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The Melded Conglomerate-Composite Hemangioendothelioma

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Abstract

Composite hemangioendothelioma is an infrequent vascular neoplasm characteristically comprised of complex intermingling of benign, low grade malignant and malignant vascular components and intermediate biological behaviour. Preponderantly confined to distal extremities, especially hands and feet, neoplasm commonly implicates the adult population. The inadequately circumscribed lesion is confined to deep seated dermis or subcutaneous tissue and represents with an admixture of vascular lesions as epithelioid hemangioendothelioma, retiform hemangioendothelioma, low grade angiosarcoma, lymphangioma or variants of haemangioma as spindle cell haemangioma, capillary haemangioma, cavernous haemangioma, hobnail haemangioma or epithelioid haemangiom.

Keywords: Commingled Vascular Lesions; Intermediate Behaviour

Abbreviations: HPF: High Power Fields; PLC: Pseudo-Lipoblastic Countenance.

Introduction

Composite hemangioendothelioma emerges as an exceptionally discerned vascular neoplasm associated with intermediate biological behaviour. Characteristically, tumefaction is comprised of complex intermingling of benign, low grade malignant and malignant vascular components. The uncommon vascular neoplasm of intermediate malignant potential is exceptionally accompanied by regional lymph node and distant metastasis. Upon low power examination, tumefaction demonstrates significant morphologic variation. Segregation of composite hemangioendothelioma from various vascular neoplasms may be challenging although the neoplasm exhibits significantly variable vascular histological configurations within a singular lesion. Tumefaction may arise as a congenital lesion, within infancy, paediatric subjects or adolescents. However, adults are predominantly implicated and neoplasm may be observed in up to eighth decade [1,2].

Majority of lesions are situated upon distal extremities, especially hands and feet. Additionally, sites such as head and neck, dorsal region, mediastinum, bone, inguinal lymph nodes, renal parenchyma or spleen may be implicated [1,2]. Of obscure aetiology, individuals delineating composite hemangioendothelioma may exhibit history of lymphedema. Tumefaction may concur with Kasabach-Merritt or Maffucci's syndrome. Clinically, lesions of extended duration appear multinodular and demonstrate a reddish blue hue [1,2]. Grossly, composite hemangioendothelioma manifests as a nodular tumefaction of bluish, purple or reddish blue hue. Multiple nodules may be discerned. Tumefaction appears to infiltrate circumscribing soft tissue [2,3].

Upon microscopy, an inadequately circumscribed lesion appears to be confined to deep seated dermis or subcutaneous tissue. Tumefaction may infiltrate into encompassing soft tissue [2,3]. Upon low power examination, morphological features appear to vary between diverse tumour zones [3,4].

Neoplasm demonstrates admixture of divergent vascular components and may represent commingling of epithelioid hemangioendothelioma, retiform hemangioendothelioma, low grade angiosarcoma, lymphangioma or variants of haemangioma as spindle cell haemangioma, capillary haemangioma, cavernous haemangioma, hobnail haemangioma or epithelioid haemangioma [3,4]. Aforesaid diverse components appear intensely intermingled and segregation of various 'pure' vascular components may be challenging. Tumour cells appear as epithelioid endothelial cells and demonstrate vacuolated, pseudo-lipoblastic appearance. Mitotic activity is minimal [3,4].

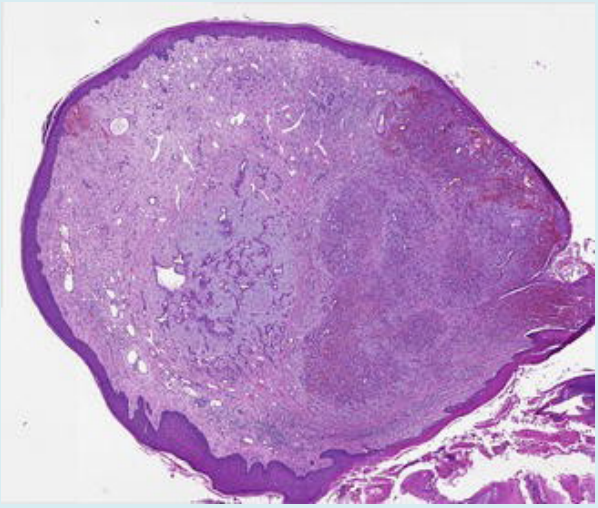

Figure 1: Composite hemangioendothelioma composed of variable proportion of spindle cell, capillary and cavernous haemangioma. Epithelioid endothelial cells exhibit a vacuolated, pseudo-lipoblastic countenance.

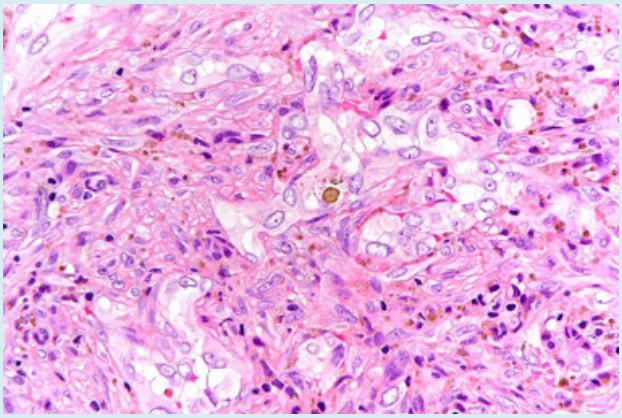

Figure 2: Composite haemangioendothelioma delineating a variable proportion of cavernous, capillary and spindle cell haemangioma component. Epithelioid endothelial cells exhibit a vacuolated, pseudo-lipoblastic countenance.

Mitotic score	Mitosis/10 HPF	Mitosis / mm^2
Score 1	0-9 mitosis/10 HPF	0-5 mitosis/mm^2
Score 2	10-19 mitosis/ HPF	6-11 mitosis/mm^2
Score 3	>19 mitosis /10 HPF	>11 mitosis/mm^2

HPF: High Power Fields.

Table 1: Mitotic score of soft tissue sarcomas.

Mitotic count is predominantly calculated from mitotically active areas, devoid of tumour necrosis. Mitosis may be quantified within 10 consecutive high power fields (HPF) upon 40x objective or 1 HPF x 400 = 0.1734 mm^2 area wherein appropriate high power fields encompassing 1 mm^2 area is contingent to individual microscope.

Score	Tumour Necrosis
Score 0	Absence of tumour necrosis
Score 1	<50% tumour necrosis
Score 2	≥50% tumour necrosis

Table 2: Tumour score associated with necrosis.

Tumour necrosis is appropriately evaluated upon cogent gross examination and categorized upon histological sections.

Tumour cells appear immune reactive to CD31, CD34, ERG, FLI1 or factor VIII. Tumour cells appear immune non-reactive to human herpes virus 8(HHV8) [5,6].

Composite hemangioendothelioma requires segregation from neoplasms as retiform hemangioendothelioma, Dabska type hemangioendothelioma, Kaposiform hemangioendothelioma, epithelioid hemangioendothelioma, Kaposi's sarcoma or angiosarcoma [5,6]. Cogent histological evaluation is an optimal, recommended methodology for obtaining a definitive diagnosis. Appropriate tumour discernment upon examination of miniature tissue samples may be challenging on account of varied morphological spectrum expounded by the neoplasm [5,6]. Optimal and recommended therapeutic guidelines remain obscure on account of infrequency of the neoplasm. However, surgical extermination of the neoplasm may be adopted with curative intent. Localized radiotherapy and chemotherapy may be employed for treating reoccurring and metastatic disease [5,6].

Prognostic outcomes are contingent to localized tumour reoccurrence and tumour aggressiveness appearing in ~50% of neoplasms. Localized tumour reoccurrence may occur within 18 months to 10 years following initial surgical excision. Proportionate regional lymph node or

distant metastasis is minimal. Tumour reoccurrence or distant metastasis may occur regardless of angiosarcoma-like foci discerned upon morphological evaluation [5,6]. Irrespective of variable morphological spectrum expounded by the neoplasm, the biological behaviour remains identical. Precise tumour discernment is crucial for accurate therapy and ascertainment of prognostic outcomes [5,6].

Conclusion

Tumour cells appear immune reactive to CD31, CD34, ERG, FLI1 or factor VIII and immune non-reactive to human herpes virus 8(HHV8). Composite hemangioendothelioma requires segregation from neoplasms as retiform hemangioendothelioma, Dabska type hemangioendothelioma,Kaposiformhemangioendothelioma, epithelioid hemangioendothelioma, Kaposi's sarcoma or angiosarcoma. Cogent histological evaluation is an optimal, recommended methodology for obtaining a definitive diagnosis. Optimal, recommended therapeutic guidelines remain obscure on account of infrequency of the neoplasm. However, surgical extermination of the neoplasm may be adopted with curative intent.

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