Case Report

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ANGIOKERATOMA: A RARE MANIFESTATION

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ABSTRACT

Angiokeratoma shows dilatation of capillaries in the upper dermis and it also shows some epidermal changes, like- hyperkeratosis, papillomatosis, and thickening of epidermis or acanthosis .Clinical presentation of angiokeratoma includes plaques over the skin of lower limbs, mostly on the buttocks or thigh. It is usually identified clinically but a definitive diagnosis is made only on histopathological examination.A deeper biopsy of the lesion is empirically essential to make a concrete diagnosis as the characteristic changes seen in angiokeratoma, are limited to superficial dermis. Our case report has a 24 years old female presenting with unilateral, linear, bluish-black verrucous plaque on the back of left elbow.

KEYWORDS: Angiokeratoma, Linear, Verrucous Plaques.

INTRODUCTION

Angiokeratoma term is originated from three Greek words-angio meaning vessels, kerat for horn and oma implies tumor. (3) It is a rare type of vascular lesion which appears as dark red papule or plaque, and occurs singly or in clusters. It is predominantly a congenital condition with no racial predisposition. It shows slight male predominance, however in case of angiokeratoma circumscriptum females are more frequently affected.(4).The exact pathogenesis of angiokeratoma is not known clearly, however, congenital factors, pregnancy, perniosis, and trauma can be the causative factors.(3).The basic underlying pathology is dilatation of capillaries in the upper dermis with evidence of associated changes in the epidermis, like-increased keratinisation and thickening. These secondary changes further prevent the dilatation and rupture of capillaries.(2).There are many variants of angiokeratoma, like- angiokeratoma circumscriptum, solitary and multiple angiokeratomas, angiokeratoma of Fordyce, angiokeratoma of Mibelli, and angiokeratoma corporis diffusum.(4).Clinically, it appears as dark red to dark blue plaques or nodules, occurring unilaterally, over the lower extremities.

CASE REPORT

Clinical History: A 24 year old female presented to the dermatology OPD (Out Patient Department) of Era's Lucknow Medical College and Hospital, Era University, Lucknow, U.P., with the chief complaint of multiple well defined coalescent bluish black papules, soft in consistency over the back of left elbow since 2 years (Fig.1). Patient also gave the history of intermittent regression in size and episodes of bleeding from the lesion, after minor trauma. However, no complaints of oral or rectal bleeding were noticed during the disease course.

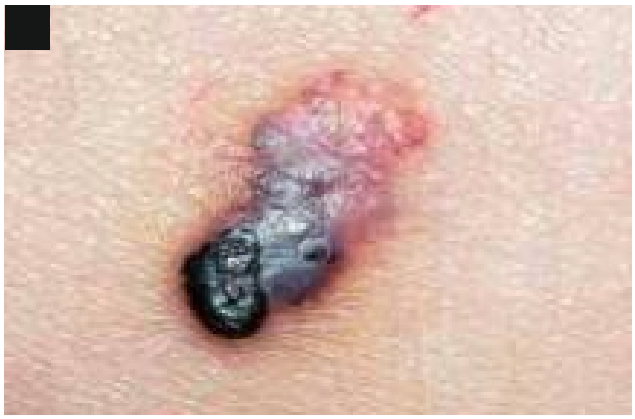

Fig. 1: Clinical Image Showing a Linear, Unilateral, Verrucous Plaque Present in the left Hand, near Elbow.

General physical examination was within normal limits. No history of pruritis, increased pigmentation or any other similar lesion was noted. Family history and medical history were insignificant. Local examination of the lesion revealed unilateral, linear, soft, bluish blackish papules of size 3x1 cm (Fig.1). Routine blood investigations were normal. Clinically, the differential diagnosis of angiokeratoma and melanoma were made. Excisional biopsy was done

and the specimen was sent to the histopathology laboratory for examination. However, we lost the patient in follow-up, so further work-up was not done.

Grossly: The specimen was solitary, gray-white skin covered soft tissue piece measuring ~0.5 cm. Routine tissue processing and H&E staining as per the protocol standardized in our histopathology laboratory was performed.

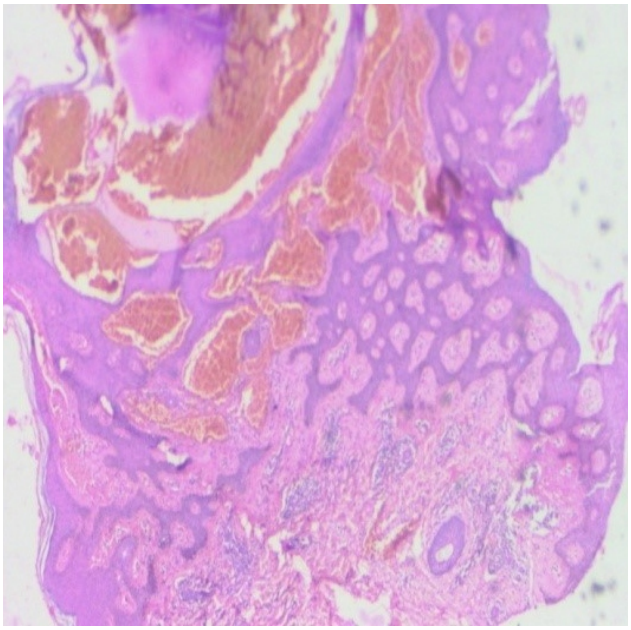

Fig. 2: (H&E,4X) Histological Examination Revealed Hyperkeratosis, Elongation of rete Ridges and Congested, Dilated Capillaries in the Upper Dermis

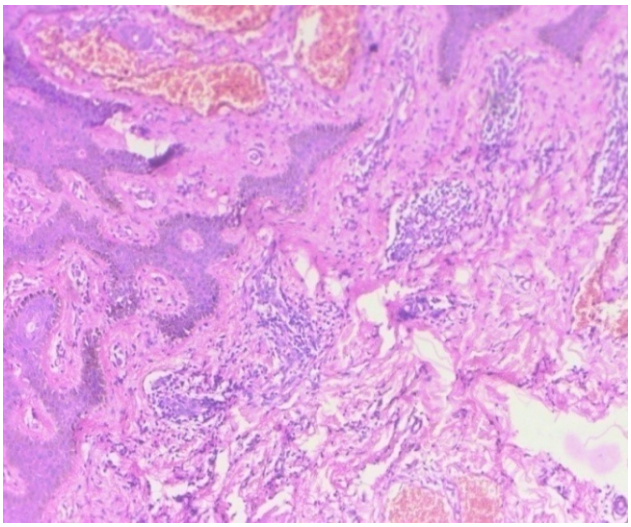

Fig. 3: (H&E, 10X), Dilated and Congested Capillaries seen in upper Dermis. No Involvement of Deeper Dermis seen.

Microscopically: H&E (Haematoxylin and Eosin) stained sections showed epidermis and dermis. Epidermis was lined by keratinized stratified squamous epithelium and showed increased keratinization, thickening and elongation of rete ridges. (Fig.2).The underlying papillary dermis shows numerous dilated and thin-walled, congested blood capillaries. (Fig.3). No involvement of deeper dermis is seen. No evidence of granulomatous or neoplastic pathology noted. Finally, the diagnosis of angiokeratoma was made, based on clinical and histopathological features.

DISCUSSION

Angiokeratoma has been described as a manifestation of different diseases, since it is first reported.(5,6). In 1967, Imperial and Helwig (7) described angiokeratomas as telangiectasias or dilatation of pre-existing vessels and not true vascular tumours or angiomas. The exact pathogenesis of angiokeratoma is not known. However, various factors, like-congenital anomaly, pregnancy, trauma, and subcutaneous hematomas have been proposed as its etiology.

It is an uncommon vascular malformation seen in the papillary dermis where the deeper dermis and subcutaneous tissue are not involved. The lesions occur typically on the legs, but can occur anywhere on the lower extremity, like- buttocks, thigh etc.(4). Also, the involvement is commonly unilateral.

Initially it presents as a reddish macule but gradually with time, some colour change is seen in the lesion from dark red to blue-violet along with features of hyperkeratotis. These lesions generally do not have any tendency of spontaneous regression. However, they may be eradicated permanently with surgery or laser therapy with rare incidence of recurrence. On clinical examination, the lesion of angiokeratoma may resemble melanoma, especially if the dilated vessels get thrombosed, giving the lesion a melanoma like darker appearance (5)(6).

On histological examination, angiokeratoma appears as dilated capillaries in the upper region of dermis. The overlying epidermis zone also shows some changes, like- thickening, elongation of the rete ridges and hyperkeratosis. But the deeper dermis is not involved. So, the characteristic feature of angiokeratoma on histopathological examination is dilated capillaries, present in the superficial dermis. Hence, it is placed among the hypertrophic vascular tumors or telangectasias, showing only dilatation of the capillaries without any cellular proliferation. These dilated vessels further, may form large vascular spaces or lacunae in the superficial dermis and even get thrombosed (1,2,8). Also, the rete pegs may get elongated and encase the vascular lacunae either

partially or completely. Similar lesions having angiomatous components in the deeper dermis are reported as verrucous hemangiomas.

Differential diagnosis of angiokeratoma includes lesions like, verrucous hemangioma, lymphangioma circumscriptum, Cobb syndrome, angioma serpiginosum, verrucae, and some tumors, like melanoma.

Verrucous hemangioma is an uncommon congenital condition of vascular lesion which on histology shows dilated vessels present in the deeper dermis and even the subcutaneous fat along with the hyperkeratinisation of epidermis.(9). It is classified among the hyperplastic vascular tumors showing proliferation of endothelial cells which leads to the formation of new blood vessels (9,10).

Lymphangioma circumscriptum is characterized by the presence of proteinaceous eosinophilic material in the lymphatic channels but the differentiation from blood may become difficult if erythrocytes are also present into the spaces. The presence of clear vesicles may help in its differentiation from angiokeratoma, clinically.

Cobb syndrome is a very rare condition defined by the presence of a congenital vascular skin nevus and a meningospinal angioma, leading to neurologic deficits, including paraplegia (11).

Angioma serpiginosum presents during childhood as small reddish macules or papules with a linear or serpiginous pattern, mainly on the lower limbs of females. However, it does not show the hyperkeratotis of the epidermal surface (12).

CONCLUSION

Diagnosis is based mainly on the histopathological examination, however, a right clinical evidence is also necessary to have an adequate deeper biopsy specimen. To the best of our knowledge, both the incidence and the site of occurrence of this entity, are rare which prompted the present case report. Commonly, this lesion is seen on the lower extremities however in our case, the patient presented with eruptions on upper extremity.

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