Bizarre tongue movements

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This 14-year-old girl is brought by her parents to your office. You take the history and do the clinical examination.

Video
(Please double click and use only the latest Adobe Reader)

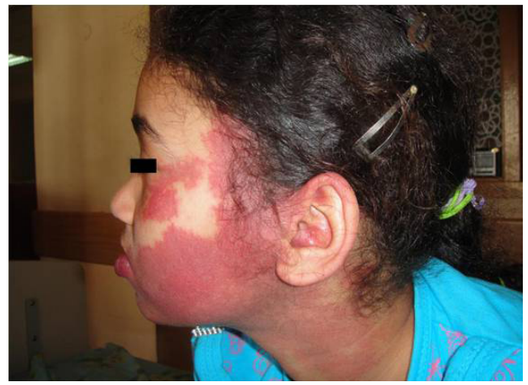

Figure 1

Questions

1. You ask the girl to stick out her tongue (video). What does her tongue reveal?
A. Spastic tongue of pseudobulbar palsy
B. Epilepsia partialis continua
C. Bilateral fasciculations
D. Tongue chorea
E. Rabbit syndrome of orofacial dyskinesia

2. You scrutinize the skin around her mouth (video) and her face (Figure 1). What do you notice?
A. Perioral dermatitis
B. Dermatitis artefacta
C. Butter-fly rash
D. Extensive skin burn
E. Port-wine stain

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Answer

Q1: D
Q2: E

This young girl has been experiencing generalized choreic movements since five years. The facial port-wine stain has been present since birth and it involves the V2 and V3 divisions of the left trigeminal nerve in addition to the second left cervical dermatome. The facial rash is a mere coincidence; the stems mentioned in the quiz may misdirect you to think of Sturge-Weber syndrome and focal seizures. In the video, you may notice overgrowth of the lower lip and port-wine stain, and the "harlequin tongue" or tongue chorea appears when the patient sticks out her tongue. Inside the mouth cavity, the tongue is relatively free of such bizarre, irregularly-timed, non-repetitive, abrupt, and randomly distributed movements. The harlequin tongue is the counterpart of milkmaid's grip. These "motor impersistences" are very characteristic of chorea.[1] Port-wine stains are found in 0.3% of newborn children. However, only 3% those babies have underlying Sturge-Weber syndrome,[2] in which this facial angioma characteristically involves the forehead and upper eyelid (rather the skin of the lower face and upper neck) and predicts the presence of an intracranial leptomeningeal angioma.[3] The patient's older sister and mother have chorea. She has benign hereditary chorea.[4]

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