Teaching Video NeuroImage: Chvostek sign with Fahr syndrome in a patient with hypoparathyroidism

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Figure 1 Chvostek sign: Repeated gentle taps in front of tragus over the superficial part of facial nerve elicits infatigable ipsilateral facial muscle contractions with each tap (see video)

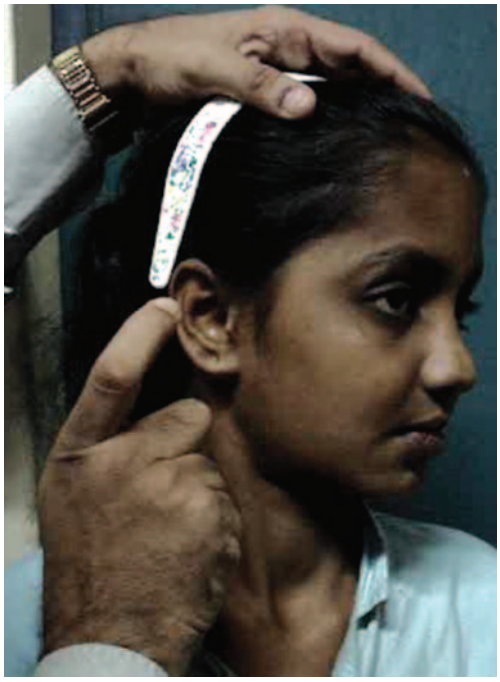
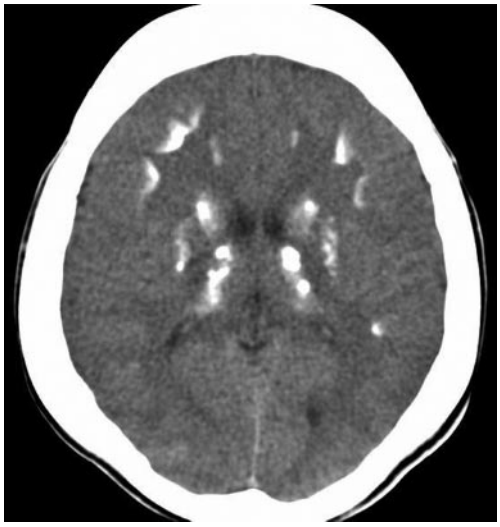


Figure 2 CT scan of the brain with plain axial cuts shows bilateral, symmetric, mirror-image cerebrostriothalamic calcifications (Fahr syndrome)



A 17-year-old short-statured girl (figure 1) presented with seizures, progressive cataracts, and tetany. Further evaluation revealed positive Chvostek sign (video), cerebrostriothalamic calcifications or Fahr syndrome, persistent hypocalcemia (total serum calcium 6.5 [normal 9–10.5] mg/100 mL), and primary hypoparathyroidism (serum parathyroid hormone <3.0 [normal 12–72] pg/mL).

Chvostek sign, seen in hypocalcemia, hypomagnesemia, or alkalosis, is also reported in children with epilepsy.[1] Calcifications in Fahr syndrome (figure 2) are distinguishable from those of physiologic and other causes, including hyperparathyroidism; phacomatoses; gliosis from CNS infections, trauma, or strokes; and neoplasms. Clinically, Fahr syndrome can be associated with rigidity, seizures, psychosis, and dementia. Short stature and primary hypoparathyroidism prompted investigations for a mitochondrial disorder,[2] but a muscle biopsy was normal.

REFERENCES

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Supplemental data at www.neurology.org

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