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Essay Paper

The Maladjusted Crystallization-Non Ossifying Fibroma

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Preface

Non ossifying fibroma (NOF) is a benign neoplasm of fibrous tissue origin appearing on account of dysfunctional ossification. The neoplasm is additionally designated as metaphyseal fibrous defect, non-osteogenic fibroma, cortical desmoid, fibrous cortical defect, fibromatosis or fibroxanthoma.

The benign neoplasm is frequently observed in childhood, resolves spontaneously and lacks a malignant potential. Upon cogent imaging, several benign tumours such as non-ossifying fibroma and non-neoplastic conditions can recapitulate malignant bone neoplasms and necessitate appropriate segregation.

Disease characteristics

Non ossifying fibroma is a benign neoplasm arising within metaphysis of long bones and is frequently discerned around the knee joint. Tumefaction is commonly delineated in children and adolescents with an estimated incidence of 20% to 30% [1,2].

As non-ossifying fibroma occurs in children with skeletal immaturity, it is frequently delineated in children between 5 years to 15 years. The condition depicts a male predominance with a male to female proportion of 2:1 [1,2].

Tumefaction is commonly located within the metaphysis of long bones. Around 80% tumours are confined to the lower extremity wherein tumours within distal femur are common, followed by proximal tibia and distal tibia. Nevertheless, tumefaction is uncommonly exemplified within proximal femur and proximal humerus. Roughly 50% neoplasms are multiple and below one centimetre magnitude [1,2].

The neoplasm is posited to arise on account of an aberrant, osteoclastic bone resorption occurring within the sub-periosteum during metaphyseal remodelling. The condition is also hypothesized to be a developmental, non-neoplastic defect which undergoes spontaneous resolution and gradual replacement with cortical bone [1,2].

Non ossifying fibroma is associated with conditions such as neurofibromatosis, aneurysmal bone cyst, familial multifocal non ossifying fibroma and the congenital Jaffe-Campanacci syndrome composed of multiple non ossifying fibromas, café au lait pigmentation, mental retardation along with incrimination of genitalia, cardiac and ocular tissue [1,2].

Incidence of pathologic fractures emerging as a complication of non-ossifying fibroma within the lower extremity is around 90%. Nearly 50% neoplasms occur within distal tibia. However, a pre-existing non ossifying fibroma is exceptionally associated with pathological fracture of the femur [1,2].

An enhanced possibility of pathologic fractures is observed with incrimination of transverse diameter of bone exceeding > 50% or tumour magnitude in excess of > 33 millimeters within weight bearing zones of femur and tibia with non-ossifying fibroma [1,2].

Clinical elucidation

Non ossifying fibroma is a frequently discerned benign fibrous lesion appearing within metaphyseal region of long bones. Tumefaction is generally asymptomatic and discovered incidentally on radiography. Clinically, the neoplasm may be associated with pain [3,4].

Specifically, non ossifying fibroma may be asymptomatic, may be discerned incidentally or appear associated with a pathological fracture. Additionally, pre-existing non ossifying fibroma can impair the bone and predispose to pathologic fractures [3,4].

Instances with an intra-medullary component and loosely cohesive lesions exceeding 5 centimetre magnitude are especially termed as non-ossifying fibroma [3,4].

Histological elucidation

On gross examination, tumefaction appears as a well circumscribed, granular, reddish brown lesion.

On microscopy, a storiform pattern of tumour architecture composed of fibroblasts is observed. Benign multinucleated giant cells and foamy macrophages are disseminated within the tumour parenchyma along with cholesterol crystals and hemosiderin pigment deposits [4,5].

Mitotic figures are commonly discerned. Occasionally, tumour cells depict bizarre nuclei with degenerative features. Non ossifying fibroma morphologically recapitulates a benign fibrous histiocytoma [4,5].

Classically, proliferation of fibroblastic, spindle-shaped cells configuring a whorled or a storiform pattern is observed. Tumour cells are intermingled with fibroblastic connective tissue, numerous lipid laden macrophages, giant cells and foci of hemosiderin pigment deposition. Occasionally, an aneurysmal bone cyst like component is discerned [4,5].

Differential diagnosis

Non ossifying fibroma requires a segregation from neoplasms such as

- Giant cell tumour which is a painful neoplasm usually discerned in older individuals and exceptional within skeletally immature subjects. The lytic, epiphyseal tumefaction is devoid of mineralization or sclerosis [1]. Multinucleated, osteoclast-like giant cells incorporated with nuclei exceeding > 40 are uniformly disseminated within the tumour parenchyma. Spindle-shaped, elliptical or spherical cells are intermixed within an extensively vascularized stroma. Foci of fibrosis and reactive, woven bone are admixed with foci of acute haemorrhage, hemosiderin pigment deposits and accumulated xanthomatous histiocytes. Enlarged neoplasms display necrosis or secondary aneurysmal bone cyst like modifications [1,2].
- Osteosarcoma is a painful, malignant neoplasm demonstrating foci irregular bone destruction along with minimally distinct zones of transition and foci of periosteal reaction. Tumefaction depicts cellular permeation with replacement of medullary spaces, erosion of native bone trabeculae, cortical destruction and infiltration of abutting soft tissue. Neoplastic cells are pleomorphic, hyperchromatic and depict multiple morphologies as epithelioid, plasmacytoid, spindle-shaped, miniature spherical, clear cell and multinucleated giant tumour cell. Foci of neoplastic osteoid are deposited upon bony trabeculae. Non neoplastic giant cells may be scattered within the tumour parenchyma Also, mineralized soft tissue nodules are exemplified [1,2].
- Aneurysmal bone cyst is a multi-loculated, cystic lesion composed of blood filled cystic spaces subdivided by cellular septa incorporating fibroblasts, giant cells and woven bone. Calcified, basophilic material is disseminated within the solid foci. Mitotic figures are discerned although cytological atypia is absent and necrosis is uncommon. Plain radiographs depict a bubbly lesion [1,2].
- Unicameral bone cyst is also designated as solitary bone cyst and demonstrates a cyst wall constituted by attenuated fibrous tissue, fibroblasts and an absence of distinctive epithelial layer. Irregular bands of fibrin-like, calcified material are observed within the cyst wall which also contains osteoclast-like, multinucleated giant cells, foamy macrophages, hemosiderin pigment deposits and cholesterol clefts. Plain radiography displays a bubbly appearance [1,2].
- Pigmented villonodular synovitis enunciates diffuse, expansive sheets of tumour cells. Hyperplastic synovium with papillary projections are imbued with foamy histiocytes and hemosiderin containing macrophages. Tumefaction depicts a variable cellularity and an infiltrative tumour perimeter. Large clefts, pseudo-glandular spaces and alveolar spaces are layered with synovial cells, osteoclastic-like multinucleated giant cells and epithelioid cells. Abundant collagen, giant hemosiderotic granules and giant siderophages are enunciated within the lesion [1,2].

- Fibrous dysplasia is composed of irregular, branching and anastomosing trabeculae of woven bone devoid of osteoblastic rimming. Intervening fibrous tissue stroma is incorporated with bland, spindle-shaped cells. Mitotic figures are exceptional. Stromal alterations as myxoid change and adipose tissue metaplasia are infrequently discerned along with secondary aneurysmal bone cyst like modifications. Simple observation may suffice as pertinent therapy [1,2].

- Benign fibrous histiocytoma is a painful neoplasm usually occurring within long bones and pelvis. Tumefaction simulates a non-ossifying fibroma on morphology and demonstrates a storiform tumour pattern configured by spindle-shaped cells, frequent foam cells and variable quantities of intermingled, benign, multinucleated giant cells [1,2].

- Enchondroma demonstrates lobules of hyaline cartilage encased by bone and superimposed with perichondrium. Tumefaction is hyper-cellular with scattered binucleate tumour cells, foci of myxoid change, calcification and endochondral ossification. Lobules of bone may extend into underlying cartilage. Necrosis is commonly observed. Simple observation may be adequate therapy [1,2].

- Osteochondroma is comprised of bony nodules covered by a cap of mature hyaline cartilage with superimposed fibrous perichondrium. Young subjects depict foci of endochondral ossification appearing within mature bone. The mature hyaline cartilage cap gradually diminishes with increasing age and may be absent in adults. Elements of bone marrow may be incorporated within stalk of bone. The neoplasm may be adequately managed with simple observation [1,2].

- Eosinophilic granuloma delineates cellular proliferation of sheets of elliptical, mononuclear cells imbued with pale cytoplasm and characteristic uniform, reniform or cleaved nuclei. Mitotic figures can be abundant. Infiltration of inflammatory cells such as eosinophils is extensive although lymphocytes and plasma cells may be discerned. Multinucleated giant cells are frequently delineated along with foci of necrosis. Simple observation may be a satisfactory therapy in several instances [1,2].

- Paget's disease of bone is a condition demonstrating enhanced osteoclastic and osteoblastic activity. Lesions of acute phase are constituted primarily by woven bone, focal mosaic pattern of lamellar bone and numerous osteoclasts incorporated with up to 100 nuclei when occurring within the osteolytic phase. Chronic instances are composed of lesions with thick bony trabeculae, thickened bone and bone marrow traversed with fine fibrotic septa. The condition can be managed with simple observation [1,2].

Investigative assay

Upon plain radiograph, an eccentric, bubbly, lytic, metaphyseal lesion is enveloped by a rim of sclerotic bone. Typically, non-ossifying fibroma demonstrates a well-defined osteo-sclerotic perimeter circumscribing a lobulated, radiolucent lesion situated within subcortical or intra-cortical region. A periosteal reaction is characteristically absent [6]. Easley and Kneisl in 1997 documented that lesions incriminating an excess of > 50% of bone diameter may predispose to pathological fractures [7].

Bone cortex may be expanded and thinned out. The elongated lesion migrates to diaphysis with eventual bone growth and enlarges to up to one centimetre to 7 centimetres. Individuals with skeletal maturity usually demonstrate sclerotic lesions [6,8].

Computerized tomography (CT) is beneficially adopted for predicting possible fractures wherein quantitative CT may depict an enhanced possibility of pathological fractures [6,8].

Therapeutic options

Non ossifying fibroma can be managed conservatively with simple, non-operative techniques such as observation. Simple observation is adopted as an initial treatment methodology as majority of lesions undergo spontaneous resolution and the bone re-ossifies progressively within the second or third decade.

Curettage of the neoplasm and bone grafting is optimal for treating enlarged neoplasms with possible occurrence of pathological fractures. Plain radiography of the lesion is indicated at six month and twelve month interval followed by annual reassessment until complete re-ossification occurs [6,8].

Pathological fractures can be treated solely according to features of individual fracture as with plaster cast for fracture of long bones such as the distal femur. Non-displaced pathological fractures are appropriately treated by immobilization within the plaster cast. Majority of pathological fractures are treated conservatively and non-operatively [6,8].

Operative methodologies such as curettage and bone grafting are indicated in symptomatic and enlarged tumours incriminating in excess of > 50% to 75% of the bone cortex [6,8].

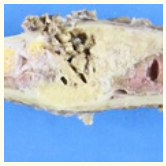

Figure 1: Non ossifying fibroma delineating a well circumscribed, granular lesion confined to the metaphysis of long bones [9].

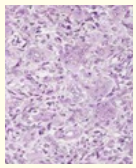

Figure 2: Non ossifying fibroma exhibiting whorls of spindle-shaped cells admixed with multinucleated giant cells and hemosiderin pigment deposits [9].

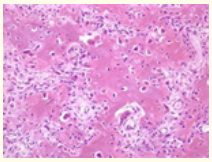

Figure 3: Non ossifying fibroma enunciating foci of reactive bone admixed with spindle-shaped tumour cells, focal whirling and foamy histiocytes [10].

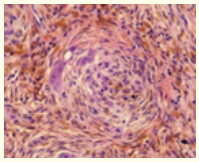

Figure 4: Non ossifying fibroma delineating a focal storiform pattern configured by spindle-shaped cells and multinucleated giant cells and hemosiderin pigment deposits [10].

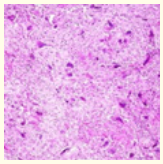

Figure 5: Non ossifying fibroma exhibiting a focal storiform pattern constituted by spindle-shaped cells, several multinucleated giant cells and hemosiderin pigment [11].

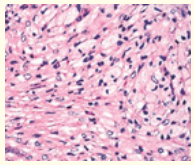

Figure 6: Non ossifying fibroma demonstrating spindle-shaped cells with focal whorls, hemosiderin pigment and lymphocytic infiltrate [12].

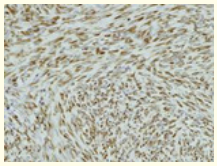

Figure 7: Non ossifying fibroma exhibiting spindle-shaped cells displaying a focal storiform pattern, significant hemosiderin pigment and a fibrotic stroma [13].

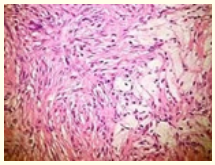

Figure 8: Non ossifying fibroma exemplifying spindle-shaped c ells articulating cellular whorls, hemosiderin pigment, few multinucleated giant cells and a fibrotic stroma [14].

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9. Image 1 and 2 Courtesy: Humpath.com

10. Image 3 and 4 Courtesy : Orthobullets.com

11. Image 5 Courtesy: Webpathology.com

12. Image 6 Courtesy: Pathology outlines.

13. Image 7 Courtesy: SAGE Journals.

14. Image 8 Courtesy: Libre Pathology.