

CASE REPORT

ANGIOKERATOMA: A CASE REPORT

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ABSTRACT: A 7 year old female had a raised rough lesion on the right knee, which was compressible. Due to cosmetic reasons, the same was excised and proved to be a case of angiokeratoma.

KEYWORDS: Vascular Nevi, Angiokeratoma.

INTRODUCTION: The neoplasms of vascular origin are mesodermal derivative which may be present from birth or can appear in early childhood; sometimes they may appear at a later stage of life. These lesions histologically may have vessels, some of which are fully developed and filled with blood and few will be presented as collection of endothelial cells.

CASE SUMMARY: A 7 year old female presented with raised plaque like lesion on the right knee. The surface had corrugated appearance and deep red to black in color; compressible.

MACROSCOPY: The specimen was 4x1cm skin covered soft tissue mass, the surface of which showed corrugated appearance. Cut section revealed tiny pores and spongy in appearance, black colored, involving the epidermis and upper dermis.

MICROSCOPY: Section revealed skin biopsy material upto deep sub-cutaneous adipose tissue level. There is hyperkeratosis, acanthosis and papillomatosis. In the areas of papillomatosis, dilated vascular channels, mostly filled with RBCs are seen. Some of the channels are occluded with thrombi. There are also few spaces filled with lymph and there is formation of granulation tissue at one focus.

There are congested vascular channels in the dermal region.

FINAL HISTOLOGICAL DIAGNOSIS: Angiokeratoma circumscriptum.

DISCUSSION:

WHO CLASSIFICATION OF CUTANEOUS VASCULAR TUMORS¹

BENIGN TUMORS AND TUMOR-LIKE CONDITIONS	INTERMEDIATE VASCULAR TUMORS
Papillary endothelial hyperplasia (Masson's tumor)	Locally Aggressive
Reactive angioendotheliomatosis	Kaposi-like hemangioendothelioma
Glomeruloid hemangioma	Giant cell angioblastoma
Bacillary angiomatosis	Rarely metastasizing
Vascular ectasias	Kaposi's sarcoma
Nevus flammeus (salmon patch and port-wine stain)	Retiform hemangioendothelioma
Angiokeratoma	Papillary intravascular
Generalized essential telangiectasia	angioendothelioma(malignant

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<p>Cutaneous collagenous vasculopathy Unilateral nevoid telangiectasia Angioma serpiginosum Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu) Nevus araneus Venous lake Congenital hemangiomas RICH NICH Capillary hemangioma Variants: Infantile hemangioma Cherry angioma Tufted angioma (angioblastoma) Lobular capillary hemangioma (pyogenic granuloma) Cavernous hemangioma Variant: Sinusoidal hemangioma Verrucous hemangioma Microvenular hemangioma Hobnail hemangioma Epithelioid hemangioma (angiolymploid hyperplasia with eosinophilia) Cutaneous Epithelioid angiomatous nodule Acquired elastotic hemangioma Arteriovenous hemangioma (angiolymploid hyperplasia with eosinophilia) Variants: Superficial (cirroid aneurysm) Deep Angiomatosis Spindle cell hemangioma Symplastic hemangioma</p>	<p>endovascular papillary angioendothelioma, Dabska tumor) Composite hemangioendothelioma</p> <hr/> <p>MALIGNANT VASCULAR TUMORS</p> <p>Epithelioid hemangioendothelioma Angiosarcoma Variants: Idiopathic (head and neck) Associated with lymphedema Postradiotherapy Epithelioid</p> <hr/> <p>TUMORS OF LYMPH VESSELS</p> <p>Lymphangioma Variants Lymphangioma circumscriptum Cavernous lymphangioma/cystic hygroma Benign lymphangioendothelioma (acquired progressive lymphangioma) Lymphangiomatosis Lymphangiomyoma</p> <p>Atypical vascular proliferation after radiotherapy</p> <hr/> <p>TUMORS OF PERIVASCULAR CELLS</p> <p>Glomus tumor Glomangiomatosis Variant: Infiltrating glomus tumor Glomangiosaroma Myopericytoma</p>
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The common congenital vascular lesions of the skin are Nevus flammeus and Strawberry mark. Nevus flammeus otherwise known as Port-wine stain is a congenital vascular lesion of telangiectatic type and the other common congenital vascular lesion Strawberry mark² is a type of capillary hemangioma. Deep-seated congenital vascular lesions will most commonly be of cavernous type.

Bartonellosis (*Verruca peruana*) simulate hemangioma, but the organism can be found in the endothelial cells.³

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The common vascular lesions that can occur in the upper dermis, involving the epidermis are Angiokeratoma and Lymphangioma circumscriptum.

ANGIOKERATOMA CAN BE CLASSIFIED AS:

ANGIOKERATOMA CORPORIS DIFFUSUM: Patients present with numerous clusters of tiny red papules in a symmetrical distribution usually in the bathing-trunk area.¹

ANGIOKERATOMA OF MIBELLI: Usually appearing during childhood or adolescence, several dark red papules with a slightly verrucous surface are seen on the dorsa of the fingers and toes measuring 3 to 5 mm in diameter.¹

ANGIOKERATOMA OF FORDYCE: Multiple vascular papules 2 to 4 mm in diameter are seen on the scrotum. They arise in middle or later life. Early lesions are red, soft, and compressible; later, they become blue, keratotic, and non-compressible.¹

SOLITARY OR MULTIPLE ANGIOKERATOMAS: Usually one and occasionally several papular lesions arise in young adults, most commonly on the lower extremities. The lesions range from 2 to 10 mm in diameter. Early lesions appear bright red and soft, but they later become blue to black, firm, and hyperkeratotic.⁴

HISTOPATHOLOGY: The histologic findings are essentially the same in all four above-mentioned types of angiokeratoma.

It consists of numerous, dilated, thin-walled, congested capillaries mainly in the papillary dermis underlying an epidermis that shows variable degrees of acanthosis with elongation of the rete ridges and hyperkeratosis.⁴

LYMPHANGIOMA CIRCUMSCRIPTUM: Lymphangioma circumscriptum is predominantly a developmental malformation of infancy with an equal gender incidence, but it may arise at any age. The proximal portions of the limbs and limb girdle are most frequently affected. Association with cavernous lymphangioma, cystic hygroma, and even lymphangiomatosis is common.¹

A typical lesion consists of collections of numerous vesicles containing clear fluid and less commonly, blood. Due to the presence of a deep component, lesions arising in infancy tend to recur after simple excision.¹

HISTOPATHOLOGY: Lymphangioma circumscriptum is composed of numerous dilated lymphatics in the superficial and papillary dermis. There is clear fluid and, less frequently, red blood cells in their lumina. In the overlying epidermis there is some degree of acanthosis and hyperkeratosis.

The surrounding stroma shows scattered lymphocytes. Lesions developing in infancy often show a large-caliber, muscular lymphatic space in the subcutaneous tissue, which has to be ligated at the time of excision to avoid recurrence.¹

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MACROSCOPIC IMAGES



MICROSCOPIC IMAGES

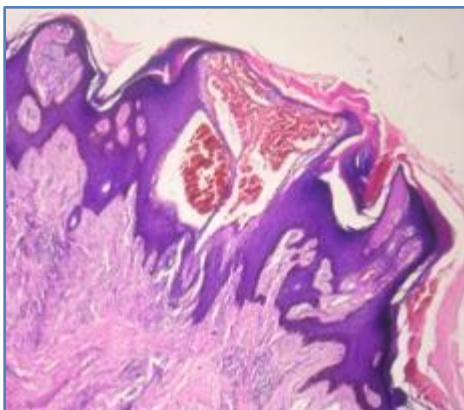


Fig. 1A: H & E stained 4 x

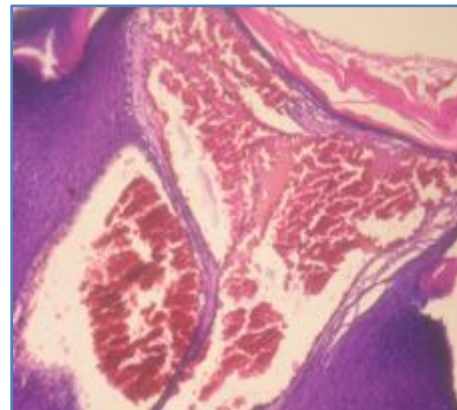


Fig. 1B: H & E stained 10 x

Showing dilated blood vessels in the epidermal zone which is lined by endothelial cells.

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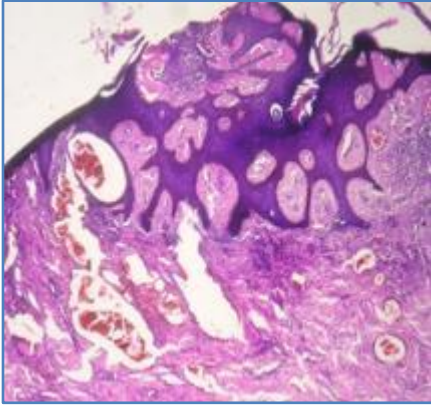


Fig. 2A: H & E stained 4 x

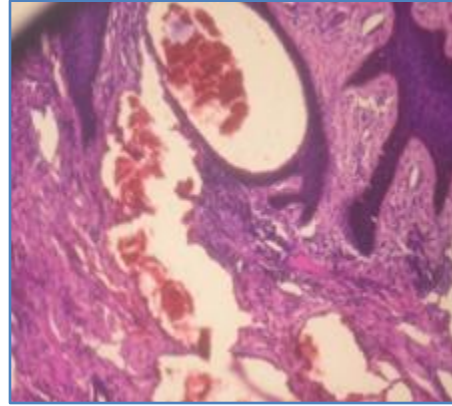


Fig. 2B: H & E stained 10 x

Showing dilated blood vessels filled with RBCs in the upper dermis. Also seen are telangiectic spaces.

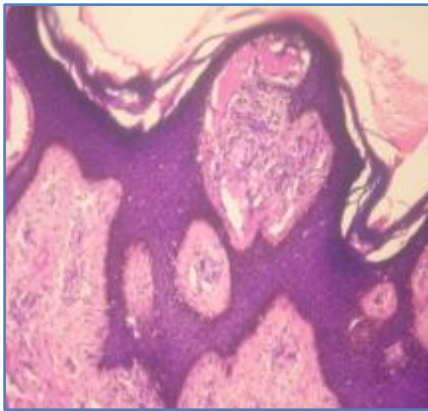


Fig. 3A: H & E stained 10 x

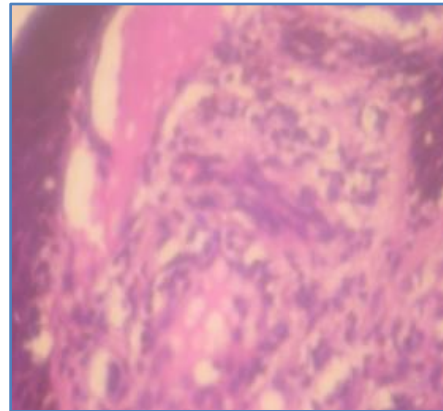


Fig. 3B: H & E stained 40 x

Dilated space filled with granulation tissue located in the epidermis.

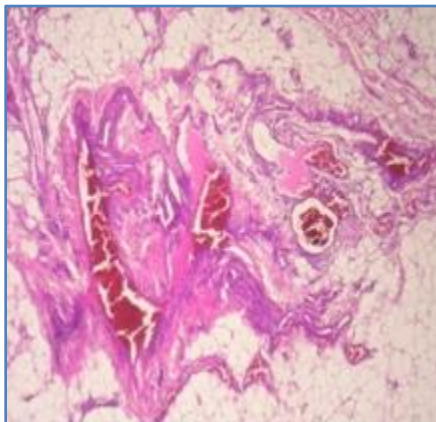


Fig. 4A: H & E stained 4 x

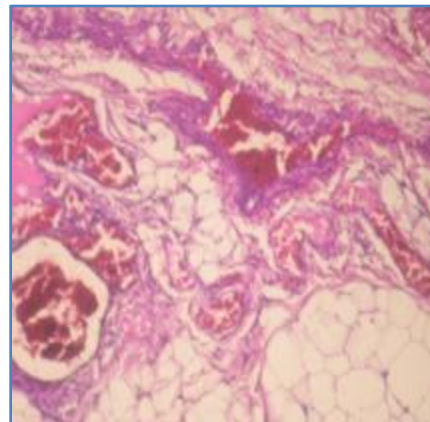


Fig. 4B: H & E stained 10 x

Dilated vascular spaces many filled with RBCs and few filled with lymph present in the deep sub-cutaneous adipose tissue level.

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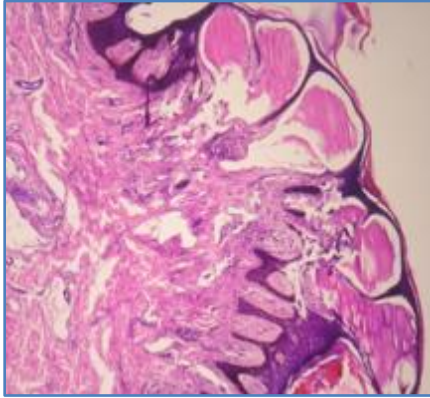


Fig. 5A: H & E stained 4 x

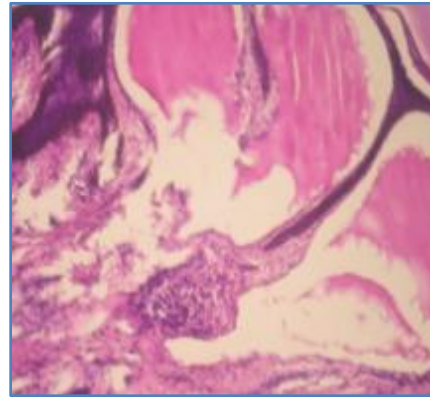


Fig. 5B: H & E stained 10 x

Raised plaque-like lesion present in the upper dermal region; lined vascular spaces filled with lymph and few spaces filled with RBCs.

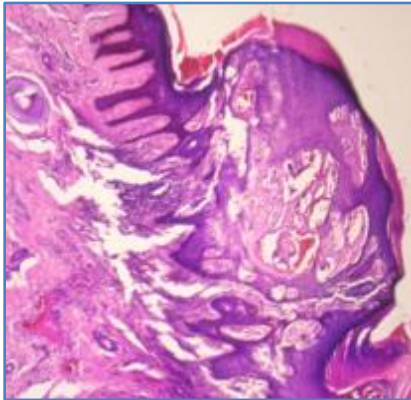


Fig. 6A: H & E stained 4 x

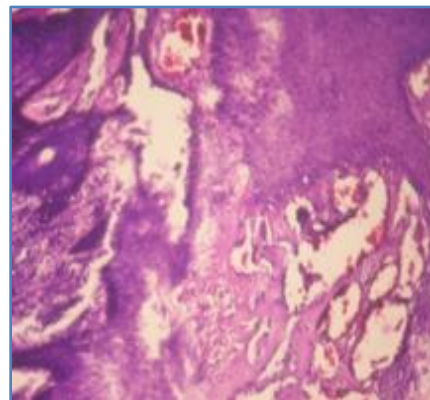


Fig. 6B: H & E stained 10 x

Raised plaque-like lesion, where telangiectatic spaces present in the epidermis. Dilated vascular channels present in the upper dermal region.

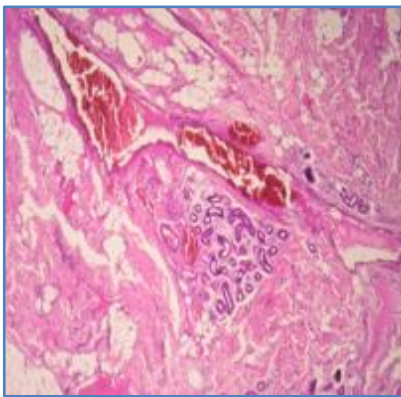


Fig. 7A: H & E stained 4 x

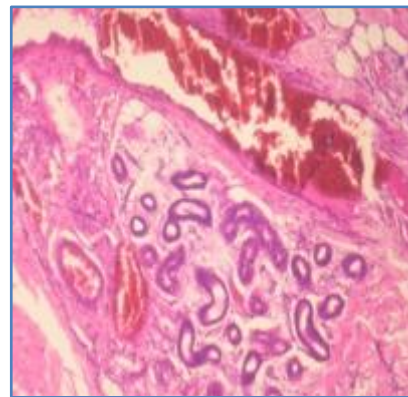


Fig. 7B: H & E stained 10 x

Dilated vascular channel in mid-dermal zone filled with RBCs.

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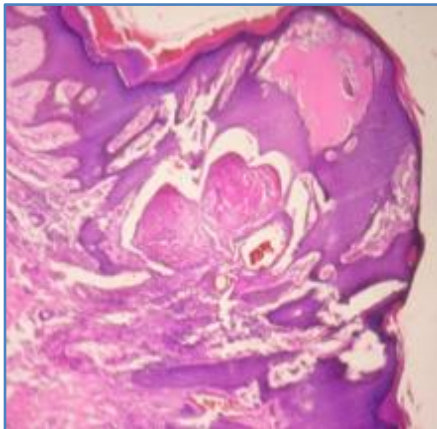


Fig. 8A: H & E stained 4 x

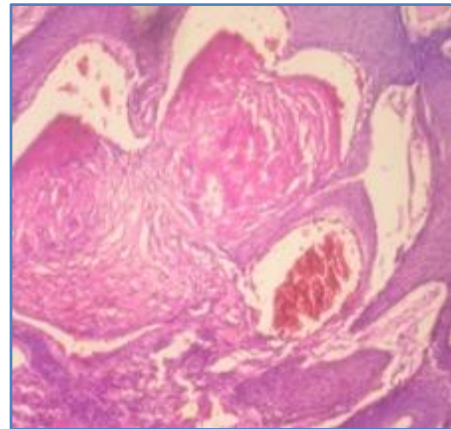


Fig. 8B: H & E stained 10 x

Plaque-like lesion, where the space is filled with recent thrombus.

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