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Editorial

The Refitted Ossein- Osteofibrous Dysplasia

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Osteofibrous dysplasia emerges as a benign fibro-osseous tumour frequently encountered within diaphysis of anterior tibia. Generally, nomenclature of ossifying fibroma of long bones or congenital fibrous defect of tibia is not recommended.

Morphologically, tumefaction comprises of trabeculae of woven bone with peripheral rimming by osteoblasts intermingled within bland stroma of fibroblastic tissue configuring a storiform pattern. Fibroblastic stroma depicts scattered epithelial cells which are immune reactive to keratin. Bone trabeculae display maturation of bone, especially towards tumour periphery, thereby configuring a 'zonal' architecture. Upon plain radiography, tumefaction appears as an intra-cortical, radio-lucent or multi-locular neoplasm with well defined, sclerotic perimeter. Tumefaction may reoccur. Also, tumour may retrogress following attainment of skeletal maturity.

Osteofibrous dysplasia configures around 0.2% of primary bone tumours. Commonly implicating the paediatric population, neoplasm occurs within first two decades. Median age of disease emergence is 10 years in males and 13.5 years in female. Besides, tumefaction may implicate neonates of 3 days or elderly subjects of 65 years. A specific gender predilection is absent [1,2].

Osteofibrous dysplasia is preponderantly (>95%) discerned within anterior tibial diaphysis. Besides, synchronous implication of fibula may occur. Few neoplasms are confined to metaphyseal region and configure as multifocal or bilateral neoplasms. Exceptionally, bones as the humerus, radius, ulna or clavicle are implicated [1,2].

Of obscure aetiology, concurrence of osteofibrous dysplasia and adamantinoma remains ambiguous. Osteofibrous dysplasia is posited to emerge as a consequence of displacement of neural crest derived epithelial cells or on account of traumatic implantation of epithelial cells into bone within the course of foetal development. Alternatively, metamorphosis of mesenchymal cells into epithelial cells may occur due to neoplastic regression [1,2]. Lesion appears devoid of specific concurrent genetic anomalies. However, germline mutations within MET gene may emerge within hereditary neoplasms. Additionally, mosaic lesions of RASopathy with specific cutaneous or skeletal neoplasms may demonstrate concurrence of osteofibrous dysplasia. Familial neoplasms are exceptionally encountered [2,3]. Generally, cytogenetic assay of the neoplasm appears superfluous. Few lesions expound trisomy of chromosomes 7, 8, 12, 21 or 22 which may be appropriately discerned with fluorescent in situ hybridization (FISH) or Giemsa banding. Trisomies appear to emerge within neoplastic, spindle shaped stromal cells [2,3]. Majority of neoplasms are incidentally discovered upon adoption of cogent imaging modalities. Generally, cogent causative factors or associations of disease emergence are absent. Neoplasm appears associated with pain, pathological fracture, bowing deformity or localized swelling. Length of tumefaction varies from 2 centimetres to 8.5 centimetres with mean length of 6.1 centimetres [2,3].

Grossly, a fibrous, whitish yellow tumefaction is confined to bony cortex. Tumour depicts a gritty consistency [3,4]. Upon microscopy, tumefaction delineates distinct components as bone trabeculae encompassed within fibrous tissue stroma. Characteristically, neoplasm delineates a zonation pattern. Centric zone of the lesion appears fibrous and admixed with attenuated, newly formed trabeculae of woven bone. Innumerable trabeculae of lamellar bone appear thickened, mature and commingle with peripherally displaced extraneous and innate bony cortices. Bone trabeculae appear circumscribed by epithelioid cells or active osteoblasts. Encompassing fibrotic stroma is composed of spindle shaped or stellate cells configuring abridges fascicles or an indistinct storiform pattern [3,4]. Cellular component appears intermingled within myxoid substance. Exceptionally, epithelial cells are disseminated within the stroma and appear immune reactive to cytokeratin. Additionally, stroma depicts scattered multinucleated giant cells, hemosiderin laden macrophages, foamy macrophages and hemosiderin pigment deposits. Ultrastructural examination of stromal cells demonstrates fibroblast-like cells imbued with prominent rough

endoplasmic reticulum. Focal epithelial differentiation appears absent. Thickened trabeculae of woven bone expound enhanced bone remodelling along with osteoblastic and osteoclastic activity [3,4].

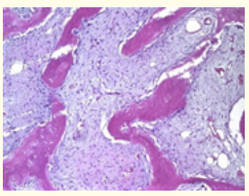

Figure 1: Osteofibrous dysplasia depicting a classic zonation pattern with centric fibrous tissue and peripheral trabeculae of woven bone surrounded by an abundant fibrous tissue stroma [7].

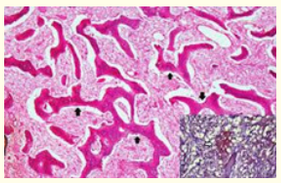

Figure 2: Osteofibrous dysplasia delineating a classic zonation pattern with centric fibrous tissue and peripheral trabeculae of woven bone encompassed within an abundant fibrous tissue stroma [8].

Tumour staging is applicable to neoplasms emerging within skeletal or facial bones, trunk or skull and is designated as [2,3]. Stage IA: Tumour is low grade and ≤8-centimetre magnitude(T1). Tumour grade appears as GX or G1. Regional lymph node or distant metastasis is absent (N0, M0). •stage IB: Tumour is low grade and > 8-centimetre magnitude (T2). Tumour grade appears as GX or G1. Primary bone site may depict >one distinct neoplasms. Regional lymph node or distant metastasis is absent (N0, M0). •stage IIA: Tumour is high grade and ≤8-centimetre magnitude(T1). Tumour grade appears as G2 or G3. Regional lymph node or distant metastasis is absent (N0, M0). •stage IIB: Tumour is high grade and > 8-centimetre magnitude (T2). Tumour grade appears as G2 or G3. Regional lymph node or distant metastasis is absent (N0, M0). •stage III: Primary bone site depicts multiple, high-grade tumours (T3). Tumour grade appears as G2 or G3. Regional lymph node or distant metastasis is absent (N0, M0). •stage IVA: Tumour magnitude and grade is variable (any T, any G). Regional lymph node metastasis is absent (N0). Distant metastasis into pulmonary parenchyma is discerned(M1a). •stage IVB: Tumour magnitude and grade is variable (any T, any G). Regional lymph node or distant metastasis occurs (N1,any M) OR tumour magnitude and grade is variable (any T, any G), regional lymph node metastasis may or may not occur (any N) and tumour metastasis into bone or distant organs besides pulmonary parenchyma (M1b) may ensue [2,3]. y: tumour subjected to preceding radiotherapy or chemotherapy r: recurrent tumour stage.

Epithelial cells disseminated within the stroma appear immune reactive to CK AE1/AE3, CK1, CK5, CK14, CK17, CK19, epithelial membrane antigen (EMA) and p63. Fibroblasts and osteoblasts appear immune reactive to transforming growth factor β1(TGFβ1) and transforming growth factor β2(TGFβ2).

Stromal component appears immune reactive to type IV collagen, laminin, galectin 3, osteonectin or fibronectin [5,6]. Osteofibrous dysplasia requires segregation from neoplasms as classic adamantinoma, osteofibrous dysplasia-like adamantinoma, fibrous dysplasia, osteoid osteoma, osteoblastoma, unicameral bone cyst, osteomyelitis, non-ossifying fibroma, aneurysmal bone cyst, chondromyxoid fibroma, langerhans cell histiocytosis or eosinophilic granuloma, osteosarcoma, chondrosarcoma, hemangioendothelioma, angiosarcoma or metastatic carcinoma [5,6]. Osteofibrous dysplasia may be subjected to plain radiography, computerized tomography (CT) or magnetic resonance imaging (MRI) which are optimally adopted to evaluate localization or extent of tumefaction, emergence of pathological fracture and appropriate surgical staging [5,6]. Upon plain radiographs, tumefaction characteristically expounds as a intra-cortical, eccentric, multi-locular, radiolucent or osteolytic lesion traversed by intrinsic septa and a sharply defined, sclerotic perimeter. Few lesions may exemplify ground glass appearance, significant sclerosis and an inadequately defined perimeter. Enlarged lesions may induce cortical expansion, bowing deformity or pathological fracture. Computerized tomography (CT) may be optimally adopted to assess features as matrix mineralization, pathological fracture, absence of periosteal reaction and a distinct transitional zone. However, diagnostic outcomes appear compatible with magnetic resonance imaging [5,6]. Magnetic resonance imaging (MRI) appears beneficial in assessing tumour emergence within the medulla or surgical tumour staging. However, appropriate tumour segregation may be challenging. Medullary cavity is infrequently implicated. However, an estimated 40% lesions appear to involve bony medulla. Medullary involvement may be partial or complete. Upon T1 weighted magnetic resonance imaging (MRI), images of intermediate signal intensity may be discerned. Upon T2 weighted imaging, images of intermediate to hyper-intense signal intensity are expounded. Contrast enhanced images depict homogenous or heterogeneous patterns [5,6]. Incisional biopsy or Tru-Cut tissue sampling may be employed to ascertain the histological diagnosis in order to undertake intervention procedures as curettage or surgical excision of the tumour. Tumour ascertainment upon miniature tissue samples may be challenging and osteofibrous dysplasia requires segregation from neoplasms as osteofirous dysplasia-like adamantinoma or classic adamantinoma [5,6]. Surgical extermination of the neoplasm along with or in the absence of bone grafting may be optimally adopted to alleviate the neoplasm. Alternatively, enlarged tumours or lesions with bowing of tibia may be managed with bone curettage. Few neoplasms may be subjected to meticulous observation as lesions may spontaneously retrogress

following puberty. As the neoplasm expounds minimal proportionate tumour reoccurrence, extra-periosteal surgical excision may be optimally employed and is recommended [5,6]. Tumefaction enunciates favourable prognostic outcomes. However, neoplasm exemplifies a tendency towards tumour reoccurrence. Besides, spontaneous tumour retrogression may ensue upon puberty. Following curettage or surgical resection, proportionate tumour reoccurrence emerges at ~25% [5,6].

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7. Image 1 Courtesy: Orthobullets.com.

8. Image 2 Courtesy: Thieme connect.