Pseudoxanthoma elasticum (PXE) is a heterogeneous inherited disorder of connective tissue, the hallmark of which is dystrophic calcification of elastic tissue of the skin, retina, and arteries. The dentist may be the first to suspect and diagnose PXE because of the oral manifestations.

The clinical presentation of PXE often includes the following:

- Persistent raised, yellowish-white plaque-like rash most often on the neck but may occur anywhere on the skin.

- Lower lip: beds or streaks of whitish-yellow patches along the labial mucosa that can often be misinterpreted as Fordyce spots.

The diagnosis of PXE rests on the clinical signs and histologic examination confirms it. The incidence of PXE in the general population is very low. However, the disease can be devastating. The hope is that through early detection of the clinical signs diagnosis and treatment can be started early in life.

For the dental profession, the key is early recognition and referral to a dermatologist and ophthalmologist for further evaluation.