Between Bone and Epithelium: A rare case of Adamantinoma

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ABSTRACT

Adamantinoma is a rare, low-grade malignant bone tumor that most commonly affects the tibial diaphysis. Due to its nonspecific clinical and radiologic presentation, diagnosis can be challenging. Immunohistochemistry plays an important role in confirming the diagnosis.

We report a case of a 38-year-old woman who presented with a one-year history of gradually progressive swelling and pain in the right leg. Imaging revealed a suspicious malignant lesion in the mid-tibia. Initial biopsy suggested a malignant mesenchymal neoplasm with a differential diagnosis of synovial sarcoma. Immunohistochemical analysis demonstrated positivity for CK, p63, p40, CK5/6, vimentin and D2-40, confirming the diagnosis of spindle cell variant of adamantinoma.

Keywords: Adamantinoma;Bone Tumor;Malignant;Tibia

INRODUCTION

Adamantinoma is an uncommon bone tumor, comprising only 0.1% to 0.5% of all primary bone tumors.[1] It commonly affects the long bones, particularly the tibia, and presents with wide range of radiologic and histologic features, often making difficulties in accurate diagnosis.[2] The underlying cause of adamantinoma remains uncertain, but recent genetic and molecular research have led to better characterizations of this bone tumor. These comprehensive studies have uncovered genetic changes that could serve as targets for future treatments.[3]

CASE REPORT

A 38-year-old housewife presented with a one-year history of gradually increasing swelling and pain in the right leg. She reported a minor trauma on same spot from a fall about six months prior. Initially the pain was mild that intensified over time and eventually cause difficulty with walking. X-ray revealed a well defined lesion in the tibial disphysis.(Figure 1) MRI was done and initially before biopsy which demonstrated a well-defined lesion within the medullary cavity of the mid-third of the tibial diaphysis. The lesion showed mild cortical thinning and extension toward the subcutaneous fat plane through a cloaca. Peripheral contrast enhancement was noted, with surrounding bone marrow edema. No significant periosteal reaction was observed. Rest portion of tibia appears normal. Fibula appeared normal.

Histopathological finding after incisional biposy shows biphasic tumor consisting of epithelial and fibrous tissue with hypocellular and hypercellular pattern. (Figure 2) Epithelial cells predominate appears basiloid and arranged in sheets, nests, islands with admixed loose fibromyxoid tissue. Individual tumor cells reveal spindled elongated nuclei

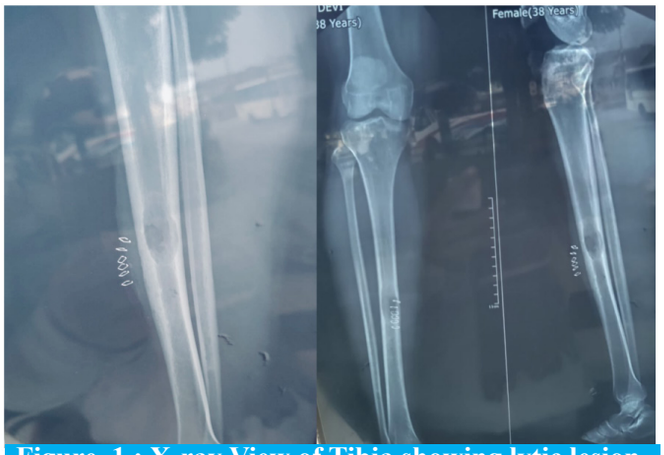

Figure 1 : X-ray View of Tibia showing lytic lesion in tibial disphysis

with eosinophilic cytoplasm. At places peripheral pallisiding is noted. Some scattered lymphocytes are also seen along with extensive areas of hemorrhages, some stag horn vessels and fragments of dead bone. The histologic features are suggestive of malignant mesenchymal neoplasm; Synovial sarcoma needed to be confirmed with application of immunohistochemistry.

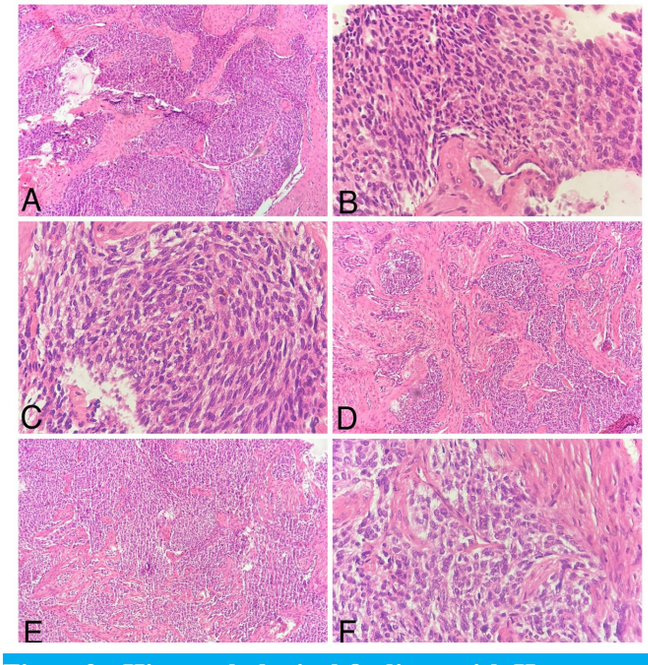

Figure2 : Histopathological findings with Hematoxylin and Eosin (H&E)

Because of the many predominant spindle shaped epithelial cells with little intervening stroma with occasional palisaded pattern synovial sarcoma was suspected. Synovial sarcoma is also the important differential diagnosis of adamantinoma because of its morphological similarity. But the treatment needs to be aggressive in synovial sarcoma and for confirmation IHC was advised.

In IHC report interpretation, The tumor cells showed positivity for CK, p63, p40, CK5/6, Vimentin & D2-40.Where as tumor cell were negetive for CK7, CK20, S100, CD34, Desmin, TLE1, PAX-8, CDX2, SOX10, GATA-3, and BerEP4. And KI-67 was 30-40%. These findings confirmed the diagnosis of spindle cell variant of adamantinoma. The patient underwent wide local excision with intercalary resection of the tibia followed by reconstruction using an endoprosthesis and soleus flap with skin grafting. Histopathological examination of the excised specimen confirmed the diagnosis of adamantinoma of tibia.

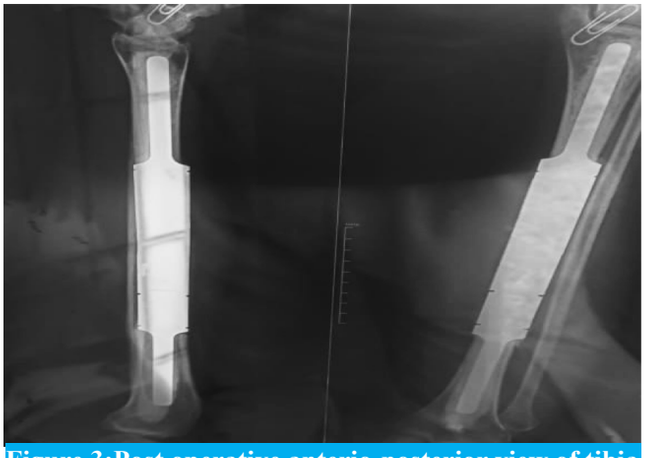

Figure 3:Post operative anterio-posterior view of tibia

DISCUSSION

The disease is predominantly seen in young adults, with a slight male predominance. Adamantinoma primarily involves the anterior metaphysis or diaphysis of the tibia in 85–90% of cases, often with multifocal tibial lesions. About 10% also have involvement of the ipsilateral fibula. Although rare, other bones like the radius, ulna, femur, and spine may be affected, but such diagnoses require careful consideration.[4] Adamantinoma typically presents with subtle and nonspecific symptoms, which vary according to the lesion's location and extent. The disease has a slow, progressive course, and patients often delay seeking medical evaluation for several years. In a review of 200 cases by Moon and Mori, a significant history of trauma was reported in approximately 60% of patients.[5] The majority of patients present with swelling, which may or may not be accompanied by pain. Involvement of the anterior surface of the tibia can lead to a bowing deformity.[6] Pathological fractures occur in up to 23% of cases. Additionally, there have been few reported cases of paraneoplastic hypercalcemia associated with tibial adamantinoma and pulmonary metastases.[7,8] Multiple imaging techniques are important for diagnosing adamantinoma. X-rays the tumor

is typically well circumscribed, cortical, often multilobulated and osteolytic - a soap bubble-like pattern, while CT scans help evaluate soft tissue involvement and cortical damage. The lesion commonly remains intracortical and extends longitudinally along the bone, but it may also destroy the cortex and invade the medullary cavity or adjacent soft tissue. Additionally, MRI is essential for determining the extent of the disease and assisting in surgical planning.[9] Although advancements in medical imaging have improved diagnostic capabilities, identifying adamantinoma remains challenging due to its often nonspecific radiological features. The radiographic appearance can range from a completely cystic lesion to a fully sclerotic one, making diagnosis more difficult. Consequently, histopathology plays a crucial role, with the presence of epithelial nests within a fibrous stroma being a key diagnostic indicator.[10] Histologically there are three subtypes, Classic adamantinoma; osteofibrous dysplasia like adamantinoma and dedifferentiated adamantinoma.[4]

Classic adamantinoma is defined by its biphasic composition, featuring both epithelial and osteofibrous elements that may be present in different proportions and display variable differentiation patterns.[11] In classic adamantinoma the epithelial component is predominant. Classic adamantinoma is classified into four patterns of growth namely basaloid, tubular, spindle cell and squamous. [12] The basaloid pattern is composed of solid clusters of basaloid cells. The tubular pattern is characterized by a row of cuboidal epithelial cells with central separation. The spindle cell pattern shows uniform spindle-shaped cells; although it stains positive for cytokeratin suggesting a possible sarcomatoid carcinoma. The differential diagnosis can be made, as adamantinoma lacks the significant cytological atypia typical of sarcomatoid carcinoma. The squamous pattern can resemble squamous cell carcinoma, but again, the presence of atypia in carcinoma helps distinguish the two.[13] Compared to classic adamantinoma, which has a dominant epithelial component, osteofibrous-like adamantinoma is composed mainly of osteofibrous tissue. The diagnostic criteria for osteofibrous like adamantinoma is the predominance of the osteofibrous tissue containing small clusters of epithelial cells and cytokeratin-positive staining. Mitotic figures are generally low.[14] By far the rarest subtype is dedifferentiated adamantinoma. In these tumors, areas of classic adamantinoma gradually merge with a diffuse growing proliferation in which the characteristic epithelial differentiation is lost and instead pleomorphic cells are present, with a high mitotic count.[4] Classic adamantinoma usually occurs in patients older than 20 years, whereas differentiated adamantinoma , Adamantinomas are immunohistochemically positive for keratins 14 and 19.[15] The epithelial component exhibits co-expression of cytokeratins 5, 14 and 19, epithelial membrane antigen and vimentin [16]. CK1, CK13, and CK17 are also variably present. CK8 and CK18 are virtually absent.[4] The stromal component displays immunohistochemical positivity for vimentin. Adamantinomas lack the immunohistochemical expression of keratins 8 and 18, which differentiates them from many other skeletal or soft-tissue tumors.

CONCLUSION

Adamantinoma should be considered in the differential diagnosis of tibial lesions with biphasic histology. Immunohistochemistry is crucial in establishing the diagnosis and differentiating it from other spindle cell neoplasms. Early diagnosis and wide surgical excision are key to successful management.

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