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NEUROLOGICAL PICTURE

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Tigroid and leopard skin pattern of dysmyelination in metachromatic leucodystrophy

A 3 year old female child, born of consanguineous parentage and full term normal delivery, developed psychomotor regression. When examined one year later, she exhibited no tracking of visual or auditory stimuli and optic fundi revealed mild pallor. She could produce no meaningful speech. She had drooling of saliva from the mouth, bilateral pyramidal signs, and was doubly incontinent. There was no family history of similar illness. Cranial MRI scan revealed hypointense radially oriented stripes and dots within the hyperintense cerebral white matter on T2WI (panels A and B). These dots were iso to hyperintense on T1WI (panel C). This pattern of dysmyelination resembled the skin of tiger (radial

stripes) and leopard (dots). A diagnosis of metachromatic leucodystrophy (MLD) was confirmed by decreased activity of arylsulfatase A in leucocytes.

Besides MLD, tigroid and leopard skin pattern of dysmyelination has also been described in Pelizaeus-Merzbacher disease and globoid cell leucodystrophy.^{1,2} Recognition of this pattern could provide valuable diagnostic clue in the proper clinical context. In the MRI of present patient, the visible dots/radial stripes with shortening of both T2 and T1 values (hypointense on T2WI and hyperintense on T1WI) probably represented areas of myelin sparing or lipid storage. This observation was confirmed by MRI and histopathological correlative study of brain in MLD.²

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(A) Hypointense radial stripes seen within the hyperintense cerebral white matter (resembling tiger skin) on T2 weighted axial MRI. (B) Hypointense dots resembling leopard skin seen in the same sequence at the level of centrum ovale. (C) Iso to hyperintense dots seen in the cerebral white matter on T1 weighted axial imaging.