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Editorial

The Thriving Consort-Epstein Barr Virus Associated Smooth Muscle Tumour

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Immunodeficiency is accompanied by enhanced incidence of various neoplasms wherein tumefaction is associated with diverse genetic disorders, acquired immunodeficiency syndrome (AIDS) or meticulous immunosuppressive therapy adopted following solid organ transplantation as renal transplant.

Post transplant smooth muscle tumour may be categorized into distinctly malignant tumours as leiomyosarcoma or 'borderline' neoplasms exhibiting minimal cellular or nuclear pleomorphism, cellular crowding, and mitotic activity and an 'uncertain biological potential'.

Epstein Barr virus (EBV) associated smooth muscle tumour is an exceptionally discerned, variably aggressive neoplasm commonly encountered within immunocompromised subjects. Epstein Barr virus associated smooth muscle tumour appears morphologically distinct from classic smooth muscle tumours of soft tissue.

Precise disease incidence remains obscure although ~ 1% subjects with immunodeficiency may be incriminated. Average duration of tumour emergence following organ transplantation is ~4 years [1,2].

Tumour discernment with conventional histological methods may be challenging as multifocal neoplasms are engendered on account of multiple and sequential viral infection rather than configuration of as metastatic tumefaction. Frequently non fatal, tumour related mortality is encountered within ~8% instances.

Epstein Barr virus(EBV) associated smooth muscle tumour commonly arises within adult population. A male predominance is observed with male to female proportion of 1.5:1 [1,2].

The infrequently discerned post transplant smooth muscle tumour emerges as a distinct entity confined within peripheral soft tissue, intracranial space or various visceral sites. Following liver transplantation, discernible lesions compatible with Epstein Barr virus associated smooth muscle tumour may manifest as colonic polyposis or appear confined to hepatic parenchyma or pulmonary parenchyma [1,2].

Majority (>50%) of neoplasms represent with multiple lesions wherein multifocal tumour dissemination appears as a consequence of multiple infectious events, in contrast to distant metastases arising from singular neoplastic site. Tumour multiplicity is encountered wherein multiple smooth muscle neoplasms engendered due to Epstein Barr virus infection concordant with a singular neoplasm appear as disparate lesions upon clone specific evaluation [1,2].

Of obscure pathogenesis, Epstein Barr virus associated smooth muscle tumour emerges due to infection with Epstein Barr virus which is a DNA herpes virus imbued with a potential to immortalize infected cells. It is posited that Epstein Barr virus associated smooth muscle tumour may originate from smooth muscle cells confined within blood vessel walls. Accompanying immunosuppression may permit an anomalous transfer of Epstein Barr virus into smooth muscle cells with consequent occurrence of a latent infection and neoplastic configuration along with altered cytogenetic influences [1,2].

Epstein Barr virus (EBV) infection is posited to engender Epstein Barr virus associated smooth muscle tumour wherein

gastrointestinal tract, hepatic parenchyma or pulmonary parenchyma may be incriminated following liver transplantation [2,3].

The infrequent, gradually progressive, locally invasive Epstein Barr virus associated smooth muscle tumour is commonly confined to hepatic parenchyma. Besides, pulmonary parenchyma, renal parenchyma, lymph nodes, adrenal gland, spleen, heart and uncommonly the central nervous system may be implicated. Subjects demonstrating autoimmune deficiency syndrome may exhibit multifocal tumours confined to hepatic parenchyma, spleen, brain, spinal cord or adrenal gland [2,3].

Generally, disease progression is gradual. Besides, immunocompromised subjects demonstrating Epstein Barr viral infection may denominate malignant mesenchymal neoplasms as leiomyosarcoma [2,3].

Grossly, multiple, miniature, submucosal, polypoid lesions may be encountered [2,3].

Upon microscopy, a well differentiated smooth muscle neoplasm is comprised of spindle shaped cells configuring a storiform pattern along with an admixture of scattered small T lymphocytes. Tumefaction is composed of zones of primitive round cells intermingled with prominent infiltration of intratumoral T lymphocytes [3,4].

Cellular and nuclear atypia may be minimal or absent. Mitotic figures are exceptional to absent. Neoplasm is devoid of cellular or nuclear pleomorphism [3,4].

Morphologically, incriminated mucosa exhibits a neoplasm demonstrating features of smooth muscle tumour. Tumefaction is composed of intersecting fascicles of monotonous, spindle shaped cells pervaded with abundant, eosinophilic cytoplasm, cigar shaped nuclei and indistinct cellular perimeter. Cellular and nuclear atypia is mild. Mitotic activity may be minimally elevated. Tumour necrosis is absent.

Occasionally, neoplasm may exhibit features of high grade sarcoma [3,4].

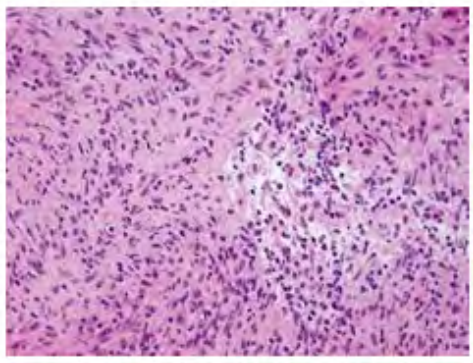

Figure 1: EBV associated smooth muscle tumour demonstrating an admixture of spindle shaped cells and primitive round cells with infiltrating lymphocytes. Tumour cells configure a storiform pattern. Cellular and nuclear pleomorphism is absent [6].

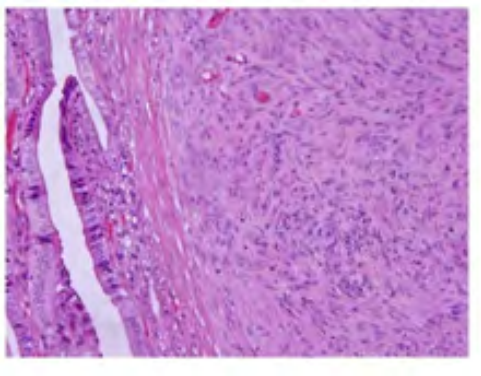

Figure 2: EBV associated smooth muscle tumour exhibiting fascicles of spindle shaped cells commingled with primitive round cells and infiltrating lymphocytes. Cellular and nuclear pleomorphism is absent [7].

Epstein Barr virus associated smooth muscle tumour is immune reactive to smooth muscle actin (SMA), desmin and Epstein Barr virus encoded small ribonucleic acid 1 (EBER1).

Upon in situ hybridization, early Epstein Barr virus ribonucleic acid (RNA) may be discerned [4,5].

Tumefaction is immune non reactive to angiogenic markers as CD31, CD34, CD117 or human melanoma black 45 (HMB45) antigen.

Ki67 proliferative index may be elevated to ~15% [4,5].

Ultrasonography exhibits a solid tumefaction confined to various sites. Contrast induced spiral chest computerized tomography (CT) and abdominopelvic computerized tomography (CT) exhibits ring enhancement of solid lesions confined to specific sites.

Colonoscopy may depict multiple, miniature, raised, polypoid lesions disseminated within the rectum and colon [4,5].

Epstein Barr virus encoded small RNAs chromogenic in situ hybridization (EBER CISH) may exhibit Epstein Barr virus RNA confined within tumour cell nuclei and immunoblasts of abutting lymph node, as discerned with tissue sampling of prospective Epstein Barr virus associated smooth muscle tumour [4,5].

In situ hybridization (ISH) and polymerase chain reaction (PCR) can be optimally employed to demonstrate Epstein Barr viral infection by discerning Epstein Barr viral ribonucleic acid(RNA) [4,5].

Serological assays can be adopted to confirm preceding Epstein Barr viral infection. However, the assay appears inadequate for cogent diagnosis, monitoring of disease reoccurrence or evaluating disease burden.

Epstein Barr virus associated smooth muscle tumour can be appropriately managed with cogent surgical extermination of the neoplasm [4,5].

Adoption of precise chemotherapy may be beneficial.

Amelioration of immune status of incriminated subject is advantageous. Occasionally, tumour associated mortality may ensue.

Unifocal lesions may be appropriately alleviated with comprehensive surgical eradication of the neoplasm with resection of tumour free surgical margins [4,5].

Reduction of immunosuppressive agents may permit Epstein Barr virus specific cytotoxic T cells to proliferate. However, concomitant graft rejection may ensue [4,5].

Antiviral drugs which decimate Epstein Barr viral load, cyclosporine or sirolimus as an oncogenesis inhibitor may ameliorate disease control with superior therapeutic outcomes [4,5].

Antiviral therapy, adjuvant chemotherapy and radiotherapy appear advantageous for treating neoplasms unamenable to surgical resection [4,5].

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6. Image 1 Courtesy: Research gate.

7. Image 2 Courtesy: Hindawi.com.