

Adams Oliver Syndrome is a rare and clinically heterogeneous anomaly characterized by the combined occurrence of congenital scalp defects and terminal transverse limb defects. It includes aplasia cutis congenita, variable limb defects, and associated anomalies ranging from skin tags to lymphedema. Other system anomalies and malformations such as cardiovascular, respiratory and orofacial defects have also been reported. With conservative therapy to prevent secondary infection and consequent tissue damage, most small defects of scalp heal well during the first few months of

life. Larger and obvious scars can be treated with plastic surgical reconstruction. The lesions of cutis marmorata may fade with time during first year of life due to skin thickening and maturation.

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Facial Angiofibromas

A nine-year-old boy presented to us with recurrent episodes of convulsions over the preceding two years. His family and birth history was unremarkable. He had learning difficulties and poor scholastic performance. Cutaneous examination revealed multiple non-tender, dark brown and skin-coloured telangiectatic papules of varying size over the face (**Fig. 1**). In addition, he had three oblong hypopigmented macules over the upper back. Systemic examination including ophthalmoscopy was normal. Computerized tomographic (CT) scan of brain revealed multiple calcified subependymal nodules of different size. Based on the features, a diagnosis of tuberous sclerosis was established. Subsequent echocardiography and ultrasonography of the abdomen were normal.

The distinctive facial lesions in the present patient were angiofibromas, also referred by a misnomer – adenoma sebaceum. These are pink, dark brown, or skin-colored telangiectatic papules, often observed in the nasolabial folds and on the cheeks and chin. Pathologically, they are hamartomas, and composed of fibro-vascular tissue. They usually present after 2 years of age and gradually increase in size and number until adolescence. Besides tuberous sclerosis, angiofibroma may also be seen in multiple endocrine neoplasia type I and Birt-Hogg-Dube syndrome. Treatment of these skin lesions is usually required for cosmetic concerns, and different forms of laser being the best option.



FIG. 1 (a) Dark brown and skin-coloured papules of varying size over the face; and (b) Close-up showing typical lesions of angiofibromas.

Common differential diagnoses of angiofibroma include, trichoepithelioma (small, firm, flesh-coloured papules, begin during early puberty), acne (polymorphic lesions, presence of comedones, also present in other acne-prone areas, usually noted during adolescence), syringoma (small, skin-colored, dermal papules, typically on the lower eyelid; occurs between 20 and 40 years of age), and sebaceous hyperplasia (<3 mm yellowish telangiectatic papules with central umbilication; common in elderly people).

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