

Menkes Kinky Hair Disease

A 10-month-old boy, born to non-consanguineous parents, presented with failure to thrive and tonic spasms. Sparse, steely hairs (**Fig. 1**), depressed nasal bridge, chubby cheeks and delayed motor development were the findings on examination. Serum copper was 14.37 mg/dL and



FIG. 1 Brittle, tangled, sparse, ivory gray kinky hairs.

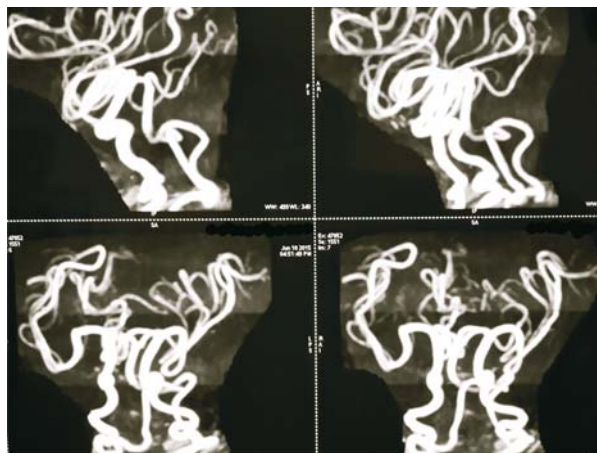


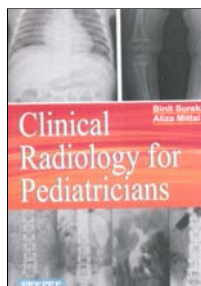
FIG. 2 MR angiography showing tortuosity of intracerebral vessels.

ceruplasmin was 9.58 mg/dL. MR angiography reveals tortuosity of cerebral vessels (**Fig. 2**).

Menkes Kinky hair disease is an X-linked recessive trait caused by mutations in the ATP7A gene leading to disturbed copper metabolism contributing to low serum copper and ceruplasmin. It is characterized by brittle, tangled, sparse, steely or kinky hairs that are often white, ivory, or gray in color with easy pluckability. Hair shaft abnormalities include pili torti, trichorrehexis nodosa, trichoclasia, and trichoptilosis.

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BOOK REVIEW



Clinical Radiology for Pediatricians

BINIT SUREKA AND ALIZA MITTAL
Peepee Publishers and Distributors
(P) Ltd.,
New Delhi: 110 002, India.
Pages: 206; Price: Rs 295/-.

This pocket-sized book is a joint effort of a young Radiologist and a Pediatrician, and provides a synopsis of the commonly encountered clinical conditions and their imaging appearance in the children. It presents the relevant information in an

attractive manner, easily comprehensible to the readers. Pertinent imaging findings are well covered for quick reference for the students along with the important differentials of the imaging diagnosis. The most attractive aspect of the book is the brainstorming part which quizzes the readers with the classical imaging appearances. This book is a good attempt to familiarize the pediatricians with the 'blacks and whites' of Radiology to add color to the clinical perspective in providing a better diagnosis.

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