2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

CASES

Wilson's disease is an inborn error of copper metabolism that is characterized by deficiency of ceruloplasmin, the serum transport protein for copper.

Copper is collected in the **liver, and after hepatic binding sites are saturated, it is released.**

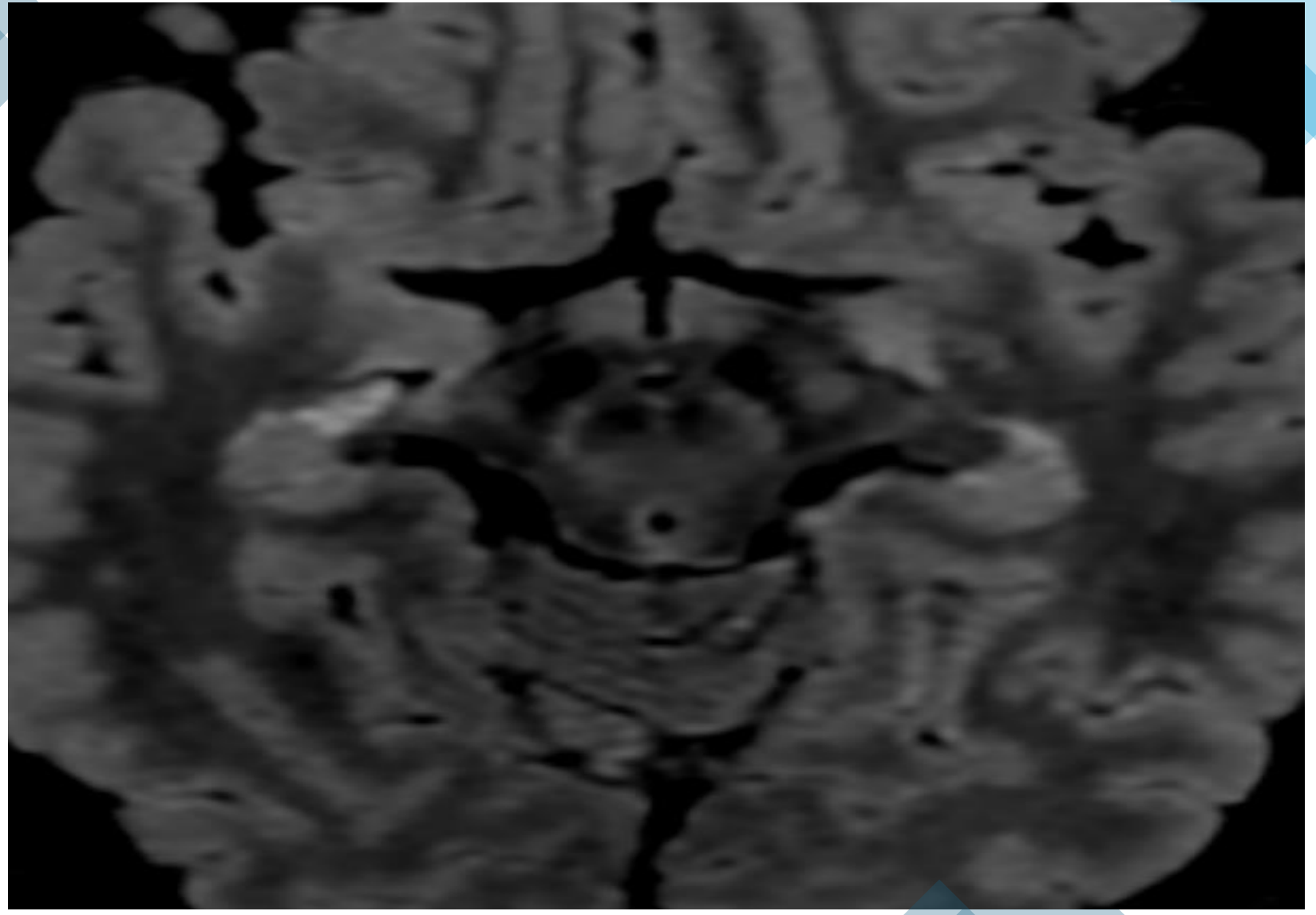
Systemic disease then develops and there is **abnormal accumulation of copper in the brain, particularly in the putamen and globus pallidus.**

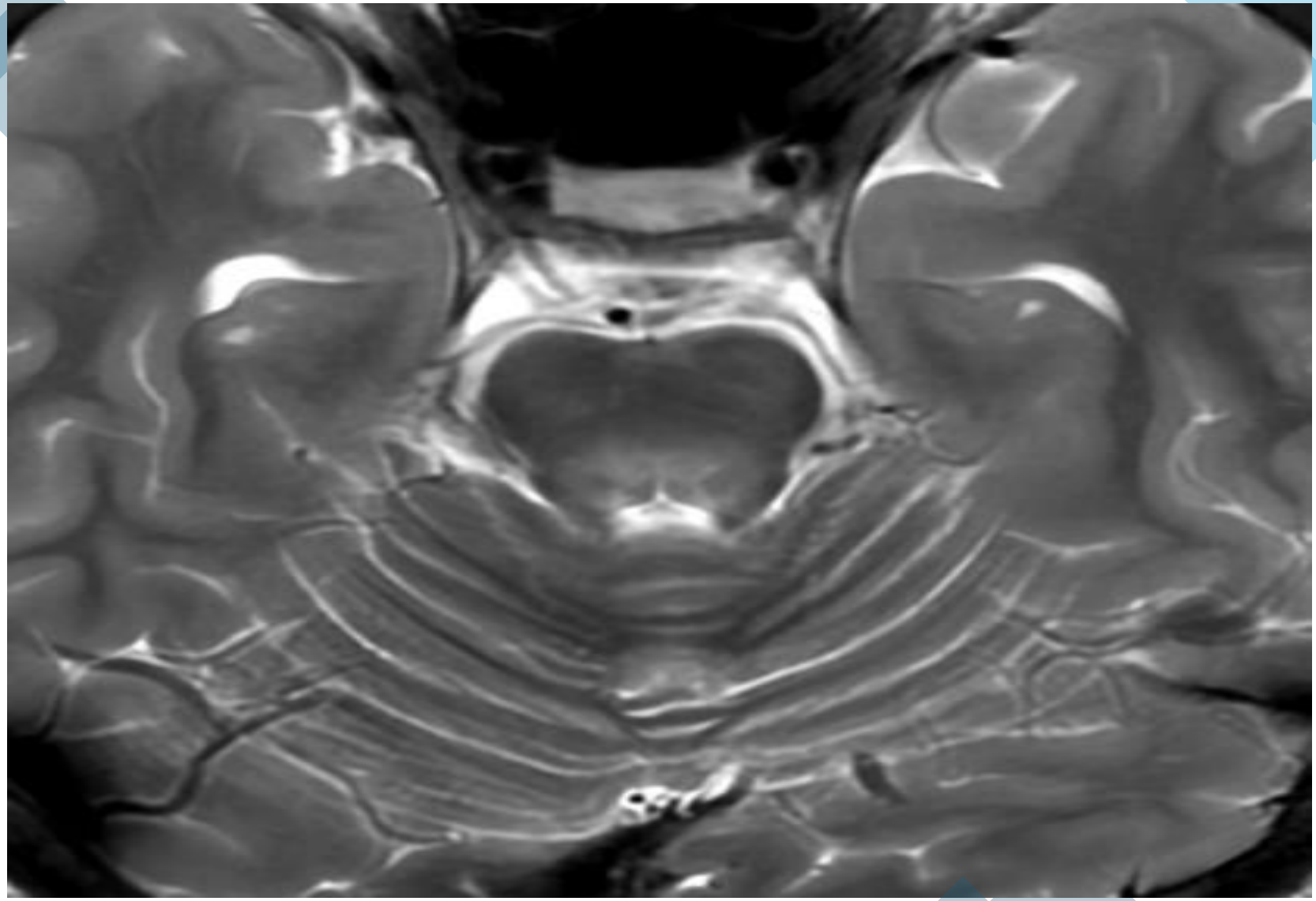
The neurologic manifestations associated with **Wilson's disease are understood to be secondary to buildup of cerebral copper at levels adequate to destroy nerve cells.**

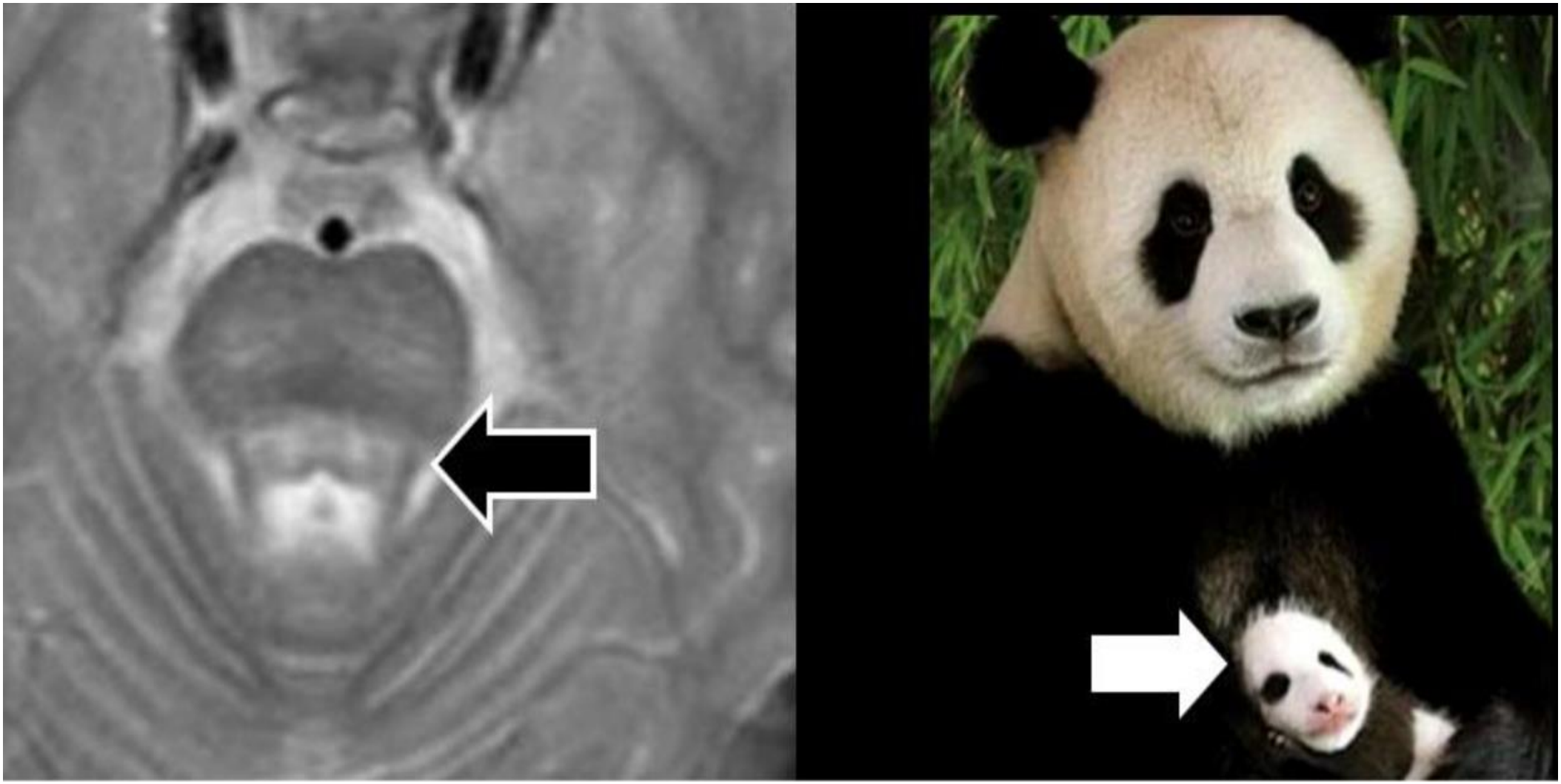
Edema, necrosis, and spongiform degeneration are the histopathological changes that are observed in Wilson's disease involving the brain.

MRI not only provides biochemical information on heavy metal distribution in brain tissue but also gives an insight into the pathologic and anatomic correlates of clinical signs and symptoms in Wilson's disease.

Interval changes seen on follow-up MR imaging have good correlation with clinical symptoms and can be useful in evaluating the clinical response to treatment of children with Wilson's disease.







IMAGING DIFFERENTIALS:

Leighs disease
CJD
HIE
Extrapontine myelinolysis.