A Rare Soft-Tissue Tumor in a 15-Year-Old Boy With Tuberous Sclerosis Complex: Challenge

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CASE REPORT

A 15-year-old boy known with tuberous sclerosis complex with a *TSC2* mutation and clinical features of hypopigmented macules, facial angiofibromas, mental retardation, epilepsy, angiomyolipoma, and cardial rhabdomyoma presented at our pediatric dermatology clinics. He had a gradually increasing large soft multinodular tumor in the neck (Fig. 1), which had been present for 5 months. At times, the lesion was painful, and the patient noticed purulent discharge. The tumor measured 5.3 × 3.5 cm and showed an irregular surface with comedo-like/cystic openings. The patient requested removal of the tumor for cosmetic reasons.

On gross pathology, the specimen showed a diffuse, pale fibrotic appearance, with fibrosis extending into the subcutaneous fat (Fig. 2A). Histopathologic examination of the lesion revealed 3 components: abundant thick collagen bundles in the dermis and underlying adipose tissue; concentric perifollicular, perieccrine, and perivascular fibrosis; and variable-sized epithelial cysts and dilated openings (Figs. 2B–D).

WHAT IS YOUR DIAGNOSIS?

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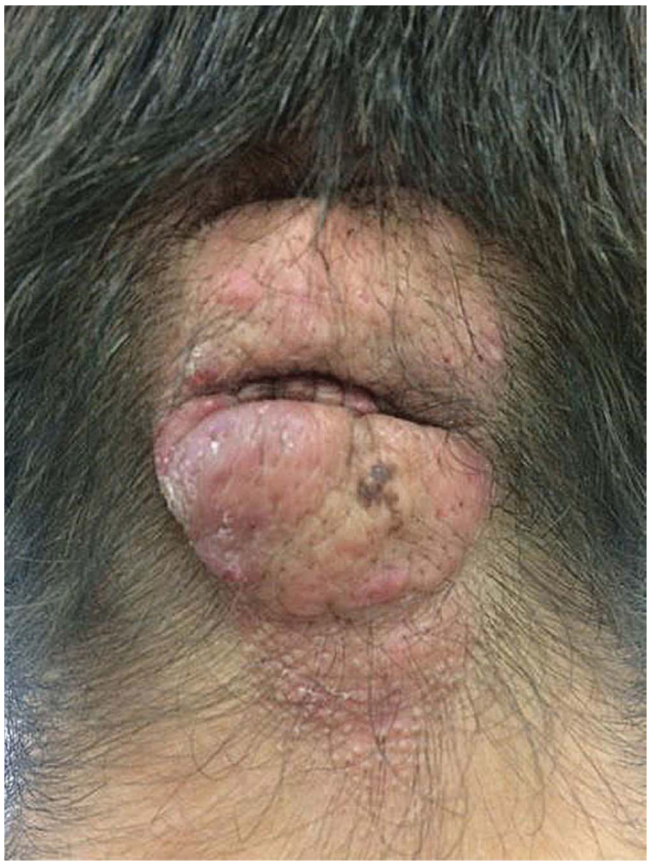

FIGURE 1. Exophytic multinodular tumor in the neck with an irregular surface and comedo-like openings.

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The authors declare no conflicts of interest.

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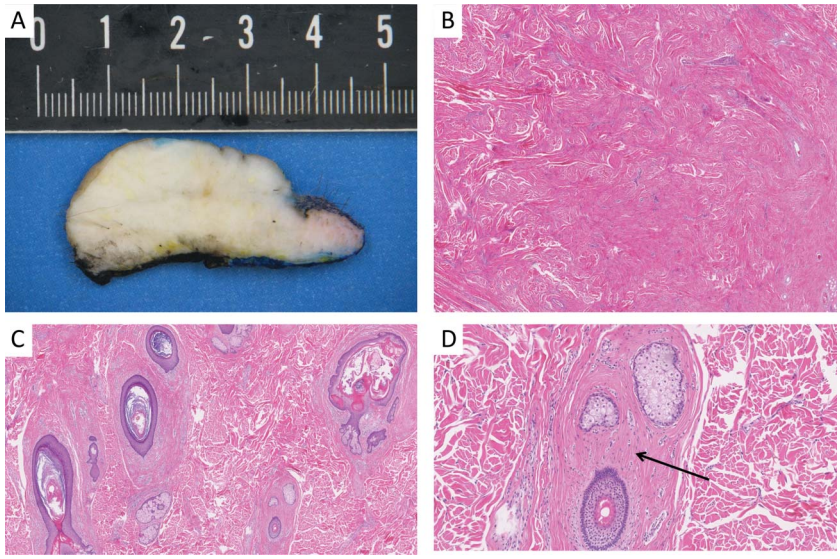

FIGURE 2. Gross pathology shows a diffuse, pale fibrotic rubbery appearance (A). Histopathology reveals abundant thick haphazardly arranged collagen bundles in the dermis and subcutis (B), concentric variable-sized epithelial cysts/dilatation of hair follicles (C), and profound perifollicular fibrosis (D, arrow).