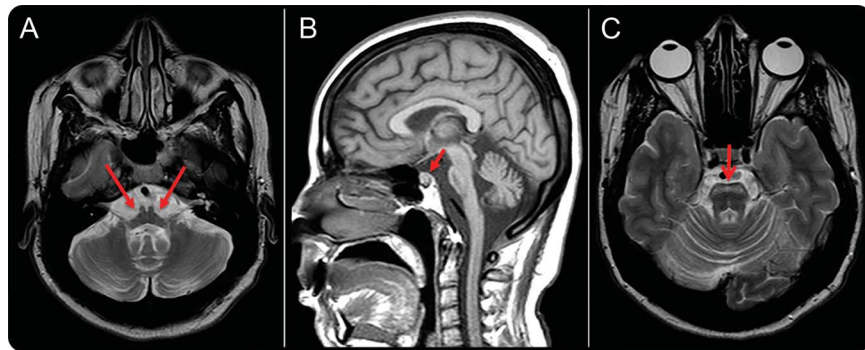


Teaching NeuroImages: MRI of brain findings of Wolfram (DIDMOAD) syndrome

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Figure 1 Brain MRI findings of a 31-year-old woman with Wolfram syndrome

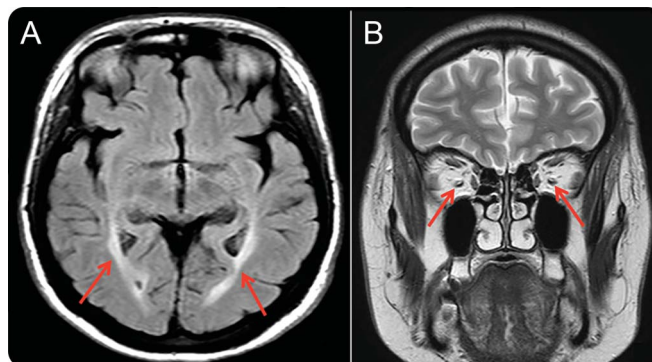


Axial T2-weighted image (A) and sagittal T1-weighted image (B) demonstrate atrophy of brainstem. Note absence of neurohypophyseal "bright signal" on sagittal T1 image (B). T2-weighted image (C) shows atrophy of cerebellum and hyperintense signal at ventral part of the pons.

A 31-year-old woman was diagnosed with type 1 diabetes mellitus (DM) at age 5 years and subsequently with hypothyroidism at age 16 years. She developed progressive visual loss at age 19 years and progressive hearing loss at age 28 years. She was clinically and radiologically diagnosed with Wolfram syndrome

(figures 1 and 2). Wolfram syndrome, first described in 1938, is a rare autosomal recessive disorder.¹ It features diabetes insipidus (DI), DM, optic atrophy (OA), and deafness (D) (DIDMOAD).^{1,2} It is caused by a mutation in the *WFS1* gene that encodes wolframin, a transmembrane protein of pancreatic β cells.¹

Figure 2 Optic nerve and optic tract findings



Axial fluid-attenuated inversion recovery image (A) demonstrates increased signal intensity in the bilateral peritrigonal areas/optic tracts. Coronal T2-weighted image (B) shows thinning/atrophy of bilateral optic nerves (arrows).

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The life expectancy of patients diagnosed with this syndrome is about 30 years.

AUTHOR CONTRIBUTIONS

Rahsan Gocmen interpreted neuroradiologic aspects of this article including formatting of the figure and other professional comments, assisted in drafting, and revised the manuscript for intellectual content. Ezgi Guler wrote the draft, collected the information, compiled the manuscript, and assisted in drafting and preparing the manuscript for publication.

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REFERENCES

1. Scolding NJ, Kellar-Wood HF, Shaw C, Shneerson JM, Antoun N. Wolfram syndrome: hereditary diabetes mellitus with brainstem and optic atrophy. *Ann Neurol* 1996;39:352–360.
2. Ito S, Sakakibara R, Hattori T. Wolfram syndrome presenting marked brain MR imaging abnormalities with few neurologic abnormalities. *AJNR Am J Neuroradiol* 2007;28:305–306.

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