Painful tumors of the skin – from ENGLAND to LEND AN EGG to BLEND TAN EGG

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Acronyms

The acronyms for painful tumors of the skin always evoke an interest among students and teachers of dermatology. In fact, the main author of this article was asked during his Post Graduate examination about the acronym ENGLAND. Over the years with the addition of newer entities, new acronyms have been developed. The only common symptom attributed to these heterogeneous benign tumors is pain. We also had the opportunity to publish a few case reports related to atypical presentations of these tumors.[1-3] The acronym ENGLAND or GLENDA is often used to recall these tumors – eccrine spiradenoma, neuroma, glomus tumor, leiomyoma, angiolipoma, neurilemmoma and dermatofibroma. Naversen *et al*. modified the acronym "LEND AN EGG" with the addition of endometrioma and granular cell tumor.[4] With the addition of blue rubber bleb nevus and tufted angioma to this list, presently the acronym stands as "BLEND TAN EGG."[2] In this article, we have briefly described the clinical features and analyzed the pathogenesis of pain in these tumors [Table 1].

Blue Rubber Bleb Nevus

It is characterized by cutaneous and gastrointestinal venous malformations. Skin lesions have a cyanotic bluish appearance with soft elevated nipple-like projections. Nocturnal pain is characteristic.[5]

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Leiomyoma

This tumor develops from smooth muscles.[6] Solitary and multiple piloleiomyomas are derived from arrector pili muscle and angioleiomyomas are derived from muscles of vein.[7] These lesions present as solitary or grouped red, pink, purple, brown, waxy or translucent nodules [Figures 1 and 2]. On histopathological examination, interlacing bundles of smooth muscle bundles with characteristic eel-shaped nuclei are seen [Figure 3].[4]

Eccrine Spiradenoma

It presents as painful, blue-colored, small, nondescript lesions showing histologic differentiatiion towards intradermal eccrine ductal and secretory cells. Histopathology shows deeply basophilic stained, sharply marginated lobules lying freely in the dermis. Two types of epithelial cells are present – small cells with dark peripheral nuclei and pale cells with central nuclei.[8-10] Characteristic appearance is called "blue balls" in the dermis.[11]

Neuromas

These tumors present as painful skin colored lesions which are characterized by proliferative bundles of nerve fibres with a surrounding capsule on histopathology. Morton's neuroma presents as a reactive, persistent and degenerative nodule commonly over the sole.[4]

Dermatofibroma

It presents as a painful hyperpigmented nodule with "dimple sign" most commonly over the legs[4] [Figure 4]. Histopathology shows a hyperplastic epidermis separated by a grenz zone from a dermal tumor composed of varying proportion of histiocytes and blood vessels [Figure 5].[12]

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Figure 1: Leiomyoma cutis: multiple, hyperpigmented, soft nodules over left shoulder, arm and upper back

Figure 2: Leiomyoma cutis: multiple skin-colored to hyperpigmented nodules with a few appearing translucent

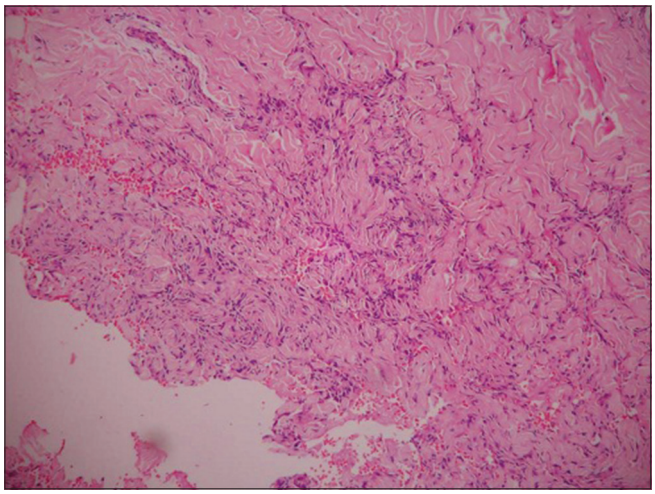

Figure 3: Histopathology of leiomyoma: Dermis shows intersecting fascicles of smooth muscle bundles intermingled with collagen bundles in histopathology on high power (hematoxylin and eosin, ×400)

Table 1: Causes of pain in "BLEND TAN EGG"[23-27]

Tumor	Pathogenesis of pain
Blue rubber bleb nevus	Pain caused by contraction of smooth muscle fibers surrounding the nevus[23]
Leiomyoma cutis	Due to presence of involuntary smooth muscle fibers
Eccrine spiradenoma	Disorganized nerve fibres encasing the spiradenoma nodules seems to contribute to intense pain[24]
Neuroma	Presence of nerve tissue
Dermatofibroma	Probable cause could be entrapped collagen bundles in between interlacing strands of fibroblasts or histiocytes[25]
Tufted angioma	Growth factors (e.g., interleukin-8) and inflammation may be responsible for pain[26]
Angiolipoma	Thrombosis of the capillaries close to the capsule of the tumor[27]
Neurilemmoma	Presence of Schwann cells
Endometrioma	Hormonal stimulation of endometrial tissue
Granular cell tumor	Schwann cell origin and possible neural differentiation
Glomus tumor	Presence of modified smooth muscle cells

Tufted Angiomas

Tufted angioma is an uncommon, benign, vascular tumor that usually develops during infancy or childhood, with a majority of lesions arising before the age of 5 years.[13] It typically presents on the neck, shoulders, trunk or groin as slowly expanding, mottled, red-to-purple patches and firm plaques superimposed with papules and nodules. Lesions are often associated with paroxysmal pain or tenderness to palpation and localized hyperhidrosis as well as hypertrichosis have been observed.[14] Kasabach–Merrit syndrome has been associated with congenital tufted angioma. Histopathologically, tufted angiomas are characterized by multiple, discrete lobules of tightly packed capillaries in a cannonball pattern within the dermis and sometimes in subcutis.[15]

Angiolipoma

It is a variant of lipoma.[4] Cutaneous angiomyolipomas are rare and differ from renal angiomyolipomas in that they are more common in males 33–77 years of age without any association with tuberous sclerosis and are HMB-45 negative. Histologically, they are composed of thick-walled blood vessels, smooth muscle cells and mature fat in variable proportions. Epithelioid cell component is usually absent in cutaneous angiomyolipomas in contrast to renal angiomyolipomas, which may be responsible for HMB-45 negativity.[16]

Neurilemmomas

These are nerve sheath tumors presenting as ovoid, rounded, firm, circumscribed nodules.[17] Neurilemmoma is

Figure 4: Dermatofibroma: solitary, hyperpigmented firm nodule over upper back

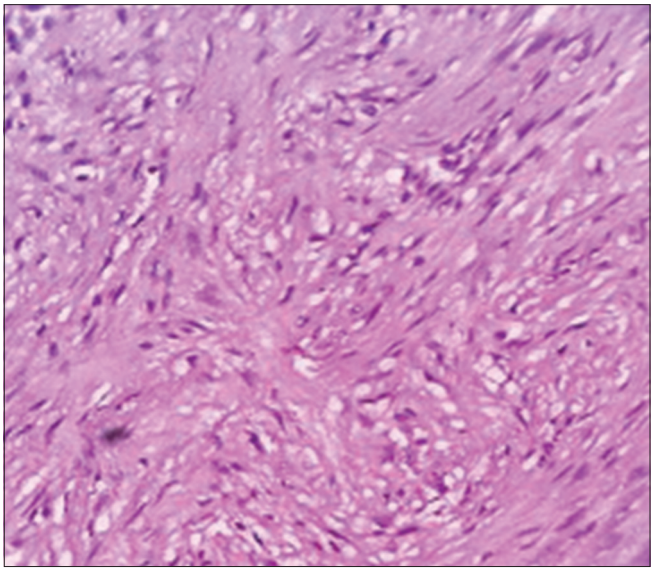

Figure 5: Histopathology of dermatofibroma: Storiform pattern of spindle-shaped cells on high power (hematoxylin and eosin, ×400)

Figure 6: Glomus tumor: dusky, cyanotic nodule over the left calf

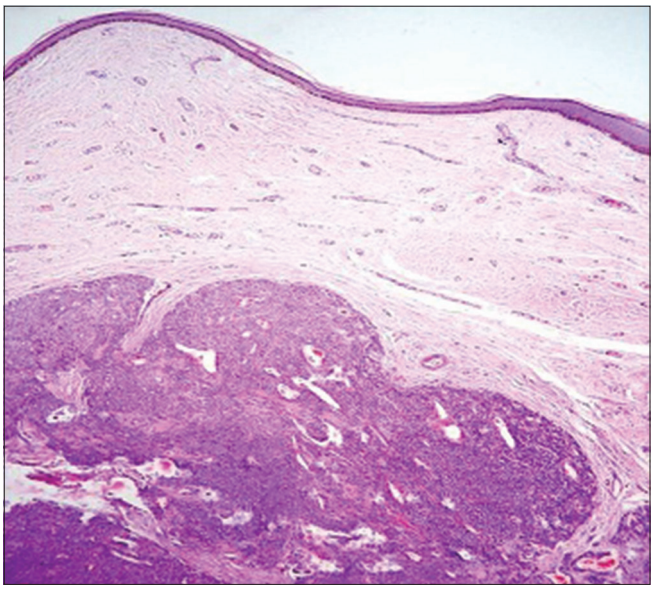

Figure 7: Histopathology of glomus tumor: with hematoxylin and eosin stain showing a well-defined tumor in the dermis with solid sheets of tumor cells which are surrounding vascular channels (×100)

one of the few truly encapsulated neoplasms of the human body.[18] Schwann cells are arranged in bands which stream and interweave. Cellular areas are known as Antoni A areas which are intermixed with areas showing predominantly myxoid change and named as Antoni B areas.[17]

Endometrioma

It is a rare skin tumor presenting with pain and bleeding which worsens at the time of menstruation. It usually appears over a scar or near umbilicus.[19] Histopathology shows a cellular vascular stroma with lumina as well as changes associated with various phases of menstrual cycle.[20]

Granular Cell Tumor

It occurs most commonly over the tongue. Histopathology shows irregular polygonal, large cells with poorly-defined membrane and granular cytoplasm.[17]

Glomus Tumor

This is a neoplasm of normal glomus body with a triad of symptoms of pain, pinpoint tenderness on blunt palpation and hypersensitivity to cold.[21] Commonly seen over digits but extradigital locations have also been described. We have reported a case of glomus tumor over the calf region [Figure 6].[22] Histopathology shows branching vascular channels separated by connective tissue stroma containing aggregates, nests and masses of glomus cells [Figure 7].[3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his

consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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