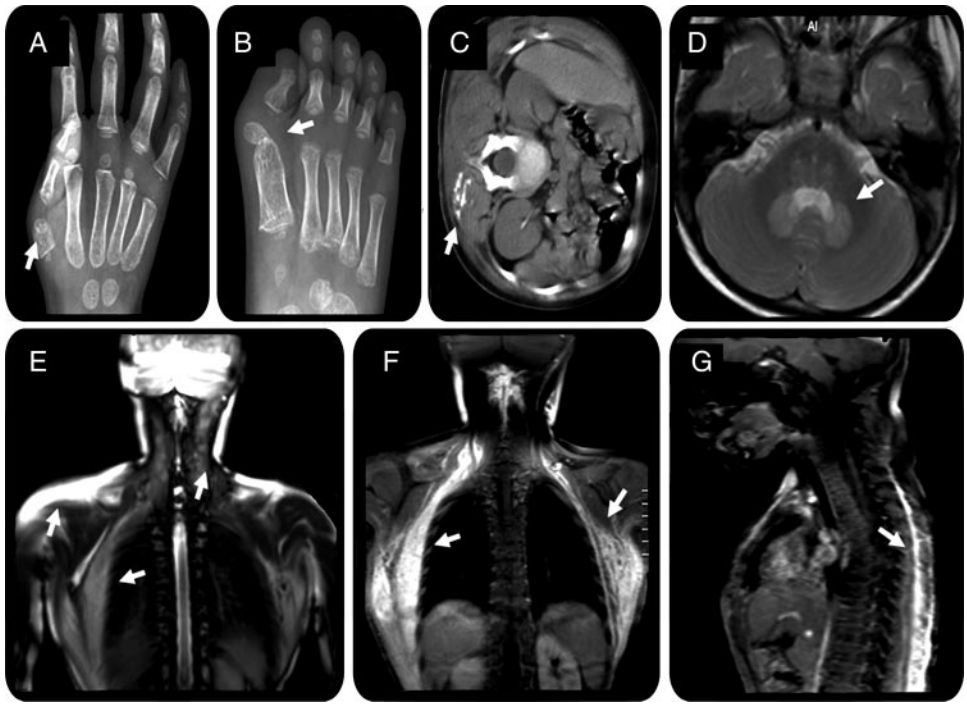
Teaching Neuro*Images*: MRI in fibrodysplasia ossificans progressiva

Shiva Kumar R., MD
Keerthiraj B., MD
Chandrasekhran Kesavadas, MD

Address correspondence and reprint requests to Dr. Shiva Kumar R., Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum 695011, Kerala, India
drshivakumar1995@yahoo.co.in

Figure Radiologic findings



Skeletal X-rays show clinodactyly, short thumb, and metacarpals (A), and hallux valgus with short phalanx (B). The CT scan shows calcification around paraspinal muscles (C). T2-weighted MRI of the brain shows dentate nucleus hyperintensity (D); STIR-coronal MRI (E), T2-weighted (F), and postcontrast (G) show increased signal and contrast enhancement in the muscles and fascial planes of the thoracic wall and paraspinal muscles.

A 3-year-old boy presented with abnormal neck posturing and rapidly growing lumps over the neck. The presence of typical malformed great toes and ectopic calcification (figure, A–C) and an affected family member confirmed the diagnosis of fibrodysplasia ossificans progressiva (FOP).[1] MRI of the cervicothoracic region (figure, C, E–G) revealed diffuse hyperintense signals extending along the neck, thoracic wall, and paraspinal muscles. The presence of dentate nucleus hyperintensity on brain MRI indicates that FOP may not be purely a musculoskeletal disorder, but a multisystem disorder including CNS involvement (figure, D). There was pseudodystonia of the neck without any other neurologic signs. Because ectopic ossification might not be present in the early stages, diagnosis of FOP based on radiographs alone may be delayed. In the presence of characteristic skeletal malformations, MRI can detect preosseous lesions and avoid additional diagnostic procedures, including biopsy.[2]

REFERENCES

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From the Departments of Neurology (S.K.R.) and Imaging Sciences and Interventional Radiology (K.B., C.K.), Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum, Kerala, India.

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