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Case Report

Dermatology Section

Solitary Angiokeratoma: Report of Two Uncommon Cases

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ABSTRACT

Angiokeratomas are rare benign vascular skin lesions arising in isolation or in groups of multiple lesions, as solitary cutaneous forms or generalized systemic forms. They are ectasias of dermal capillaries with an acanthotic and hyperkeratotic epidermis. They can occur in both healthy individuals and in those with underlying systemic disease due to inherited enzyme deficiency or other acquired predisposing factors. The identification and reporting of these lesions is important as patients with these lesions should be evaluated to rule out underlying pathogenic conditions.

We report two rare cases of isolated solitary cutaneous angiokeratoma occurring in two patients.

Keywords: Benign vascular lesion, Cutaneous, Vascular ectasia

CASE REPORT

Case 1

A 21-year-old female patient presented with a single asymptomatic pigmented patch over her left thigh of one year duration. Examination showed an irregular brownish and nodular lesion measuring 0.7 x 0.6 cm. There was no tenderness, bleeding on touch or induration of the base of the lesion.

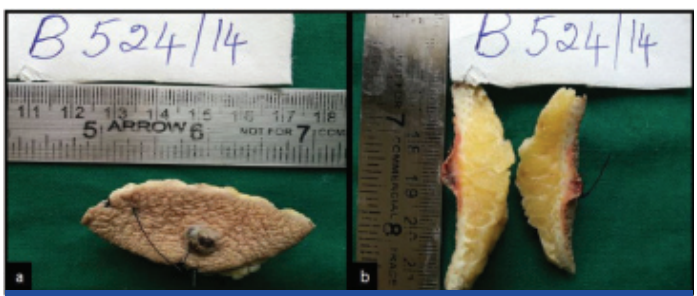
Gross examination: Specimen consisted of an elliptical wide excision skin biopsy with a central raised brownish papulonodular lesion measuring 0.7x0.6 cm. Cut section showed dilated channels filled with blood, within the lesion [Table/Fig-1a&b].

Case 2

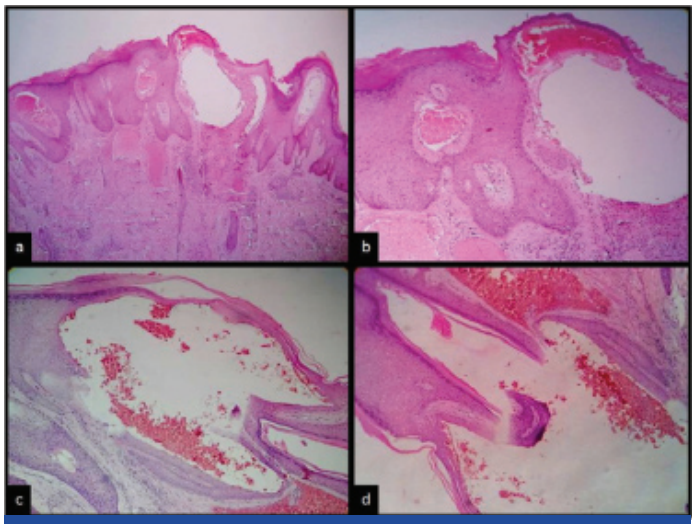
A 17-year-old boy came with the chief complaints of swelling over the back since eight months. There was no history of bleeding, pain or discharge from the swelling. On examination it measured around 1 x 1 cm and was non-tender with firm consistency.

Gross examination: Specimen consisted of a small bit of skin biopsy measuring 1 x 0.7 x 0.5 cm. External surface showed a raised brownish papulonodular lesion. Cut section showed haemorrhagic areas in the lesion.

In both these cases, there was no regional lymphadenopathy. There were no other similar lesions in the body. Their past, personal and family history did not yield any significant information. With the provisional clinical diagnoses of nevus, haemangioma, pyogenic granuloma and melanoma, excision biopsies were performed under local anaesthesia and sent for histopathological examination.



[Table/Fig-1]: Gross pictures showing a) the papulonodular lesion with pigmented external surface; b) cut section showing dilated channels filled with blood



[Table/Fig-2a-d]: Case 1: Photomicrographs showing a) dilated and ectatic vascular channels in papillary dermis immediately beneath the epidermis (H&E;x100). b)epidermal reaction, seen as hyperkeratosis, papillomatosis and hypergranulosis with ecstatic subepidermal vessels lined by flattened cells filled with blood and scattered dermal lymphocytes (H&E;x400). Case 2: c) Ectatic blood vessels in papillary dermis filled with blood(H&E;x100). d) Hyperkeratotic epidermis and vascular channels lined by flattened endothelial cells in dermis (H&E;x400)

Microscopy of both these lesions showed dilated and ectatic vascular channels lined by flattened endothelial cells, filled with blood in dermis closely abutting epidermis. The covering epidermis showed hyperplasia acanthosis, papillomatosis and hyperkeratosis. There was no atypia, increased melanocytes, viral inclusions or ulcerations of epidermis Case 1: [Table/Fig-2a,b] Case 2: [Table/Fig-2c,d].

Thus, a final histopathological diagnosis of angiokeratoma was offered. In both the cases there were no additional similar lesions on the body. Associated symptoms and syndromes were ruled out. Hence these cases were typed as solitary angiokeratoma.

DISCUSSION

The accurate prevalence of angiokeratoma is difficult to calculate as these lesions are asymptomatic. The prevalence rises from 0.6% in first and second decades to 16.6% in the 7th decade of life. Males are commonly affected than females. It is frequently encountered in Caucasian and Japanese individuals [1].

It presents as single or multiple reddish or blackish hyperkeratotic papules, punctuate-to-10-mm in diameter. It can be localized and generalized. Solitary angiokeratoma occurs most frequently on the lower extremities, penis, vulva and clitoris and rarely in the oral cavity [1,2].

Localized angiokeratoma may be solitary or multiple. It can be classified into Fordyce's angiokeratoma (arising on the genitals), Mibelli's angiokeratoma (dorsum of toes and fingers) and angiokeratoma circumscriptum naeviforme (unilateral large keratotic plaques). In the generalized form, angiokeratoma corporis diffusum (ACD), lesions are usually concentrated between the umbilicus and knees. ACD has been observed in both healthy individuals as well as persons with various enzyme deficiency disorders [3].

Enzyme deficiencies include α-fucosidase (fucosidosis), neuraminidase (sialodosis), aspartyl glycosaminase (aspartyl glucosaminuria), β-mannosidase (β- mannosidosis), α-N-acetyl galactosaminidase (Kansaki disease), and β-galactosidase (adult onset GM1 gangliosidosis) [3].

Clinically, angiokeratomas appear as well-circumscribed, red to blue or black verrucous papules and can resemble melanocytic nevus, malignant melanoma, verruca vulgaris, haemangioma, capillary aneurysm, Spitz nevus or focal epithelial hyperplasia [4].

Cavernous haemangiomas are rare in skin and superficial soft tissues. Here, the vascular channels are separated by thick fibro connective tissue in case of cavernous haemangioma which was lacking in our case [4]. Pyogenic granulomas show numerous inflammatory cells and oedema of dermis in addition to vascular channels which were lacking in this case [4]. Nevi and melanoma were ruled out as the melanocytic cells were distributed normally in the epidermis and there was no atypia. Lymphangiomas were ruled out as the channels contained blood and not lymph [4].

Verrucous haemangioma is a congenital lesion. It is a localized vascular malformation involving the dermis and subcutaneous fat. Histopathological examination shows hyperkeratosis, dilated capillaries, large cavernous, endothelial-lined, and blood-filled spaces extending deep into the reticular dermis and subcutaneous fat [2].

The aetiopathogenesis of angiokeratoma is uncertain. The first event is the vascular ectasia within the papillary dermis just beneath the basement membrane. The epidermal hyperplasia is the secondary reaction. Increased venous pressure proximal to the site and raised intra-abdominal pressure can predispose to the formation of angiokeratoma Fordyce [2,5,6]. Angiokeratoma scroti is associated with varicocele, hernia, prostatitis, lymphogranuloma venereum, carcinoma of the bladder, and thrombophlebitis. Vulvar form was associated with pregnancy, vulvar varicosity, postpartum period, and hysterectomy. These conditions cause increase in the local venous blood pressure [2,5].Thus these patients should also be examined both clinically and radiologically for the abdominal masses.

The electron microscopy shows vascular channels lined by very thin endothelial wall. Their cytoplasm shows microfilaments, pinocytosis vesicles and vacuoles with scarcely electron-opaque content, with a diameter of 2,000 and 4,000 A and scanty cytoplasmic organelles, such as mitochondria and endoplasmic reticulum [7]. They can arise at the site of lymphangioma circumscriptum and melanocytic nevi [8]. The complications include sudden bleeding from the lesions which may be spontaneous or may follow trivial trauma [1,2]. This can be controlled with direct local pressure followed by definitive therapy. Excision, electrodessication, cryotherapy, or laser ablations are the various modes of therapy [5]. Recurrences have been reported after surgical excision. Malignant transformation has not been reported in angiokeratoma [4]. Upon further evaluation no other underlying pathological conditions were encountered in both our patients and both were asymptomatic at the end of 2 months of follow-up.

CONCLUSION

We presented two rare cases of solitary angiokeratoma arising in young patients without any underlying pathology.These cases are rare and require extensive patient evaluation to rule out the predisposing conditions. Thus, the knowledge of this entity helps to make accurate histopathological diagnosis and to rule out all the benign and malignant clinical differential diagnosis.

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