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Commentary

Mime and Mummer-Osteofibrous dysplasia-like Adamantinoma

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Received: July 14, 2025; **Published:** July 26, 2025

Osteofibrous dysplasia-like adamantinoma emerges as an exceptionally encountered, malignant, primary bone tumour of uncertain genesis. Tumefaction is preponderantly confined to tibia or fibula. The alternative terminology of well differentiated adamantinoma is not recommended.

Neoplasm depicts predominant clinical and pathological variants denominated as

- Osteofibrous dysplasia (OFD)-like differentiated adamantinoma comprised of clusters of inconspicuous epithelial cells enmeshed within a fibro-osseous stroma.
- Classic adamantinoma displays distinct epithelial elements entangled within a fibro-osseous stroma.
- Dedifferentiated adamantinoma denominates absence of epithelial differentiation and foci of sarcomatoid alterations.

Neoplasm expounds characteristic biphasic fibro-osseous morphology of epithelial structures embedded within a mesenchymal or osteofibrous dysplasia-like stroma [1,2].

The exceptionally encountered neoplasm configures < 1% of primary bone tumours. Tumefaction may be observed within 4 years to 75 years with median age of disease occurrence at 30.8 years. A male to female proportion of 5:4 is enunciated [1,2].

Majority (~90%) of lesions appear localized to median one/third of tibial diaphysis. Additionally, neoplasm is encountered within ulna, femur, humerus, radius, ribs, tarsal bones, metatarsal bones and extra-skeletal pretibial soft tissue. Synchronous tumour emergence within the tibia and fibula occurs in ~10% instances. Multifocal tumour occurrence within the tibia may frequently occur [1,2].

Epithelial component of adamantinoma is posited to derive directly from the mesenchymal tissue with gradual quantifiable tumour expansion. Tumour progression into an aggressive subtype may concur with transition of epithelial cells into mesenchymal cells with sarcomatoid dedifferentiation, a feature associated with epithelial cell immunoreactivity [2,3].

The neoplasm depicts repetitive genomic mutations within KMT2D (MLL2) genes. Few tumours may depict somatic genetic fusions within EPHB4-MARCH10 gene [2,3].

Elevated expression of DLK1 gene as encountered within adamantinoma may be applicable as a possible molecular biomarker. A reoccurring pattern of numerical anomalies with extra copies of chromosomes 7, 8, 12, 19 and 21 may be observed within osteofibrous

dysplasia, osteofibrous dysplasia-like and classic adamantinoma, thereby indicating a common histogenesis of aforesaid lesions with perpetuating neoplastic progression [2,3].

Osteofibrous dysplasia emerges as a precursor of adamantinoma.

Pre-emptive clinical symptoms of the indolent neoplasm are nonspecific and contingent to extent and location of disease.

Onset is insidious and the disease demonstrates a protracted, progressive clinical course [2,3].

Cytological examination depicts a biphasic cell population comprised of epithelioid cells and significant percentage of singly disseminated or fragments of spindle shaped cells [3,4].

Epithelioid cells appear impregnated with indistinct cytoplasm, bland spherical to elliptical nuclei, finely dispersed nuclear chromatin, distinctive nuclear grooves and occasional micro-nucleoli. A percentage of cells depict spindle shaped cell morphology, copious clear cytoplasm and elongated nuclei [3,4].

Grossly, tumefaction appears fleshy, firm and yellowish/grey or greyish white. Occasionally, tumour parenchyma depicts macroscopic cysts impregnated with straw coloured or haemorrhagic fluid [3,4].

Upon microscopy, the characteristically biphasic neoplasm delineates commingling of varying proportions of epithelial and osteofibrous components. Diverse tumour configurations may be encountered.

The fibrous component appears loose myxoid, hyalinized or sclerotic. Mitotic figures appear infrequent at 0 - 2 mitoses per 10 high power fields [3,4].

Neoplasm depicts distinctive morphological variants as

- Classic adamantinoma delineating a prominent epithelial component expounded by minimally atypical epithelial cells entangled within an osteofibrous dysplasia-like stroma configured of conspicuous, solid or basaloid cellular nests demonstrating peripheral palisading. Alternatively, tubular structures, keratinized squamous cell nests or bundles of spindle shaped cells may be encountered [3,4].
- Osteofibrous dysplasia-like adamantinoma typically expounds miniature clusters of scattered epithelial cells enmeshed within a predominantly osteofibrous dysplasia-like stroma [3,4].
- Dedifferentiated adamantinoma exemplifies sarcomatoid features. Aggregates of significantly pleomorphic cells with frequent mitosis and focal deposits of osteoid, chondroid or clear cell change may be observed. Sarcomatous zones appear immune non reactive to keratin. Ultrastructural examination demonstrates epithelioid tumour cells impregnated with tonofilaments, hemidesmosomes and desmosomes [3,4].

Osteofibrous dysplasia-like adamantinoma appears immune reactive to cytokeratin AE1/AE3, basal epithelial cell keratins as CK5, CK14 and CK19, vimentin, epithelial membrane antigen (EMA), p63 and podoplanin (D2-40) [5,6].

Grade I
Low grade central osteosarcoma
Clear cell chondrosarcoma
Grade I chondrosarcoma
Parosteal osteosarcoma
Adamantinoma
Grade II
Periosteal osteosarcoma
Grade II chondrosarcoma
Grade III
Conventional osteosarcoma
Telangiectatic osteosarcoma
Small cell osteosarcoma
Secondary osteosarcoma
High grade surface osteosarcoma
Malignant giant cell tumour
Ewing sarcoma
Grade III chondrosarcoma
Mesenchymal chondrosarcoma
Dedifferentiated chondrosarcoma

Table: *Grading of bone tumours [4,5].*

Tumour cells appear immune non reactive to CK8, CK18 and CD34.

Osteofibrous dysplasia-like adamantinoma requires segregation from neoplasms as osteofibrous dysplasia, distant metastasis from various primary carcinomas or adamantinoma-like Ewing sarcoma [5,6].

Cogent imaging may be adopted for discerning the neoplasm. Upon imaging, singular or multiple lytic lesions of variable magnitude demonstrating sclerotic perimeter with the configuration of classic 'soap bubble' appearance may be discerned within the diaphyseal region or metaphyseal cortex [5,6].

Computerized tomography of primary tumour site elucidates involvement of bony cortex or precise enunciation of distant metastases [5,6].

Akin to osteofibrous dysplasia, osteofibrous dysplasia-like adamantinoma is pre-eminently an intra-cortical lesion.

Magnetic resonance imaging appears suitable in detecting multi-focal bony lesions or lesions demonstrating invasion of medullary cavity or extra-osseous soft tissue [5,6].

Notwithstanding, a definitive diagnosis may be ascertained with precise histopathological examination wherein additional tissue samples may be obtained with 'open' tissue sampling techniques. Classically, surgical extermination with removal of broad perimeter

of uninvolved tissue is recommended for eradication of an adamantinoma. Procedures as intralesional or marginal surgical excision are associated with enhanced possible localized tumour reoccurrence [5,6].

Meticulous monitoring of extended duration is necessitated on account of possible emergence of delayed complications.

The classic variant appears indolent and is associated with an unpredictable clinical course. Enhanced proportionate localized tumour reoccurrence may ensue with inadequate surgical excision. Besides, possible occurrence of tumour reappearance appears concurrent with an elevated neoplastic epithelial to stroma ratio [5,6].

Comprehensive and radical surgical excision is associated with superior prognostic outcomes [5,6].

Osteofibrous dysplasia-like adamantinoma demonstrates a favourable prognostic outcome, in contrast to classic adamantinoma. Dedifferentiated adamantinoma expounds an aggressive clinical course [5,6].

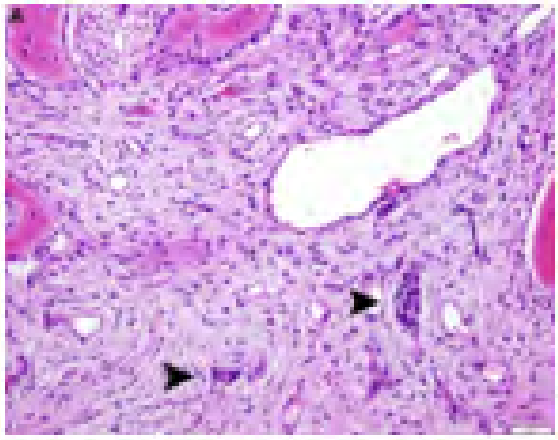

Figure 1: *Osteofibrous dysplasia-like adamantinoma delineating a biphasic countenance composed of epithelial and osteofibrous components intermingled within a loose hyaline and myxoid stroma [7].*

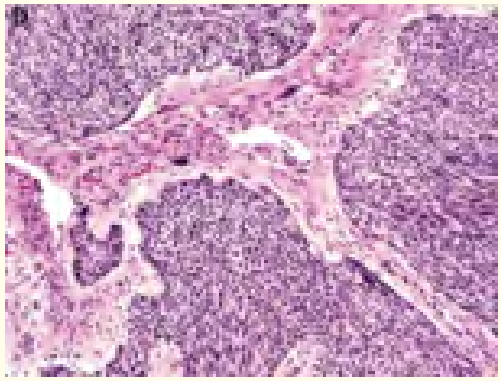

Figure 2: *Osteofibrous dysplasia-like adamantinoma expounding a biphasic composition comprised of epithelial cells and osteofibrous tissue enmeshed within a loose, hyaline and myxoid matrix [7].*

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7. Image 1 and 2 Courtesy: Surgical pathology clinics.

Volume 24 Issue 8 August 2025