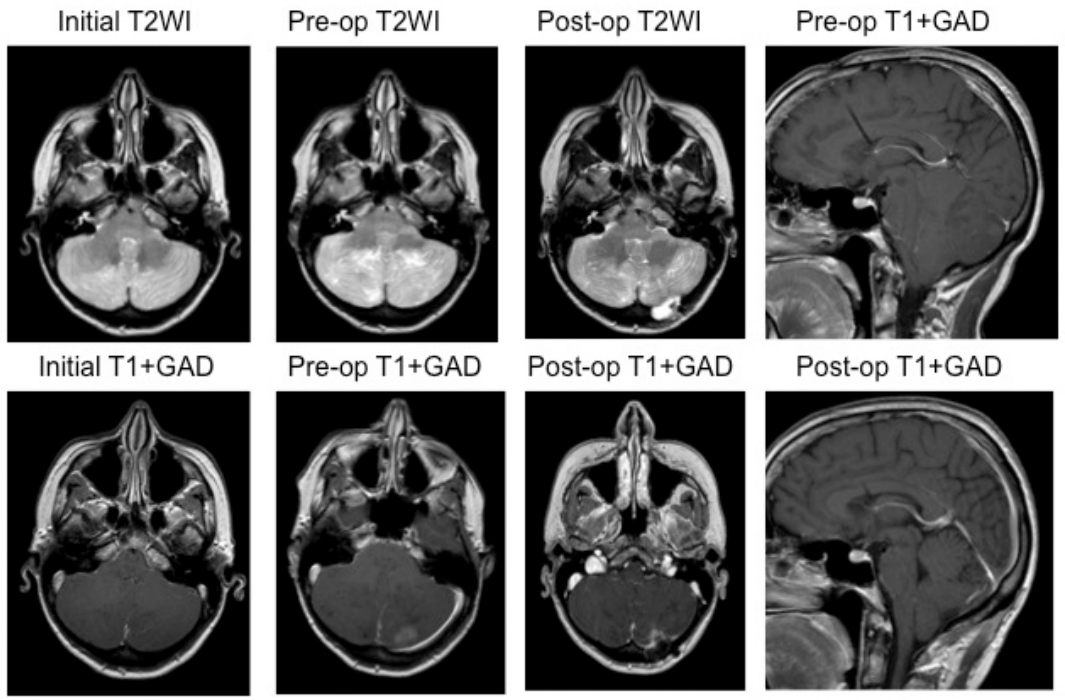
Table 1

Study	Age	Sex	Presentation	MRI	MR Spectroscopy	Management	Histological diagnosis
Savardekar et al., 2012 [4]	28 y	M	Raised ICP	Tiger-striped appearance. No enhancement. No restricted diffusion.	Increased choline. Cho:Cr and Cho:NAA ratio Mitotic pathology	VP shunt Midline suboccipital craniectomy and decompression of left cerebellar hemisphere	Medulloblastoma WHO grade IV
	19 y	M	Raised ICP	Tiger striped. Small enhancing nodule in midline.	Mitotic pathology	Midline suboccipital craniectomy and excision of enhancing nodule	Medulloblastoma WHO grade IV
Mittal et al., 2009 [5]	19 y	M	Raised ICP	Right cerebellar mass. Hypointense T1, isointense to mildly hyperintense T2. Mild patchy enhancement and restricted diffusion.	-	Not specified	Medulloblastoma
Douglas-Akinwande et al., 2009 [6]	27 y	F	Raised ICP	Folia pattern. No enhancement or restricted diffusion.	-	Subtotal surgical resection	Medulloblastoma WHO grade IV
Kamble 2012 [3]	3 y	-	Ataxia	Hyperintense right cerebellum with enlargement, cerebellar folia well preserved. Hyperintense vermis. No enhancement. Repeat MRI in 1 month showed increased size of vermis but no enhancement. No restricted diffusion.	Elevated choline and reduced NAA from vermis	Not specified	Medulloblastoma with infiltration into cerebellum
Chen et al., 2002 [7]	18 months	M	Developmental delay and ataxia	Tiger-striped appearance. Enhancement with gadolinium.	-	Suboccipital craniotomy and excision of lesion followed later by VP shunt	Medulloblastoma

Summary of published case reports of medulloblastoma mimicking LDD

Figure 1



MRI appearances at presentation, pre-operatively and at 30 days post-op.