

Figure 2. First spinal radiographs performed, at the age of 11 months, showing **A**, severe left dorsal scoliosis and **B**, kyphosis.

References

1. Baumann M, Giunta C, Krabichler B, Ruschendorf F, Zoppi N, Colombi M, et al. Mutations in FKBP14 cause a variant of Ehlers–Danlos syndrome with progressive kyphoscoliosis, myopathy, and hearing loss. *Am J Hum Genet* 2012;90:201–16.
2. Giunta C, Baumann M, Fauth C, Lindert U, Abdalla EM, Brady AF, et al. A cohort of 17 patients with lyphoscoliotic Ehlers–Danlos syndrome caused by biallelic mutations in FKBP14: expansion of the clinical and mutational spectrum and description of the natural history. *Genet Med* 2018;20:42–54.
3. Brady AF, Demirdas S, Fournel-Gigleux S, Ghali N, Giunta C, Kapferer I, et al. The Ehlers–Danlos syndromes, rare types. *Am J Med Genet C Semin Med Genet* 2017;175:70–115.
4. Bursztejn AC, Baumann M, Lipsker D. Ehlers–Danlos syndrome related to FKBP14 mutations: detailed cutaneous phenotype. *Clin Exp Dermatol* 2017;42:64–7.

Tick-Tock on the Ward: Essential Palatal Tremor in a Pediatric Patient



A previously healthy 14-year-old girl presented with a 2-week history of tinnitus and continuous movement in the throat. There was no impairment of speech or swallow. Sleep was disturbed due to

tinnitus, with a “tick-tock” sound; sensory relieving techniques were not identified. Examination revealed a repetitive rhythmic movement of the uvula and soft palate (levator veli palatini) with an audible click (**Video** and **Figure**). There was inability to voluntarily suppress the movement. Neurologic examination was otherwise normal. Brain magnetic resonance imaging

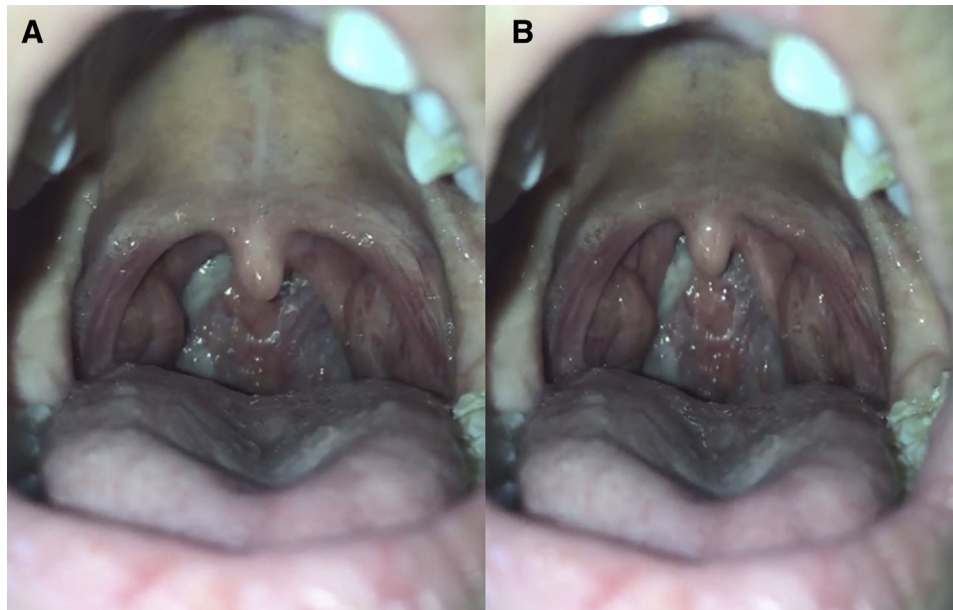


Figure. A and B, Contraction of the levator veli palatini (uvula and soft palate).

was normal, in particular pontine-olivary hypertrophy was not seen. The auditory click, normal neuroimaging, and absence of other neurological signs are in keeping with essential palatal tremor (myoclonus). Clonazepam trial was ineffective, and a trial of botulinum toxin is pending.

Palatal tremor is a rare hyperkinetic movement disorder characterized by rhythmic movements of the soft palate.¹ Recognition is important because it may be the first presentation of an underlying neurologic disorder. Two forms of palatal tremor, essential and symptomatic, are recognized. They seem to be 2 separate clinical entities with differing etiologies and presentations. Essential palatal tremor is characterized by normal neuroimaging and objective tinnitus or audible click, caused by peritubal muscle contractions. As in this case, tinnitus may be the primary complaint, affecting quality of life. Symptomatic palatal tremor, owing to an underlying structural lesion (tumor, hemorrhage, stroke, demyelination) of the brainstem or cerebellum in the dentato-rubro-olivary pathway, is differentiated clinically by the presence of additional neurologic signs such as tremor, nystagmus or ataxia and less frequently an audible click.² Reports on the benefits of treatment of this rare disorder are limited to case reports and small cohort studies. Antiepileptic and anxiolytic drugs have been used in adults with variable results.³ Significant improvement has been reported in some patients after injection of botulinum toxin into tensor veli palatini muscle.⁴ ■

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References

1. Margari F, Giannella G, Lecce PA, Fanizzi P, Toto M, Margari L. A childhood case of symptomatic essential and psychogenic palatal tremor. *Neuropsychiatr Dis Treat* 2011;7:223-7.
2. Kojovic M, Cordivari C, Bhatia K. Myoclonic disorders: a practical approach for diagnosis and treatment. *Ther Adv Neurol Disord* 2011;4:47-62.

3. Saeed RR, Mohammed ZA. Essential rhythmic palatal myoclonus in a 51-year-old man. *Oxford Med Case Rep* 2016;2016:omw056.
4. Penney SE, Bruce IA, Saeed SR. Botulinum toxin is effective and safe for palatal tremor: a report of five cases and a review of the literature. *J Neurol* 2006;253:857-60.

Blaschkoid Angioma Serpiginosum: A Clinicodermoscopic Diagnosis



An otherwise healthy 12-year-old boy presented with gradually progressive (over 2 years), asymptomatic reddish skin eruptions over his right upper limb and chest. There was no history of local trauma or bleeding from any site. No other family member had similar symptoms. Examination revealed bright-red grouped punctate macules and irregular patches arranged in a linear distribution, extending from the lateral aspect of right arm to the right pectoral area, following the lines of Blaschko (**Figure**, A). On diascopy, lesions were nonblanchable. Dermoscopy showed multiple, round-to-oval red lagoons (**Figure**, B) corresponding with dilated papillary dermal

vessels. Other mucocutaneous areas were uninvolved. Systemic examination and routine laboratory measures, including coagulation profile, were normal. Based on clinicodermoscopic findings, a diagnosis of blaschkoid angioma serpiginosum was established. Laser therapy was advised for cosmetic correction.

Angioma serpiginosum is a rare, benign, acquired nevoid vascular anomaly that usually appears in childhood or early adolescence with a predilection for the extremities and sparing mucosal areas. The typical presentation of angioma serpiginosum is copper to bright red, punctate, grouped macules that may transform into papules over an erythema-

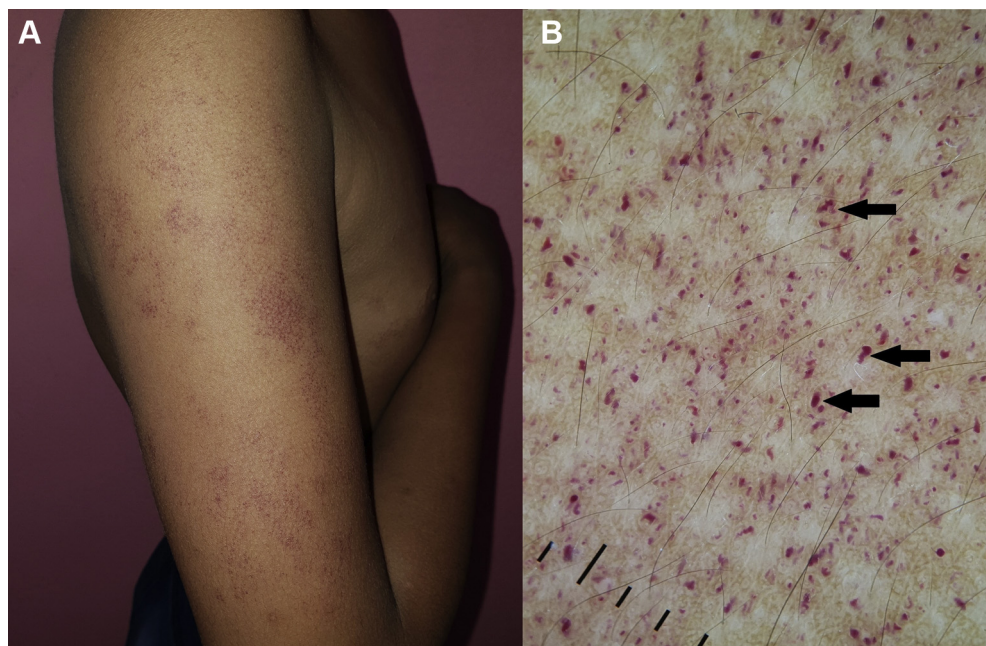


Figure. **A**, Bright-red grouped punctate macules and irregular patches arranged in a linear configuration, following the lines of Blaschko. **B**, Dermoscopy (Dermlite DL4, contact/polarized mode; original magnification $\times 10$) showing multiple, round-to-oval red lagoons (black arrows).

The authors declare no conflicts of interest.