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The Interposed Smudge-Kaposiform Haemangioendothelioma

Bajaj A*

Department of Pathology, AB Diagnostics, India

***Corresponding author:** Anubha Bajaj, Department of Pathology, AB Diagnostics, Rajauri Garden, New Delhi, India, Tel: +91 9811693956; Email: anubha.bajaj@gmail.com

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Abstract

Kaposiform hemangioendothelioma emerges as an exceptionally discerned endothelial cell neoplasm. Commonly, tumefaction disseminates within entire dermis with expansion into the subcutaneous tissue and exhibits localized tumour aggressiveness. Characteristically, proliferation of spindle shaped endothelial cells is commingled with lymphatic vessels demonstrating capillary haemangioma-like articulations. Glomeruloid structures are configured of discrete capillary lobules demonstrating a swirling pattern. Nodules of spindle shaped endothelial cells are encompassed by peripheral lymphatic vessels articulating lymphangioma-like areas. Tufted angioma configures as a benign, superficial, vascular neoplasm arising as a congenital lesion or appears within infants and paediatric population. The aggressive neoplasm is associated with enhanced morbidity and may undergo spontaneous retrogression, in contrast to Kaposiform hemangioendothelioma.

Keywords: Hemangioendothelioma; Herpesvirus; Retrogression; Lymphatic Vessels; Kasabach Merritt

Abbreviations: HHV-8: Herpesvirus-8; PTT: Partial Thromboplastin Time: PT: Prothrombin Time; MRI: Magnetic Resonance Imaging.

Introduction

Kasabach-Merritt phenomenon is intensely concurrent with Kaposiform hemangioendothelioma and tufted angioma. The neoplasm preponderantly appears at birth, within infancy or adolescence. Although tumours arising in adults are documented, an estimated ~50% lesions occur within first year of life [1,2].

Kaposiform haemangioendothelioma commonly incriminates cutaneous surfaces and subcutaneous tissue of extremities. Besides, soft tissue and non-cutaneous locales may be implicated wherein retroperitoneum and peritoneum are frequently involved. Sites as head and neck, mediastinum or trunk may delineate the lesions [1,2]. Of obscure aetiology and pathogenesis, tumefaction is posited to emerge from lymphatic endothelium which is immune reactive to vascular biomarkers as CD31 and CD34. Besides, immune reactivity to vascular endothelial growth factor receptor 3 (VEGFR3) or lymphatic markers D2-40 and PROX1 may be observed [1,2].

Genesis is postulated to be multifactorial with contribution of sporadic genomic mutations and various genetic factors. Neoplasm demonstrates genomic mutations within GNA14 gene. Tumefaction is non-concurrent with human herpesvirus 8 (HHV8) infections. Surgical intervention of lesions or trauma may engender Kasabach-Merritt phenomenon [2,3].

Implicated infants and children represent with cutaneous lesions composed of atypical, indurated, reddish purple plaques with inadequately defined perimeter. Occasional foci of telangiectasia along with or devoid of purpura or petechiae may be observed [2,3].Characteristically, lesion demonstrates superimposed foci of cutaneous hypertrichosis or hyperhidrosis. Predominantly confined to extremities, neoplasm may infrequently appear within the trunk, head and neck or mediastinum. Deep seated lesions confined to retroperitoneum or intrathoracic region may represent with bluish purpuric tinge superimposed upon the cutaneous surface and require distinction from coagulopathy or vascular disorder [2,3].

Tumour is associated with pain or functional impairment of implicated zone. Nearly 70% lesions, especially enlarged, congenital tumours or retroperitoneal or intrathoracic tumefaction concur with Kasabach-Merritt phenomenon comprised preponderantly of consumptive coagulopathy and thrombocytopenia [2,3]. Characteristically, Kaposiform haemangioendothelioma lacks spontaneous retrogression. Regional lymph node metastases is exceptional and distant metastases remain undocumented. Consequent to occurrence of Kasabach-Merritt phenomenon, associated mortality appears at ~10% [2,3].

Grossly

Cutaneous surface exhibits inadequately demarcated violaceous plaques admixed with occasional foci of telangiectasia and an absence of purpura or petechial spots. Deep seated soft tissue expounds multiple neoplastic nodules along with prominent desmoplastic reaction appearing as tumour infiltrate [2,3].

Upon microscopy, tumour is composed of sheets, nodules or abridged fascicles of spindle-shaped cells. Cellular component is commingled with slit-like vascular lumens. Infiltration into circumscribing stroma is observed. Constituent spindle-shaped cells delineate cellular swirls, thereby imparting a 'glomeruloid' neoplastic appearance [2,3].

Characteristic

Glomeruloid structures are impregnated with red cell fragments, intracellular hyaline granules, extracellular hyaline granules, hemosiderin pigment deposits or microthrombi indicative of red cell and platelet sequestration and destruction [3,4]. Lymphangioma-like zones are comprised of peripheral, thin walled, ectatic vascular articulations which circumscribe spindle-shaped tumour cell nodules. Occasionally, a discrete lymphangioma can be discerned. Well-formed vascular articulations are admixed with foci of pericytes. Mitotic figures may be discerned. Cytological atypia is minimal [3,4].

Tufted angioma incriminates the dermis with expansion into superimposed fascia and skeletal muscle. Lesion expounds multiple dermal lobules composed of intensely adherent spindle-shaped endothelial cells intermingled with slit-like lumens and disseminated tufts of capillaries, thereby delineating a 'cannonball' appearance. Lymphangioma-like areas are comprised of lymphatic vessels amalgamated upon periphery of spindle-shaped, endothelial cell clusters and nodules [3,4].

Ultrastructural examination depicts spindle-shaped endothelial cells layering slit-like vascular channels. Endothelial cells are permeated with plump nuclei protruding into vascular lumen. Circumscribing basement membrane appears attenuated and is incorporated withpericytes. Slit-like vascular channels are pervaded with platelets, lymphocytes and macrophages. Neoplastic cells are imbued with cytoplasmic phagocytic vesicles [3,4].

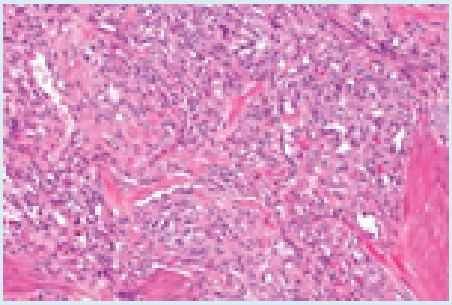

Figure 1: Kaposiform hemangioendothelioma delineating clusters and aggregates of spindle-shaped cells surrounded by thin walled, ectatic vascular articulations. Infiltration into circumscribing stroma is observed [5].

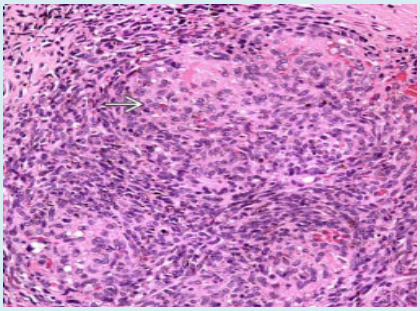

Figure 2: Kaposiform hemangioednothelioma demonstrating clusters and aggregates of spindle-shaped cells encompassed within thin walled ectatic vascular structures. Infiltration into surrounding stroma is exemplified [6].

Histological Subtype	Score
Atypical lipomatous tumour/Well differentiated liposarcoma	1
Well differentiated leiomyosarcoma	1
Malignant neurofibroma	1
Well differentiated fibrosarcoma	1
Myxoid liposarcoma	2
Conventional leiomyosarcoma	2
Conventional fibrosarcoma	2
Myxofibrosarcoma	2
High grade myxoid (round cell) liposarcoma	3
Pleomorphic liposarcoma	3
Dedifferentiated liposarcoma	3
Pleomorphic rhabdomyosarcoma	3
Poorly differentiated/ pleomorphic leiomyosarcoma	3
Biphasic/monophasic/poorly differentiated synovial sarcoma	3
Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Extraskeletal Ewing's sarcoma	3
Malignant rhabdoid tumour	3
Undifferentiated pleomorphic sarcoma	3
Undifferentiated sarcoma, not otherwise specified	3

Table 1: Differentiation of Soft Tissue Tumours [3,4].

Tumour cells appear immune reactive to endothelial cell markers as CD31, CD34, ERG or FL1. Peripheral lymphatic component delineating Kaposi-like areas appear immune reactive to PROX1 or podoplanin. However, glomeruloid structures appear devoid of aforesaid antigens. Neoplastic cells appear immune non-reactive to GLUT1 or HHV8.

Kaposiform haemangioendothelioma requires segregation from neoplasms as Kaposi sarcoma, infantile haemangioma or spindle cell haemangioma [7,8].

Kaposiform haemangioendothelioma may be appropriately ascertained with clinical representation wherein the lesion is indicated with atypical, indurated, reddish purple plaques with inadequately defined perimeter commonly confined to trunk and extremities. Infants are commonly implicated and lesion may be associated with lanugo hair hypertrichosis and enhanced sweating.

- Imaging techniques as ultrasonography which is beneficial for discerning miniature, superficial lesions [7,8].

  Magnetic resonance imaging (MRI) is optimal for evaluating deep seated, infiltrative neoplasms and demarcating the tumefaction from various vascular tumours along with assessing extent of disease and response to therapy. MRI is preferentially employed for evaluating severe unexplained coagulopathy and thrombocytopenia or coexistent cutaneous purpura which may concur with deep seated lesions of Kaposiform haemangioendothelioma [7,8].

Histological evaluation of the lesion is recommended for appropriate categorization. However, morphological assessment is not recommended when lesions concur with severe Kasabach-Merritt phenomenon [7,8].

For appropriate disease ascertainment, haematological parameters as complete blood count, platelet count, coagulation studies with prothrombin time (PT), partial thromboplastin time (PTT), fibrinogen or D dimer necessitate evaluation. Kasabach-Merritt phenomenon may demonstrate severe thrombocytopenia, hypofibrinogenemia and elevated D dimer levels. Individuals with acute disease require assessment of liver and renal function tests [7,8].

Upon magnetic resonance imaging (MRI), neoplasm

displays multi-planar extension with diffuse image enhancement and inadequately defined tumour perimeter.

- T1 weighted imaging exhibits iso-intense signal intensity, in contrast to adjacent skeletal muscle.
- T2 weighted imaging expounds hyper-intense signal intensity along with frequent alterations within adjacent bone or joints, destruction of bony cortex or epiphysis and infiltration of abutting joints [7,8].

Conclusion

Kaposiform haemangioendothelioma can be appropriately managed with diverse modalities as surgical eradication of lesion, pulsed dye laser or multiple pharmacological agents. Surgical extermination may be adopted with curative intent. However, comprehensive surgical resection may be challenging to achieve on account of factors as inadequately defined tumour perimeter, incrimination of vital organs or concordance of Kasabach-Merritt phenomenon. Symptomatic lesions and Kasabach-Merritt phenomenon may be alleviated with cogent pharmacological therapy with consequent decimation of tumour mass, amelioration of clinical symptoms and resolution of coagulopathies [7,8].

Intervention manoeuvers as embolization decimates tumour mass and may be beneficially adopted prior to commencement of precise pharmacological therapy. Radiation therapy appears advantageous for treating neoplasms refractory to diverse therapeutic modalities [7,8]. Typically, lesion is devoid of spontaneous retrogression. Surgical extermination of tumefaction appears preponderantly curative. Prognostic outcomes are intensely concurrent to factors as

- Site of tumour emergence
- Tumour magnitude
- Progression of consumptive coagulopathy

Subjects with Kasabach-Merritt phenomenon demonstrate associated mortality at ~10%. However, superior pharmacological therapy and supportive measures delineate decimated tumour associated mortality. Commonly, mortality ensues due to factors as haemorrhage, sepsis or multi-organ failure [7,8].

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5. Image 1 Courtesy: Research gate

6. Image 2 Courtesy: Basic medical key

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