**Facial Angiofibromas**

Tuberous sclerosis complex is an inherited neurocutaneous multisystem disorder characterized by the potential for the development of hamartomas in almost every organ, most notably in the skin, brain, kidneys, heart, and eyes.\(^1,2\) Facial angiofibromas are hamartomas composed of vascular and connective tissue elements and are found in approximately 75% of patients with tuberous sclerosis complex.\(^1,6\) Histologically, the lesion is characterized by dermal fibrosis and capillary dilatation.\(^7\) Sebaceous glands are often atrophic.\(^1,2,7\) As the lesion of angiofibroma is not related to sebaceous glands, the traditional term “adenoma sebaceum” is a misnomer and should be avoided.

Facial angiofibromas typically appear during preschool years in the malar area (“butterfly distribution”), nasolabial folds, and chin as small pink to red-brown dome-shaped papules with a smooth, glistening surface (Figures 1 and 2).\(^3,6,8\) The lesions gradually enlarge and become more numerous with age until adolescence, and they remain unchanged thereafter.\(^3\) Fibrous cephalic plaque, a variant of angiofibroma, is seen in approximately 20% of patients with tuberous sclerosis complex (Figure 3).\(^5,9\) The lesion may present at birth and commonly becomes more noticeable in early childhood, grows very slowly, and presents as a soft to firm, elevated plaque.\(^10\) The color varies from yellow, pink, tan, and brown.\(^4,5,9\) The lesion is usually on the forehead, but may occur on the scalp or any part of the face including the eyelid.\(^10\) A cephalic fibrous plaque may be the first and most readily recognized feature of tuberous sclerosis complex.

According to a National Institutes of Health consensus conference, a definitive diagnosis of tuberous sclerosis complex can be made when 2 major features or 1 major feature plus 2 minor features are demonstrated.\(^12\) In this regard, 3 or more facial angiofibromas or 1 or more cephalic plaques constitute a major feature of tuberous sclerosis complex.\(^3,4\) Multiple angiofibromas are found...
in approximately 88% of patients with multiple endocrine neoplasia type 1.\textsuperscript{13,14} The angiofibromas in patients with multiple endocrine neoplasia type 1 tend to be smaller and fewer than in patients with tuberous sclerosis complex.\textsuperscript{13,14} They also have a later age of onset. In addition, angiofibromas in patients with multiple endocrine neoplasia type 1 are often observed on the vermilion border of the upper lip—an area that tends to be spared in patients with tuberous sclerosis complex.\textsuperscript{13,14} Patients with multiple endocrine neoplasia type 1 do not have other features of tuberous sclerosis complex.

**Ungual Fibromas**

Ungual fibromas (Koenen tumors), a term for periungual fibromas and subungual fibroma, are hamartomatous fibromas. Generally, periungual fibromas are more common than subungual fibroma.\textsuperscript{4} Periungual and ungual fibromas are more commonly observed on toenails rather than fingernails.\textsuperscript{2,3} Typically, ungual fibromas present as smooth, firm, nodular, or fleshy lesions that are adjacent to the nails near the proximal nail fold (Figures 4 and 5).\textsuperscript{8,16} At times, they may appear underneath the nail plate or over the lateral nail groove (Figure 6).\textsuperscript{4} One of the initial features of ungual fibroma is a groove in the nail plate in the absence of an obvious tumor (Figure 7).\textsuperscript{8} Ungual fibromas are slow-growing.\textsuperscript{6,10} They are found in approximately 20% of unselected patients with tuberous sclerosis complex and are more commonly observed in adolescents and adults than in young children.\textsuperscript{116} These lesions occasionally develop subsequent to trauma.\textsuperscript{3,28} Two or more nontraumatic ungual fibromas constitute a major feature of tuberous sclerosis complex.\textsuperscript{3,28} Before correlating an ungual fibroma to tuberous sclerosis complex, one should explore the possibility whether the lesion is trauma-induced.\textsuperscript{16}

**Shagreen Patch**

The shagreen or “leather” patch is a connective tissue hamartoma made up of collagen and reduced elastic fibers.\textsuperscript{4} Typically, the patch is found in the lumbar region but can also be found on the neck, chest, abdomen, and thighs.\textsuperscript{4,8} Shagreen patches often occur in the first decade of life, although they might not be apparent in young children (Figures 8 and 9).\textsuperscript{8,16}

Characteristically, the lesion presents as an irregularly shaped, unevenly thickened plaque with a cobblestone or orange-peel appearance with a texture of pigskin (Figure 10).\textsuperscript{2,8} The color ranges from pink, grayish green to light brown.\textsuperscript{4,6,16} Shagreen patches are observed in approximately 50% of patients with tuberous sclerosis complex and are a major feature of the disease.\textsuperscript{2,4,8}

**REFERENCES**


tol. 2007;57(2):189-202. https://doi.org/10.1016/j.jaad.200705.004


