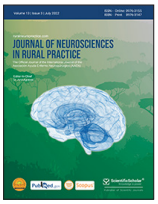
ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://ruralneuropractice.com

Journal of Neurosciences in Rural Practice



Images

Wing beating tremors in Wilson's disease: An important clinical clue

Alvee Saluja[1], Prerna Sinha[1], Shahbaz Anees[1], Jyoti Verma[1], L. H. Ghotekar[2], Rajinder K. Dhamija[3]

[1]Department of Neurology, Lady Hardinge Medical College, [2]Department of Medicine, Lady Hardinge Medical College and Associated Hospitals, [3]Institute of Human Behaviour and Allied Sciences, New Delhi, India.

A 30-year-old gentleman complained of tremors in his left hand for the past 2 years. The tremors occurred while holding objects and were absent at rest. Due to the tremors, the patient often complained of spillage of water from his glass such that he started drinking water from a half-filled glass. Over the next 6 months, the tremors gradually progressed to involve the right hand. One year after symptom onset, he noticed tremors in his left foot which gradually progressed to involve the right lower limb in the past 3 months. For the past 8 months, the patient complained of progressively increasing slowness in daily activities along with slowness in walking. He also complained of slurring of speech for the past 2 months. Apart from one episode of jaundice that occurred 5 months ago, the rest of the history was insignificant. He had a history of alcohol intake and tobacco chewing for the past 10 years but had abstained from alcohol for the past 3 years. There was no history of motor weakness, sensory impairment, cranial nerve palsies, headaches, prior strokes, imbalance while walking, falls, gait freezing, decreased perception of smell, psychiatric disturbances, hallucinations, delusions, fever, rash, joint pains, oral or genital ulcers, dry eyes/dry mouth, and Raynaud's phenomenon. The family history was negative, and there was no history of chronic antipsychotic or alternative medication use.

On examination, the patient had bilateral high-amplitude, low-frequency, asymmetric (left > right) postural tremors in bilateral upper limbs (wing beating tremors) [Video 1]. On ocular examination, he had Kayser-Fleischer rings in both eyes that were confirmed on slit lamp examination [Figure 1a and b]. He had mild symmetrical bradykinesia in all four limbs and mixed dysarthria (spastic + extrapyramidal). The rest of the neurological examination was normal. The complete hemogram showed mild thrombocytopenia with normal hemoglobin and total leukocyte counts. His liver and renal function tests were also normal. The serum ceruloplasmin levels were markedly low – 9.70 mg/dL (reference – 20–60 mg/dL) and 24-h urinary copper levels were elevated (480 mg/day; reference – 3–50 mcg/day). Ultrasound abdomen showed coarsened liver echotexture suggestive of

Video 1: Asymmetric Wing beating postural tremors (left > right) are best demonstrated with the arms abducted at 90°, elbows flexed, and palms facing the floor.

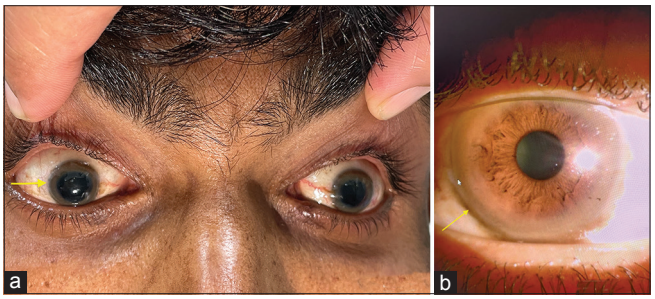


Figure 1: (a) Bilateral Kayser-Fleischer (KF) ring (yellow pointer) completely circumscribing the cornea. (b) Slit lamp examination of the right eye showing a brown KF ring (yellow pointer) at the sclera-corneal junction.

***Corresponding author:** Rajinder K. Dhamija, Institute of Human Behaviour and Allied Sciences, New Delhi, India. dhamijark@gov.in

Received: 13 May 2023 **Accepted:** 08 June 2023 **EPub Ahead of Print:** 24 June 2023 **Published:** 16 August 2023 **DOI:** 10.25259/JNRP_264_2023

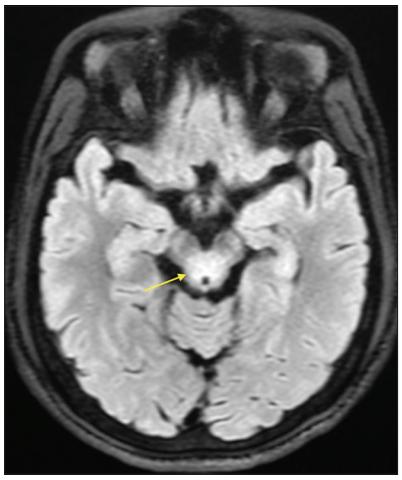


Figure 2: Magnetic resonance imaging brain axial fluid attenuated inversion recovery sequence showing hyperintensity in the midbrain tegmentum and the periaqueductal region along with hypointensity in the bilateral substantia nigra.

Video 2: A reduction in the severity of the patient's tremors was noted 2 months after initiating D-Penicillamine treatment during the follow-up visit.

liver parenchymal disease. Magnetic resonance imaging brain showed symmetrical T2/fluid attenuated inversion recovery hyperintensities in the periaqueductal region, midbrain tegmentum, and pontine tegmentum with hypointensity in bilateral red nuclei and substantia nigra [Figure 2]. Two months after starting D-penicillamine, the 24 h urinary copper excretion had decreased to 164.4 mcg/day, and his tremors had improved such that he was able to independently feed and dress without difficulty [Video 2].

Wilson's disease (WD), an autosomal recessive genetic disorder, causes the deposition of copper in the liver, cornea, and central nervous system due to a copper-transporting ATP7B gene mutation.[1] Wing-beating tremors are characteristically found in WD due to dentatorubrothalamic pathway involvement, and their presence should alert the neurologist to the possibility of this disease.[2] The tremor is best demonstrated by keeping the arms abducted and elbows flexed with the palms facing the floor.[3] Thus, astute clinical examination of this tremor pattern is key in suspecting WD, so that unnecessary investigations are avoided, and treatment can be promptly initiated.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dev S, Kruse RL, Hamilton JP, Lutsenko S. Wilson disease: Update on pathophysiology and treatment. Front Cell Dev Biol 2022;10:871877.
2. Maciel R, Portela DC, Xavier-Souza G, Maia D, Cardoso F. Improvement of "wing-beating" tremor in Wilson's disease with high dose of zolpidem: A case report. Mov Disord Clin Pract 2019;6:608-9.
3. Mahajan R, Zachariah U. Wing-beating tremor. N Engl J Med 2014;371:e1.

How to cite this article: Saluja A, Sinha P, Anees S, Verma J, Ghotekar LH, Dhamija RK. Wing beating tremors in Wilson's disease: An important clinical clue. J Neurosci Rural Pract 2023;14:547-8.