

Cervical Spondylotic Myelopathy Secondary to Ochronotic Vertebral Arthropathy

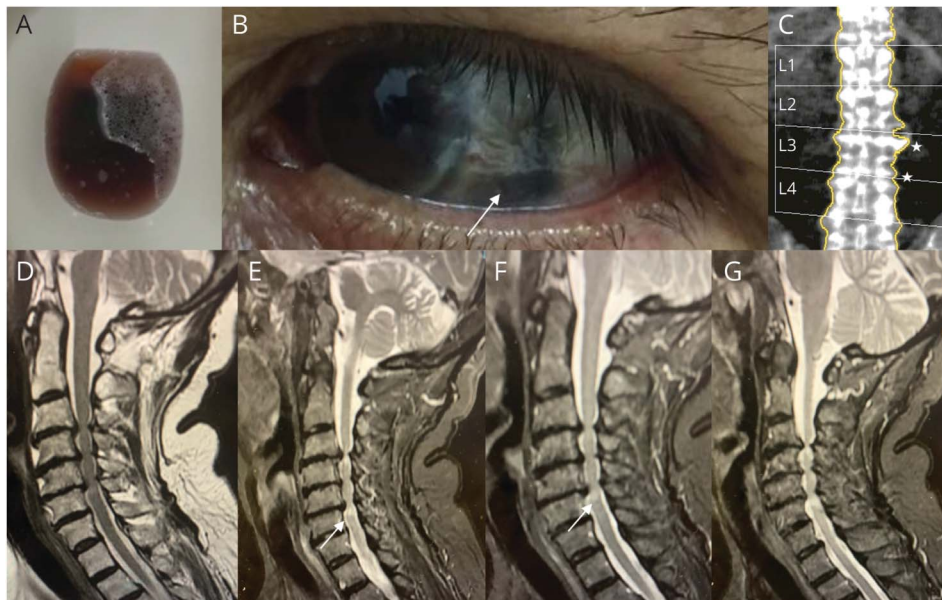
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Figure Examination and Neuroimaging in Alkaptonuria



(A) Dark urine and (B) dark sclerae (white arrow). (C) Bone densitometry disclosing osteopenia and increased density in intervertebral lumbar disks. (D-G) Sagittal spine MR imaging showing severe osteoarthropathy, disk herniation at C3-C6 levels, cervical stenosis and hyperintense spine change in T2-weighted sequence (white arrow).

A 39-year-old man presented with progressive gait disturbance and urinary incontinence. Medical history disclosed nephrolithiasis, dark urine, osteopenia and osteoarthritis for 15 years. Examination disclosed crural spastic paraparesis with sensory level at T1 and hyperpigmentation of the sclerae (figure). Neuroimaging studies disclosed cervical spondylotic myelopathy and several discs prolapses. Homogentisic acid levels by HPLC were high in 24-hour urine.

Alkaptonuria or inherited ochronosis is an autosomal recessive inherited metabolic disorder involving mainly the joints, cardiovascular and genitourinary systems. Dark urine and sclerae and diffuse osteoarthropathy represent clues for clinical suspicion.^{1,2} Early adult-onset cervical spondylotic myelopathy represents a severe neurologic complication.^{1,2}

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix Authors

Name	Location	Contribution
Wladimir Bocca Vieira de Rezende Pinto, MD, MSc	Federal University of São Paulo (UNIFESP), São Paulo, SP, Brazil	Case report project (conception, organization, execution); Writing of the first draft, review and critique
Igor Braga Farias, MD	Federal University of São Paulo (UNIFESP), São Paulo, SP, Brazil	Case report project (conception, organization, execution); Writing of the first draft, review and critique
Bruno de Mattos Lombardi Badia, MD	Federal University of São Paulo (UNIFESP), São Paulo, SP, Brazil	Case report project (conception, organization, execution); Writing of the first draft, review and critique

Appendix (continued)

Name	Location	Contribution
José Marcos Vieira de Albuquerque Filho, MD	Federal University of São Paulo (UNIFESP), São Paulo, SP, Brazil	Case report project (conception, organization, execution); Writing of the first draft, review and critique
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Acary Souza Bulle Oliveira, MD, PhD	Federal University of São Paulo (UNIFESP), São Paulo, SP, Brazil	Case report project (conception); Review and critique

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