Teaching NeuroImages: Wishbone pattern of iron accumulation

A characteristic imaging sign in GM1 gangliosidosis

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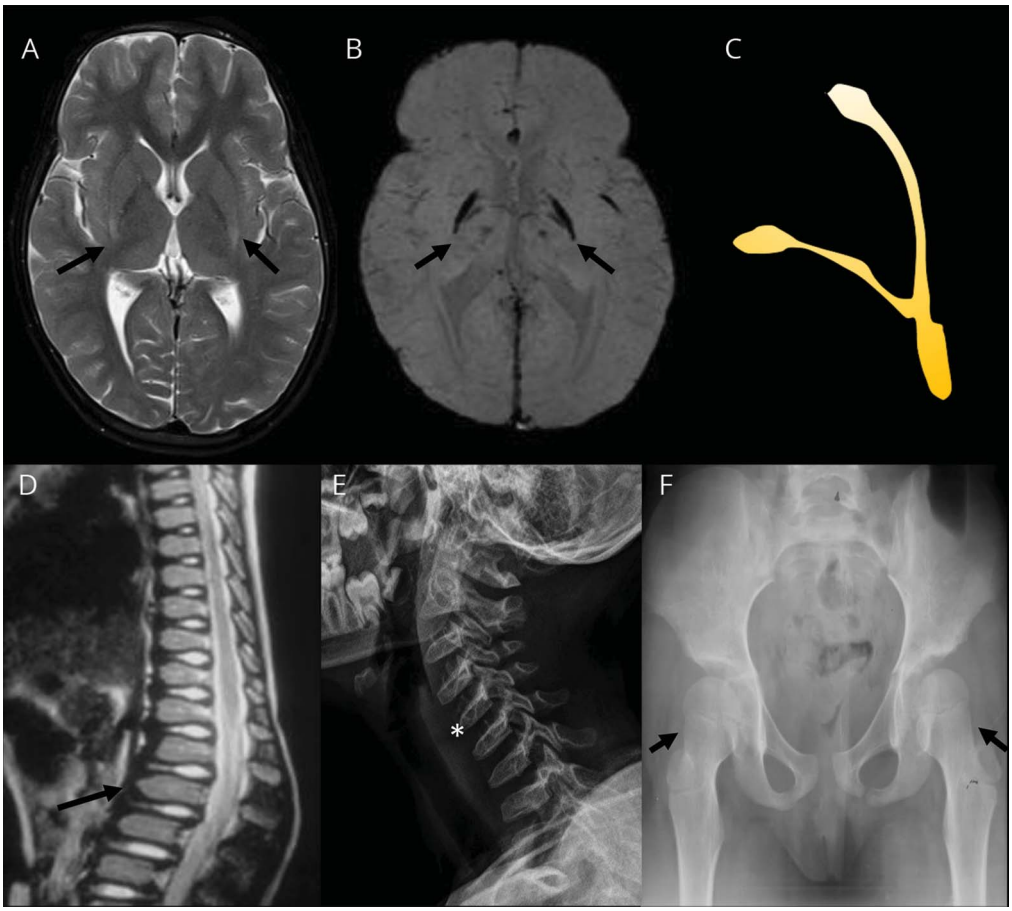
Figure Axial T2-weighted susceptibility-weighted imaging (SWI), animation, and sagittal T2-weighted spine, cervical, and pelvis radiograph



Axial T2-weighted imaging shows (A) posterior putamen hyperintensity and volume loss. SWI (B) and animation (C) shows blooming in globus pallidus in a wishbone pattern. Sagittal T2-weighted (C) spine and (D) cervical radiographs show platyspondyly (asterisk) with beaking of vertebra (arrow). Pelvis radiograph (D) shows bilateral coxa valga.

An 8-year-old girl, with no relevant developmental or family history, presented with progressive orolingual and limb dystonia since 3 years of age. MRI brain showed bilateral posterior putamen volume loss and hyperintensity. Susceptibility-weighted images showed globus pallidus blooming in characteristic wishbone pattern with medial and lateral parts forming the forked ends (figure). MRI spine showed features of dysostosis with platyspondyly and vertebral

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Prateek Malik, MD	Radiodiagnosis, Christian Medical College, Vellore	Concept, image interpretation and preparation, write-up of manuscript
Sniya Valsa Sudhakar, DNB	Radiodiagnosis, Christian Medical College, Vellore	Concept, image interpretation and preparation, critical revision
Karthik Muthusamy, MD	Neurology, Christian Medical College, Vellore	Clinical input, critical revision for intellectual content
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beaking. β-Galactosidase assay was low and genetic workup revealed compound heterozygous pathogenic mutation in *GLB1* gene. Combination of putaminal finding and wishbone pattern of iron deposition is highly diagnostic of late onset/type 3 GM1 gangliosidosis[1,2] and helps differentiate from neurodegeneration with brain iron accumulation.

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Disclosure

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