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The Concrete Paltriness- Calcifying Fibrous Tumour

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1. Abstract

Calcifying fibrous tumour is an exceptional, benign, soft tissue neoplasm initially scripted by Rosenthal et al in 1988 as a "childhood fibrous tumour with psammoma bodies" [1]. The neoplasm was subsequently nomenclated as "calcifying fibrous tumour" by Larson et al [2].

Calcifying fibrous tumour (CFT) is a distinctive, solitary, hypo-cellular neoplasm emerging from subcutaneous fibrous soft tissue. The neoplasm exhibits a proliferation of bland, spindle-shaped fibroblasts enmeshed within dense, hyalinised collagenous tissue, intermingled with an infiltrate of mature lymphocytes and plasma cell along with foci of dystrophic calcification or incorporated calcific, psammoma-like bodies [3].

However, paucity of information on clinical representation and pathogenesis about the exceptional neoplasm persists.Tumour discernment may be challenging as calcifying fibrous tumour histologically recapitulates several mesenchymal neoplasms. Nevertheless, appropriate histological assessment is definitive and categorical.

2. Disease Pathogenesis

Of obscure pathogenesis, diverse hypothesis is denominated in the genesis of calcifying fibrous tumour. Posttraumatic tumour emergence, genetic anomalies, preceding infection, surgical procedures or concurrence with immunoglobulin 4 (IgG4)- related disease and inflammatory myofibroblastictumour (IMT) is implicated [3, 4].Calcifying fibrous tumour may represent diverging phases of immunoglobulin G4 (IgG4)- related disease and manifest an immunoglobulin G4 (IgG4)- related pseudo-tumour. Identically, gastrointestinal calcifying fibrous tumour is contemplated to be a gastrointestinal lesion of immunoglobulin G4 (IgG4)- related disease [3].

The neoplasm was contemplated to be a reactive process on account of an abundance of accompanying inflammatory cells or emerging as a terminal phase of inflammatory myofibroblastictumour. It was subsequently classified as a soft tissue tumour and nomenclated as calcifying fibrous tumour secondary to multifocal sites of tumour incrimination and a potential for tumour reoccurrence [4].

Calcifying fibrous tumour is morphologically concordant with inflammatory myofibroblastictumour and may be contemplated as a terminal stage of the neoplasm. Genomic methylation profiling of calcifying fibrous tumour and inflammatory myofibroblastictumour delineate an identical epigenetic profile although characteristic genetic fusion ALK, ROS1 and RET, as discerned in inflammatory myofibroblastictumour, is absent in calcifying fibrous tumour [3,4].

Calcifying fibrous tumour is a tumefaction of controversial neoplastic disposition. Novel, deleterious chromosomal mutations of ZN 717, FRG1 or CDC27 gene are discerned in association with copy number deletion upon chromosome 8 and extensive loss common upon chromosome 6.

Whole exome sequencing of calcifying fibrous tumour is posited to display loss of copy number of genes within chromosome 6 and 8, associated with genesis of the neoplasm. The neoplasm harbours specific genetic mutations accompanied by a capacity for distant metastasis [3,4].

3. Disease Characteristics

The neoplasm occurs in adolescents and young adults and delineates a distinctive age of tumour emergence from birth to 4 years

followed by second to third decade between 25 years to 34 years and an average age of disease occurrence at 33.58 years.

Tumefaction denominates a female preponderance with a female to male proportion of 1.27:1 [4,5]. Calcifying fibrous tumour predominantly emerges as a solitary neoplasm in diverse locations such as gastrointestinal tract, pleura, peritoneum, abdominal cavity, solid viscera or neck. The non-invasive tumefaction arises from deep-seated soft tissue and is frequently discerned within stomach or small intestine [4].

Commonly incriminated sites are stomach (18%), pleura (9.9%), small intestine (8.7%), peritoneum (6.8%), neck (6.2%), mediastinum (5%) or mesentery (5%). Nevertheless, no site of disease emergence is exempt and multifocal tumours are documented. Accurate assessment of tumour incidence of the exceptional neoplasm is challenging [3,5].

4. Clinical Elucidation

An estimated 70% of calcifying fibrous tumours are asymptomatic and emerge as incidental discoveries.

Symptomatic tumefaction may represent nonspecific features such as lack of appetite, fever, weight loss, fatigue, progressive weakness, dyspnoea, chest or back pain. Alternatively, specific clinical symptoms may arise pertaining to site of tumour incrimination wherein involvement of chest wall depicts discomfort, pain, dyspnoea, cough or wheezing secondary to compression of airways [4, 5].

Pleural neoplasms are uncommon and may not incriminate pulmonary parenchyma. Pleural lesions are accompanied with stippled calcification. Enlarged neoplasms of pulmonary parenchyma are associated with obstructive symptoms arising on account of airway compression and prospective tumour progression [4,5].

Tumours arising within gastrointestinal tract exceptionally manifest complications such as bowel obstruction, intussusception or ischemic bowel necrosis [5].

The non-aggressive, enlarged neoplasm is devoid of invasion of circumscribing soft tissue. Non infiltrative entrapment of nerve bundles is observed [5].

5. Histological Elucidation

On gross examination, the neoplasm is solitary or multiple, well circumscribed, un-encapsulated, solid, spherical, lobulated, firm to hard, smooth with a grey/ white extraneous surface, mass of variable magnitude which may adhere to circumscribing soft tissue although localized tumour infiltration is absent. Partially translucent or opaque, firm or rubbery tumour fragments with whitish flecks or pink areas are discerned [5,6].

Macroscopic magnitude of calcifying fibrous tumour of the gastrointestinal tract varies from 0.5 centimetres to 11 centimetres with an average diameter of 2.6 centimetres. Cut surface is solid, gritty, white or grey with yellowish foci of calcification [5,6].

On microscopy, the pauci-cellular neoplasm demonstrates abundant, hyalinised collagen intermingled with sparse chronic inflammatory infiltrate composed of mature lymphocytes and plasma cells along with foci of normal or dystrophic calcification. Tumour cells are intermingled with abutting soft tissue with an absence of cellular invasion [5,6].

Characteristic morphological features of the hypo-cellular calcifying fibrous tumour are abundant, hyalinised collagen, foci of dystrophic calcification, psammoma-like bodies and an interspersed mononuclear inflammatory infiltrate. Intervening collagenous matrix often depicts a storiform pattern although a “pattern-less” configuration can be observed [5,6].

Dense, collagenous matrix with scattered foci of dystrophic calcification, psammoma-like bodies, aggregates of plasma cells, mature lymphocytes, minimal mast cells and eosinophils is exhibited. Spindle-shaped cells with an abundant, eosinophilic cytoplasm are embedded within the collagenous matrix. Tumour cell nuclei are elliptical and vesicular with fine chromatin and inconspicuous nucleoli. Cellular or nuclear atypia or mitotic figures are absent [5,6].

Pauci-cellular, fibroblastic proliferation is exemplified by uniform, spindle-shaped cells enmeshed within dense, collagenous tissue. Variable quantification of mature lymphocytes with possible configuration of lymphoid follicles and plasma cells is enunciated. Scattered foci of dystrophic or psammoma-like calcification accompanies the cellular exudates [5,6].

Stromal calcification and an inflammatory exudate comprised of mature lymphocytes and plasma cells are preponderant morphological manifestations of calcifying fibrous tumour.

On ultrastructural examination, immature fibroblastic cells, collagen fibrils and foci of dystrophic or psammoma-like calcification is exhibited [5,6].

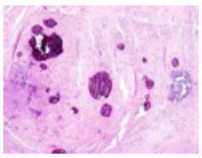

Figure 1: Calcifying fibrous tumour depicting bland, fibroblastic cells interspersed in dense, collagenous stroma and foci of dystrophic calcification [9].

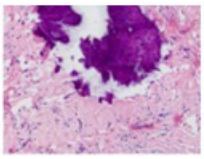

Figure 2: Calcifying fibrous tumour delineating uniform, fibroblastic cells intermixed in dense collagenous tissue and foci of psammoma-like calcification[10].

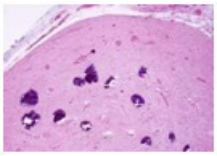

Figure 3: Calcifying fibrous tumour exhibiting bland, fibroblastic cells intermingled with dense collagen and focal dystrophic calcification [11].

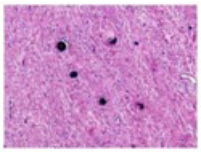

Figure 4: Calcifying fibrous tumour exemplifying uniform, spindle-shaped cells commingled with hyalinised collagen and foci of dystrophic calcification [12].

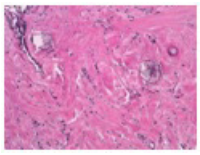

Figure 5: Calcifying fibrous tumour displaying uniform fibroblasts admixed with dense collagenous tissue and stippled calcification [13].

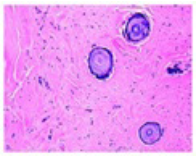

Figure 6: Calcifying fibrous tumour demonstrating bland fibroblasts commingled with dense collagenous tissue and psammoma-like calcification [14].

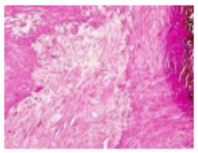

Figure 7: Calcifying fibrous tumour exemplifying uniform fibroblasts admixed within dense collagenous tissue and stippled calcification [15].

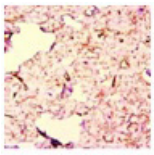

Figure 8: Calcifying fibrous tumour immune reactive to CD34 [16].

6. Immune Histochemical Elucidation

The neoplasm is immune reactive to CD34, factor XIIIa, B cell lymphoma 2 (Bcl-2), vimentin and focally immune reactive to CD99. Plasma cells are immune reactive to CD138.

Spindle-shaped cells are intensely and diffusely immune reactive to vimentin and factor XIIIa. The neoplasm is variably immune reactive to CD34 and infrequently immune reactive to smooth muscle actin (SMA). Elevated immunoglobulin G to immunoglobulin G4 (IgG: IgG4) proportion is discerned within the plasma cells, thereby indicating a concurrence between immunoglobulin G4 (IgG4) -related disease and calcifying fibrous tumour (3,4). The neoplasm is immune non-reactive to smooth muscle actin (SMA), muscle specific actin (MSA), myosin, desmin, CD117, anaplastic lymphoma kinase 1(ALK-1), S100 protein, cytokeratin (CK 8/18), discovered on GIST-1(DOG1) or β- catenin. Tumour proliferative index Ki-67 is usually below<1% [3,4].

7. Differential Diagnosis

Calcifying fibrous tumour requires a histological demarcation from diverse spindle-shaped cellular tumours such as sclerosing, calcified gastrointestinal stromal tumour (GIST), schwannoma, hyalinised leiomyoma, immunoglobulin G4 (IgG4)- related sclerosing disease, inflammatory myofibroblastictumour (IMT), solitary fibrous tumour and reactive nodular fibrous pseudo-tumour.

•Inflammatory myofibroblastictumour is a cellular neoplasm devoid of calcification. It is immune reactive to anaplastic lymphoma kinase (ALK), actin, CD34 and focally immune reactive to factor XIIIa [6, 7].

•Gastrointestinal stromal tumour (GIST) is a cellular lesion accompanied with minimal quantities of inflammatory exudate. Hyalinised collagenous stroma and focal calcification is absent. The neoplasm is immune reactive to CD117and DOG1 [6,7].

•Calcifying aponeurotic fibroma is a miniature, cellular neoplasm usually situated within distal locations or extremities.

•Desmoplastic fibroblastoma is a minimally cellular tumefaction arising in elderly subjects. The neoplasm is composed of enlarged, prominent fibroblasts. Micro-calcification and preponderant inflammatory infiltrate is usually absent [6,7].

•Idiopathic retroperitoneal fibrosis and related sclerosing, fibroinflammatory lesions are associated with significant inflammation, especially infiltration of plasma cells and eosinophils [6,7].

Cogent diagnosis mandates elimination of common neoplasms with pertinent morphological assessment, immunohistochemistry and genetic assay. Characteristically, calcifying fibrous tumour is a well circumscribed, pauci-cellular neoplasm with an encompassing collagenous matrix, bland, spindle-shaped cells, focal calcification along with disseminated, chronic lymphocytic and plasma cell inflammatory infiltrate [7].

8. Investigative Assay

Imaging evaluation of calcifying fibrous tumour is concordant to histological composition of the neoplasm. Upon ultrasonography and computerized tomography (CT), a characteristic, well circumscribed mass with disseminated foci of calcification is denominated [7,8].

Endoscopic ultrasonography of calcifying fibrous tumour exhibits hypoechoic nodules with dispersed, hyperechoic areas concordant to calcification [8].

Computerized tomography depicts an enlarged, irregular tumefaction extending into abutting soft tissues. The neoplasm is associated with significant intrinsic calcification [7,8].

Magnetic resonance imaging (MRI) demonstrates an isoechoic signal intensity upon gadolinium-contrast enhanced T1 weighted imaging. Hypoechoic signal intensity is delineated upon T2 weighted imaging [7,8].

9. Therapeutic Options

Initiation of therapy may be challenging with asymptomatic tumefaction, incidental tumour discernment or rapid tumour progression with consequent compression of adjacent organs or viscera. Calcifying fibrous tumour is appropriately alleviated with localized surgical excision [7,8].

Localized surgical extermination with a tumour- free perimeter of uninvolved soft tissue is the optimal treatment strategy. Prompt discernment of tumefaction and treatment is advantageous. Laproscopic surgical manoeuvers are beneficial for miniature lesions [7,8].

Extensive surgical eradication, elimination of incriminated pleura or pulmonary parenchyma with chest wall reconstruction is required for enlarged tumefaction. Complications engendered with surgical manoeuvers may require additional surgical procedures[7, 8].

Pulmonary tumefaction can be eradicated with pertinent surgery or wedge resection with concurrent tissue reconstruction. Pulmonary nodules necessitate and are adequately treated with multiple surgical procedures. Lethal complications mandate efficacious postoperative management wherein alleviation of clinical symptoms is necessitated.

Prognostic outcomes are excellent although localized tumour reoccurrence is documented. Extended follow up may be required to detect localized tumour reoccurrence which is delineated in an estimated 10% instances. Distant metastasis or tumour mortality is absent with extended survival of 100% [7,8].

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