Case Report

COMPOSITE HAEMANGIOENDOTHELIOMA WITH ZYGOMYCOTIC NECROTIZING FASCITIS OF RIGHT LEG: A RARE CASE REPORT

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INTRODUCTION

Composite Haemangioendothelioma with zygomycotic necrotizing fascitis is a very rare infective disease. Haemangioendothelioma is used to described as a group of vascular neoplasms and can be classified as Epithelioid, Composite, Spindle cell, Retiform, Kaposiform and Infantile haemangioendothelioma.

Composite HE was first explained by Nayler et al. in 2000. Most significantly, it is the very rare low grade vascular tumour with an admixture of concurrent various benign vascular lesions (e.g., spindle cell haemangiomias), intermediate (e.g., retiform HE), and malignant (e.g., angiosarcoma-like). Retiform and epithelioid HEs represent the two most common histological types of composite HE. In composite HEs Local relapses tend to occur several years after tumour resection, however the potential of distal and regional nodal metastases is considerable.

There were only few of previously published congenital composite HEs that had been noted since birth with longstanding presentation before excision. Exceptionally, composite HE may also arise from atypical locations, particularly the oral cavity or comitant or with certain syndromes or diseases, such as Maffucci syndrome, Kasabach-Merritt syndrome, lymphoedema.

Necrotizing Fascitis is most oftenly caused by flesh eating bacteria that is Streptococcus Pneumoniae but only small number of cases of Necrotizing fascitis were caused by Zygomycetes were reported till date.

Zygomycotic Necrotizing Fascitis is mainly found in the patients with history of trauma, Immunosuppressed, diabetics, alcoholics, i.v. drug abusers and patients with peripheral vascular disease. They can occur at any part of the body, abdominal wall, perineum and extremities are the most common site of the infection. They damage the blood vessels due to their highly invasive nature by invading with hyphae and may lead to emboli formation and ultimately causes necrosis of surrounding tissue.

Case Report

A 52 years male presented to the General Surgery Outpatient with multiple ulceroproliferetive lesions over right leg involving anteromedial and anterolateral aspect extending from ankle joint to knee joint with multiple small nodes on the medial aspect of thigh for 7 months. Left lower limb was normal. It was associated with blood mixed purulent discharge. Initially haemoglobin was 4.3mg/dl so 4 unit blood was transfused on consequent days. Liver function test, Renal Function test, blood sugar, X ray chest and ECG were with in normal limits.

USG colour Doppler of right lower limb shows moderate to gross subcutaneous edema in right leg & feet region mainly with few enlarged right inguinal lymph nodes. Mycology report (KOH Mount) was positive for fungal hyphae for Zygomycetes species. He was planned for serial surgical debridement, intravenous Amphotericin B, close monitoring including hyperbaric oxygen therapy.

Histopathological report confirms the possibility of composite haemangioendothelioma with epithelioid and spindle cell variants with CD31 & CD34 markers were diffusely positive and Cytokeratin & desmin were negative in immunohistochemistry.

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ABSTRACT

A 52 years male presented to the General Surgery Outpatient with multiple ulceroproliferetive lesions over right leg involving anteromedial and anterolateral aspect extending from ankle joint to knee joint with multiple small nodes on the medial aspect of thigh for 7 months. Left lower limb was normal. It was associated with blood mixed purulent discharge. Initially haemoglobin was 4.3mg/dl so 4 unit blood was transfused on consequent days. Liver function test, Renal Function test, blood sugar, X ray chest and ECG were with in normal limits.
Composite hemangioendothelioma with Zygomycotic necrotizing fascitis is very rare association and presently, no case reported till date. Composite HE is the very rare low grade vascular tumour with an admixture of concurrent various vascular lesions, e.g. spindle cell haemangiomas, retiform HE and angiosarcoma.\(^3,4\) Retiform and epithelioid HEs represent the two most common histological types of composite HE.\(^3,4\) in composite HEs Local relapses tend to occur several years after tumour resection, however the potential of distal and regional nodal metastases is considered very low.\(^3,5\)

In present case the composite haemangioendothelioma of right leg comprises of hyperplastic squamous epithelium with epithelioid and spindle cell variants with vascular channeles containing lobules with full of endothelial cells hobnailing admixed with spindle cells. Most of these tumors have been found on the distal extremities. Exceptions include the tongue, mandibular vestibule, achilles tendon, axilla, thigh, upper arm, cheek mucosa, upper back, mediastinum, hypopharynx, scalp, nose and kidney.\(^12\)

Necrotizing fascitis mainly caused by beta haemolytic Streptococci and Zygomycotic fungal infection is very uncommon. Human Zygomycosis is caused by order Mucorales (3 orders of Zygomycetes- Mucorales, Mortierellales & Entomophthorales) and it used to occur in immunocompromised hosts as opportunistic infections.\(^13\) Histologically Zygomycetes class of fungi exhibit broad thin walled, hyaline, often asperate or pauciseptate hyphae with frequent angioinvasion and accounts for weaker staining with Gomori methanamine silver& PAS techniques comparative to fungi with thickened walls.\(^14\)

Vascular invasion that causes necrosis of infected tissue & perineural invasion are most morbid features of these infections.\(^15\)

**CONCLUSION**

This case illustrates a very rare association of composite haemangioendothelioma with zygomatic necrotizing fascitis of leg. Although surgical resection and antifungal medications can be considered but definitive treatment strategy for this condition is still need to be find out.
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