Letters to the Editor

Conflicts of interest
There are no conflicts of interest.

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Figure 3: Digitate structures of loosely arranged subdermal tissue covered by normal mucosal epithelium (H and E, ×40)

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Segmental neurofibromatosis: An unusual association with ocular, skeletal, and cerebral anomalies

Sir,

Segmental neurofibromatosis (SNF) is an uncommon variant of neurofibromatosis that is characterized by café au lait macules and freckles or neurofibromas limited to one region of the body.[1] It is unilateral in majority with right side of the body being more commonly affected. Cervical and thoracic regions are usually involved. It affects all age groups with slight male preponderance.[2] Riccardi included it in his classification as neurofibromatosis type 5.[3] Roth et al. further classified SNF into four subtypes: true segmental, localized cases with deep involvement (nonfamilial), hereditary segmental (no deep involvement, familial), and bilateral segmental (no deep involvement, nonfamilial).[4] Systemic involvement is rare in SNF.[5]

Herein we describe a 43-year-old woman presented with asymptomatic raised skin lesions on left side of face since 3 years. Lesions were gradually increasing in size and number. On examination she had apparent facial asymmetry with left side of the face being less prominent than right side [Figure 1]. Eight skin-colored and hyperpigmented papules and nodules varying in size from 0.3 to 1 cm were present on left preauricular, retroauricular, and mandibular areas [Figure 2]. They were discrete, smooth, sessile, and soft in consistency.

Left eye revealed irregular pupil [Figure 3] and reduced visual acuity. Slit lamp examination showed pear-shaped pupil and absence of iris in temporal region. A part of
the iris showed forward extension into anterior chamber and was attached to the cornea giving a colobomatous appearance. Fundus examination showed deep optic disc cup with optic atrophy. Gonioscopy revealed poorly developed anterior chamber angle. Applanation tonometry showed raised intraocular pressure. Right eye was normal. Other systems were within normal limits.

Histopathology of skin lesions showed a well-circumscribed neoplasm composed of spindle-shaped cells with wavy nuclei and moderate eosinophilic cytoplasm in dermis consistent with neurofibroma [Figure 4].

Plain computed tomography (CT) scan of head showed hypoplasia of maxillary sinus, alveolar arch, and inferior
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