Proximal Humerus Pecoma: First Ever Case Report.

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Introduction

PEComas (tumours showing perivascular epitheloid cell differentiation) are a family of related mesenchymal neoplasms that include angiomyolipoma, lymphangiomyomatosis, clear cell "sugar" tumour of the lung, and a group of rare, morphologically and immunophenotypically similar lesions arising at a variety of visceral and soft tissue sites. PEComa manifesting as a primary bone lesion is extremely rare. This case report describes a rare case of proximal humerus PEComa.

Case report

A 16 year old boy presented with swelling over the left shoulder following a fall. His MRI of left arm mass showed pathological fracture of left neck of proximal humerus and large soft tissue component seen. Tissue biopsy of the shoulder mass noted to be Ewings Sarcoma and he underwent neoadjuvant chemotherapy. He then subsequently underwent wide tumour resection and endoprosthesis recostruction following discouraging response to the chemotherapy. His intraop histopathology report shows an aggressive epitheloid tumour with immunostaining pattern consistent with malignant melanoma. After discussion with prominent pathologist in paediatric sarcoma, diagnosis of PEComa was derived.

Discussion

The first case of primary bone PEComa was described by Insabato et al. in 2002 which occurred in the tibia of a 30-year-old male. The presentation of PEComa as a primary bone lesion is extremely rare. In addition to imaging, histological and immunohistochemical studies are crucial for the diagnosis of PEComa. These can be carried out following biopsy, intralesional curettage or wide resection of the tumor. Cytologic findings are significant for epithelioid cells with clear and eosinophilic cytoplasm, intermixed with spindle cells. In this case, the initial tissue diagnosis was Ewings sarcoma and leads to a delay in the exact treatment of the patient.

Conclusion

Although very rare, PEComa can present as a primary bone lesion. Clinical and pathological correlation is mandatory in arriving at the correct diagnosis.