and more recently oral propranolol. Surgical correction is recommended in the involuting or involuted phases (preceded by sclerotherapy).

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Cerebriform Nevus Sebaceous of Jadassohn

An 11-year-old boy presented with raised pigmented lesion over the scalp since 10 years. Parents gave history of hairless yellowish plaque present over the scalp at birth which gradually increased to present size to take cerebriform appearance. There was no history of trauma. All routine hematological investigations were normal. No systemic and developmental defect was noted. *X*-ray skull, eye and neurological examination were normal.

On cutaneous examination, single, 17×8 cm brownish, soft, cerebriform and well demarcated nodular plaques was present over the scalp (***Fig.* 1**). It had multiple folds. Histopathology showed marked papillomatous epidermal hyperplesia with hyperkeratosis and large numbers of mature sebaceous glands in the dermis along with follicular plugging with malformed hair follicles were also present. The correlation diagnosis of cerebriform type of nevus sebaceous was made.

Nevus sebaceous of Jadassohn (NS) is an epidermal nevus, predominantly congenital sebaceous hamartoma with an estimated incidence of 0.3% in the neonates. PTCH gene deletion is proposed mechanism for development of nevus sebaceous. Cerebriform type is a very rare morphologic variant of NS.

It is usually located over head and neck region as solitary lesion and often present at birth as single hairless yellowish plaque with a smooth velvety surface. Multiple extensive lesions may develop with linear, blaschkoid pattern. It becomes verrucous and nodular at puberty indicating role of hormones. Common sites are scalp, forehead, centrofacial, periauricular, and genital area. It may be associated with other developmental defects which are included as epidermal nevus syndrome. Though it occurs sporadically, autosomal dominant transmission was suggested by many case reports. Trichoblastoma is most common benign tumor which develops secondarily in NS, while malignant tumor is basal cell carcinoma (<5%).

The clinical differential diagnosis is congenital melanocytic nevi, epidermal nevus syndrome, giant seborrheic keratosis and warts, while the histopathological differential diagnosis is sebaceous hyperplesia, adenoma, sebaceous carcinoma and sebaceoma.

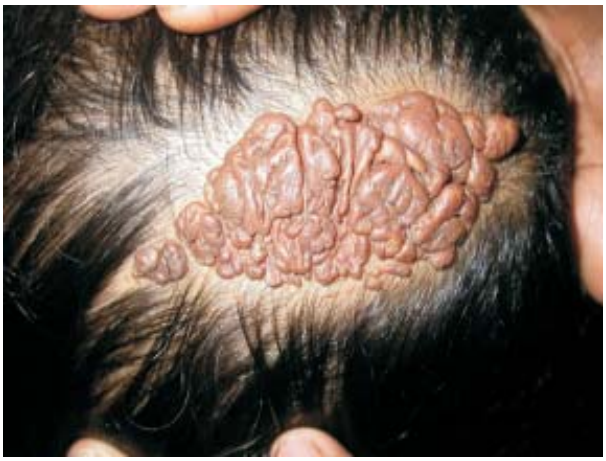

FIG.1 *Cerebriform nevus sebaceous.*

Seborrheic keratosis and epidermal nevus may be difficult to differentiate clinically. Sebaceous adenomas is sharply demarcated structure made up of immature lobules while in sebaceoma basaloid cells predominate along with sebaceous cells ducts. In contrast to nevus sebaceous, sebaceous hyperplasia shows mature sebaceous gland lobules and prominent sebaceous ductal structures. Sebaceous carcinoma shows mitotic cells with undifferentiated growth.

Wide excision remains treatment of choice and patient mainly present for cosmetic purpose. It may be done prophylactically during childhood as there is a risk of malignant transformation, but most tumors remain benign. So regular clinical follow up is necessary. Many other treatment modalities like CO2 laser and photodynamic therapy have been tried.

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