

Erythematous to Bluish Nodule on the Face in a Young Male

Clinical findings

A 21-year-old male presented with an asymptomatic erythematous nodule of 10 years' duration on his left cheek. The lesion started as a small papule and gradually increased in size. He denied any history of trauma to the site involved. Clinical examination revealed a solitary well-demarcated firm, non-tender nodule measuring 3 cm in diameter on the left cheek [Figure 1]. The central portion of the nodule was erythematous with slight scaling, while the inferolateral margin was focally tinged with bluish-black color. It was non-adherent to the underlying structures. There was no regional lymphadenopathy. A skin biopsy sample was obtained from the nodule and sent for histopathological examination.

Histopathological findings

On histopathological examination, the epidermis was slightly hyperkeratotic, acanthotic with basal layer hyperpigmentation, and Grenz zone present focally. The dermis revealed proliferation of spindle cells admixed with histiocytes around thin-walled endothelium lined dilated vascular spaces, along with extravasation of erythrocytes at places [Figure 2a]. The spindle cells showed mild atypia, elongated nuclei with pointed ends and scanty cytoplasm. Mitotic figures and necrosis were absent. Adjacent to the cellular areas at the periphery of the lesion, tumor cells were arranged in a storiform pattern with foci of collagen entrapment. Prominent intracellular and extracellular brown pigment deposits were present [Figure 2b]. To differentiate between hemosiderin and melanin pigment, we performed histochemical stains- Perls' Prussian Blue and Masson-Fontana. The clumps of golden brown pigment were

highlighted as bluish in color on Perls' Prussian blue stain, corresponding to the presence of hemosiderin [Figure 3a] and were negative for Masson Fontana. Masson-Fontana stained only the melanin pigment in the basal layer of the epidermis. The spindle cells were negative for CD34 [Figure 3b], S-100 and smooth muscle actin.

Diagnosis

Hemosiderotic dermatofibroma

Discussion

Dermatofibroma is one of the most common benign soft tissue tumors of the skin. The clinical and histopathological diagnosis of a classic dermatofibroma is usually straightforward, but it can be challenging in rare subtypes. A wide spectrum of histopathological variants are described, the important ones include cellular, aneurysmal, hemosiderotic, atypical, and epithelioid [Table 1].^[1-4]

Based on the histopathological features, dermatofibroma can be divided into three categories: a) those with cellular/stromal peculiarities, b) with architectural peculiarities, and c) with both cellular/stromal and architectural peculiarities. Hemosiderotic dermatofibroma is included in the first category.^[5] It is a rare though distinct variant, comprising 2–5.7% of all dermatofibromas.^[1,6] It has been described at various anatomical sites like the abdomen, breast, and leg as a blue-gray, dark brown or black nodule or plaque, which may sometimes simulate melanoma.^[5-8]

Histopathologically, hemosiderotic dermatofibroma is composed of numerous small vessels, extravasated red blood cells, intra- and extracellular hemosiderin deposition, in addition

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Table 1: Clinical and histopathological features of important subtypes of dermatofibromas^[1-4]

Type	Site	Age	Sex	Clinical morphology and behaviour	Histopathology
Classical	Limbs	Young to middle-aged	F>M	Firm yellow-brown papule, dimple sign positive	Localized proliferation of histiocyte and fibroblast-like cells with mononuclear inflammatory cells, foamy macrophages, siderophages and multinucleated giant cells Collagen bundles at periphery appear hyalinized with surrounding scattered tumor cells
Cellular	Limbs, head and neck, fingers, toes	Young adults	M>F	Size larger than typical DF Higher local recurrence rate (25%) Few reports of malignancy	More cellular, less polymorphic Focal storiform pattern and variable mitosis- resembles dermatofibrosarcoma protuberans Extension into subcutaneous tissue more prominent than ordinary DF
Aneurysmal	No predilection	Young to middle- aged	F>M	Rapidly growing, may resemble a vascular tumour Risk of local recurrence - 19%	Extensive hemorrhage with prominent cavernous-like pseudovascular spaces not lined by endothelial cells, on the background of ordinary DF Mitosis- variable
Hemosiderotic	No predilection	Young to middle- aged	F>=M	Bluish pink tumor, may resemble a vascular tumor	Numerous small vessels, extravasated RBCs and intra- and extracellular hemosiderin deposition with solid areas of DF
Atypical	No predilection	Young adults	M>F	Papule, nodule or plaque <1.5 cm Local recurrence -14% Rarely metastasis reported	Variable mono- or multinucleated, pleomorphic, spindle-shaped or histiocyte-like cells on a background of an ordinary DF Atypical mitotic figures may be present
Epithelioid	Limbs	Young adults	F>M	Polypoid, vascular morphology like a non-ulcerated pyogenic granuloma	Predominant population of cells with abundant eosinophilic cytoplasm and vesicular nuclei Myxoid change may be seen with prominent vascular component Should be differentiated from spitz nevus

M: male, F: female. DF: dermatofibroma



Figure 1: Single skin-coloured nodule of 3 cm in diameter with an erythematous centre and bluish-black tinge at the inferolateral margin on the left cheek

to features of a classic dermatofibroma like storiform pattern.^[1] It is hypothesized that slow extravasation of blood from the capillaries within the tumor results in the production of hemosiderin, which is engulfed by the tumor cells. Extravasation of blood may occur because of microhaemorrhages triggered by trivial trauma in a vascular dermatofibroma.^[5,6]

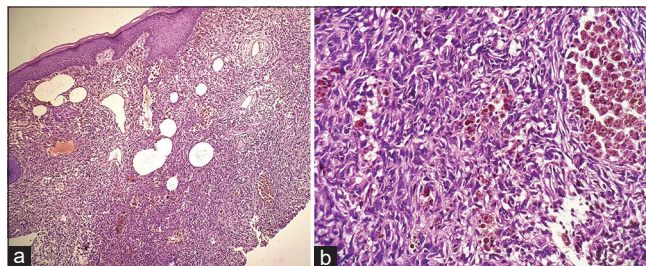


Figure 2: (a) Epidermis is slightly hyperkeratotic, acanthotic with basal layer pigmentation and focal grenz zone. Dermis shows dense proliferation of spindle cells admixed with histiocytes around capillaries lined with endothelium. Extravasation of erythrocytes is present at places, along with abundant brown pigment clumps (hematoxylin-eosin, ×100). (b) The tumor is cellular with spindle cells showing mild pleomorphism, elongated nuclei and scanty cytoplasm. There are prominent brown pigment clumps present inside the cytoplasm of histiocytes and extracellularly (hematoxylin-eosin, ×400)

Aneurysmal dermatofibroma is another uncommon variant, comprising around 1.7% of dermatofibromas.^[3] It presents as a bluish-brown nodule on the limbs, often clinically misdiagnosed as a vascular, melanocytic, adnexal tumor or a cyst. Histologically, it consists of blood-filled pseudovascular spaces varying from narrow clefts to large cavernous cysts, which are devoid of endothelial lining. The vascular channels are surrounded by siderophages, foam cells and fibroblasts. Solid areas resembling classical

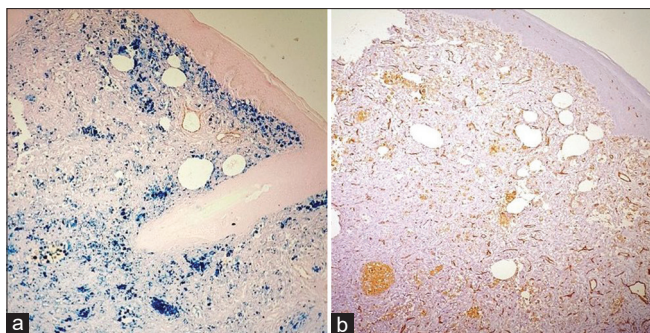


Figure 3: (a) Bluish deposits present as granules and clumps within the cytoplasm of histiocytes and extracellularly in the dermis, corresponding to hemosiderin pigment (Perls' Prussian blue stain, ×200), (b) The tumour cells are negative for CD34, positive control being the endothelial cells lining the blood vessels (CD34, ×200)

dermatofibroma are also present. Other features include interstitial hemorrhage, intra and extracellular hemosiderin deposition, cellular atypia, and mitotic figures.^[1,3] It is believed that hemosiderotic dermatofibroma is a precursor stage in the development of aneurysmal variant. Continuous extravasation of blood in hemosiderotic dermatofibroma results in the formation of dilated channels because of loss of stromal support and increase in intratumoral pressure.^[5] Histologically, aneurysmal dermatofibroma should be differentiated from angiosarcoma, Kaposi sarcoma, and angiomatoid malignant fibrous histiocyoma.^[3]

The present case is interesting as hemosiderotic variant of dermatofibroma is rare. Furthermore, the patient had onset of lesion in childhood and involvement of the face, which are unusual features for dermatofibroma and its hemosiderotic subtype. Histopathologically, the tumor was relatively cellular with mild to moderate atypia, abundant siderophages, and capillary proliferation, thus posing a

diagnostic challenge. It is therefore important to exclude benign and malignant vascular and melanocytic tumors that may mimic hemosiderotic dermatofibroma.

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Conflicts of interest

There are no conflicts of interest.

References

1. Alves JV, Matos DM, Barreiros HF, Bartolo EA. Variants of dermatofibroma-a histopathological study. *An Bras Dermatol* 2014;89:472-7.
2. Calonje E, Mentzel T, Fletcher CDM. Cellular benign fibrous histiocytoma: Clinicopathologic analysis of 74 cases of a distinctive variant of cutaneous fibrous histiocytoma with frequent recurrence. *Am J Surg Pathol* 1994;18:668-76.
3. Calonje E, Fletcher CD. Aneurysmal benign fibrous histiocytoma: Clinicopathological analysis of 40 cases of a tumour frequently misdiagnosed as a vascular neoplasm. *Histopathology* 1995;26:323-31.
4. Kaddu S, McMenamin M, Fletcher CD. Atypical fibrous histiocytoma of the skin: Clinicopathologic analysis of 59 cases with evidence of infrequent metastasis. *Am J Surg Pathol* 2002;26:35-46.
5. Scalvenzi M, Balato A, De Natale F, Francia MG, Mignogna C, De Rosa G. Hemosiderotic dermatofibroma: Report of one case. *Dermatology* 2007;214:82-4.
6. Villarreal DJ, Luz AT, Bucard AM, Abreu L, Cuzzi T. Hemosiderotic dermatofibroma. *An Bras Dermatol* 2017;92:92-4.
7. Kalsi H, Rahman A, Harbol T, Sidhu J. Giant hemosiderotic dermatofibroma: The largest giant dermatofibroma reported to date. *Am J Dermatopathol* 2015;37:778-82.
8. Acar EM, Tad M, Kilitci A, Kemeriz F. Hemosiderotic dermatofibroma mimicking melanoma in a 12-year-old boy: A case report. *Clin Case Rep* 2018;6:1006-9.