

ULNAR ADAMANTINOMA - A RARE TUMOUR AT A RARE SITE

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ABSTRACT

Background: Adamantinoma is a rare slow growing tumour with malignant potential. Males of 20-50 years are usually affected. This neoplasm has a predilection for tibia and upper extremity involvement is very rare. **Case Presentation:** A 24 year old male patient presented with a swelling in the left forearm. X-ray showed a lytic lesion in the mid-shaft of ulna. Imprint cytology revealed a spindle cell lesion. Curettage and biopsy was done. Microscopy revealed a spindle cell neoplasm arranged in storiform pattern showing positivity for CK and vimentin with immunohistochemistry. **Conclusion:** Our histopathological and immunohistochemical features were suggestive of Adamantinoma. The case is unusual because of the rarity of the tumour and even rare occurrence of the neoplasm in upper limb. Hence adamantinoma should be considered in differentials of lytic lesion of the upper limb even though the predilection for these sites are rare.

KEYWORDS: Adamantinoma, Ulna, Lytic lesion.

MESH TERMS: Adamantinoma, Ulna, Lytic lesion.

BACKGROUND

Adamantinoma was first described by Fisher in 1913. It is a rare slow growing tumour of long bones. Incidence of this tumour accounts for only <1% of all malignant bone tumours. Males in second to third decade of life are mainly affected. Common site of involvement is tibial diaphysis. Upper extremity involvement is rare accounting for less than 10 case reports in the world literature. Here we present a case of adamantinoma in the left ulna.

CASE PRESENTATION

24 year old male presented with swelling in left forearm of 7 months duration which was gradually increasing in size. No history of pain, discharge or fever was present. Physical examination revealed a swelling of 4x2x1cm in mid-shaft of left ulna. Skin over the swelling was normal. No visible pulsations seen. Plane of swelling was bone. No tenderness or local rise of temperature noted. Radiological evaluation revealed a lytic lesion in the mid-shaft of left ulna. Cortical break on volar aspect also noted [Figure 1]. The differentials considered were non ossifying fibroma and langerhans cell histiocytosis(LCH). Curettage and open biopsy was done. Intraoperatively a well circumscribed lesion with thick capsule seen. Imprint cytology revealed a spindle cell lesion [Figure 2]. Microscopy showed an infiltrating neoplasm composed of spindle shaped cells arranged in

storiform pattern and sheets. The cells had moderate to scanty cytoplasm, elongated vesicular nuclei, fine chromatin and prominent nucleoli [Figure 3]. Clusters of foam cells were seen. Thick hyalinised vessels were also seen. The stroma was myxoid in nature. These features favoured a diagnosis of Adamantinoma. Immunohistochemistry (IHC) study was done to rule out osteofibrous dysplasia. Tumour cells were positive for CK and vimentin confirming the diagnosis of adamantinoma [Figure 4]. Patient is on follow-up since recurrence is common.

Figures with legends



Figure 1: Lytic lesion with cortical break in the mid-shaft of left ulna.

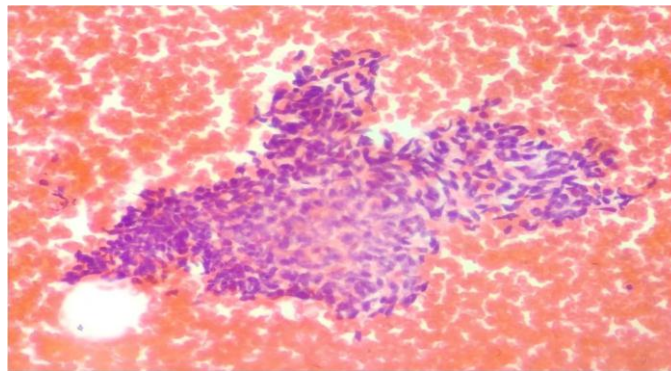


Figure 2: Cohesive clusters of oval to spindle cells with moderate cytoplasm and oval elongated nuclei with finely clumped chromatin.(Pap stain-40x)

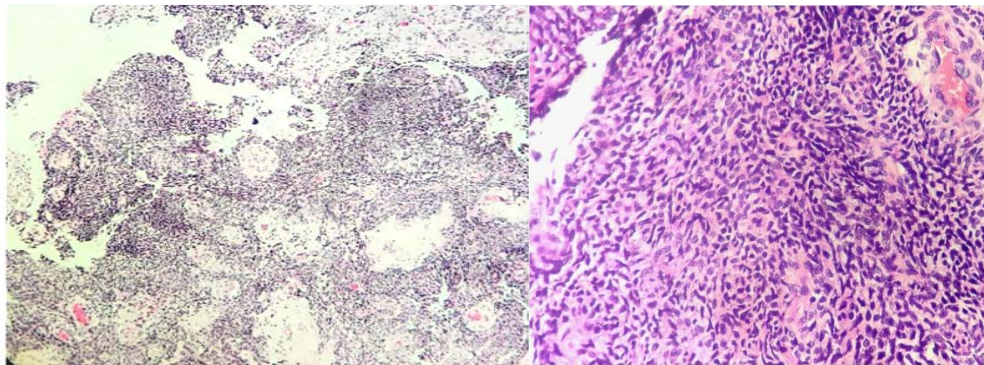


Figure 3. a: Fibro-osseous tissue with an infiltrating neoplasm arranged in storiform pattern and sheets. Thick hyalinised vessels and myxoid stroma present.(H&E 10 x)

3. b: Individual cells have moderate to scanty cytoplasm elongated vesicular nuclei fine chromatin, and prominent nucleoli.(H&E 40 x)

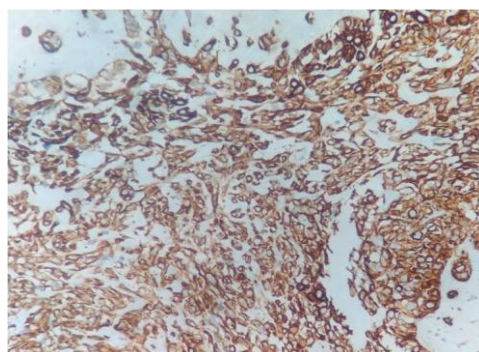


Figure 4- Pan CK- cytoplasmic positivity in neoplastic cells.

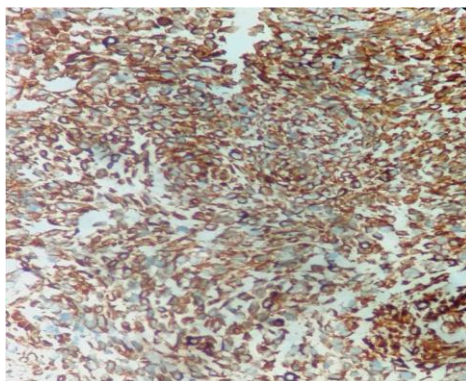


Figure 5- Vimentin - cytoplasmic positivity in neoplastic cells.

DISCUSSION AND CONCLUSION

Adamantinoma is a rare malignant tumour with an incidence of <1%. It is a slow growing tumour with malignant potential. Ultrastructural and IHC studies suggest that adamantinoma is a tumour of epithelial origin.^[1] The tumour has a predilection for males. The age group 20-50 years is more frequently affected. Most common affected bone is tibial diaphysis or anterior metaphysis. Other uncommon sites of involvement include humerus, ulna, femur, fibula, radius, innominate bone, rib, spine and rarely small bones of the hand and foot.^[2] The usual clinical presentation is local swelling or deformity with or without pain. The skin over the lesion may be shiny or tense. Pathological fractures are seen in 23 % of cases at time of presentation.^[3] Xrays are diagnostic showing expansile osteolytic lesions. Soap bubble appearance can be seen. Cortical destruction with extension into the medullary canal and/or the extraosseous soft tissues may be seen in computed tomography (CT) or Magnetic Resonance Imaging (MRI). Grossly tumour is well demarcated with firm to bony consistency and grey white to yellowish appearance. There can be lobulation with peripheral sclerosis. Histopathologically it is composed of bland epithelial cells with peripheral palisading in fibrous stroma. Foam cells and myxoid changes may also be present. IHC shows that tumor cells contain both epithelial and neural antigens. Surgery is the mainstay of treatment. These tumours are resistant to chemotherapy and radiotherapy. 15 to 20% of the cases shows metastatic potential.^[4] Usual sites for metastasis are lungs, lymph nodes, other long bones and rarely abdominal viscera. Male sex, pain, symptoms of < 5 years duration and initial treatment by biopsy, curettage or resection are risk factors for recurrence or metastatic disease. Mortality rates range from 13%–18%.^[5] Local recurrence can occur after 5–15 years of diagnosis, hence follow up is essential indefinitely.^[6]

Though adamantinoma is a tumour with predilection for tibia, it should be considered in the differentials of lytic lesion in upper extremities also.

List of Abbreviations

Computed tomography (CT)
Immunohistochemistry (IHC)

Langerhans cell histiocytosis (LCH)
Magnetic Resonance Imaging (MRI)

Declarations

Ethics approval and consent to participate - This case report was approved by the Ethics committee of Amala institute of medical sciences.

Consent for publication: Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

Patient Declaration Statement

“The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.”

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study

Competing interests - The authors declare that they have no competing interests.

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Authors' contributions

Dr Dominic K Puthoor did clinical examination and curettage and biopsy. Dr Maymol P Varghese, Dr Lekha K Nair and Dr Joy Augustine performed the histological examination. All authors read and approved the final manuscript.

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