

Myositis ossificans with hoffa's disease (Lipoma Arborescens): Usual mimicker of soft tissue sarcoma

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Abstract

Myositis Ossificans is a benign heterotopic ossification mainly occurs in muscles and soft tissue. Hoffa's disease (Lipoma Arborescens) is a rare benign lesion characterized by villous lipomatous proliferation of synovial membrane. Due to its unusual property of abnormal ossification in muscles and soft tissue, myositis ossificans presents a diagnostic and therapeutic problem to pathologist and clinician respectively, as it mimics soft tissue sarcoma.

We present a case of 14 year male presented with left thigh swelling near knee joint with restricted movements. Clinically it was suspected as synovial sarcoma. Wide local excision of mass and synovial membrane excision was done. Final diagnosis on histopathology was given as Myositis Ossificans with Hoffa's disease.

The aim of this case is to highlight the unusual combination as myositis ossificans with lipoma arborescens together due to its rarity and sarcoma mimics. Such type of combination was reported for the first time in present literature in Indian context.

Keywords: Myositis ossificans, Hoffa's disease, Lipoma arborescens, Synovial sarcoma, Tumor, Mimics.

Introduction

Myositis ossificans (MO) is a rare benign disorder with heterotopic ossification of the muscle and soft tissue.¹ Due to its abnormal bone formation at different locations it pose a diagnostic as well as therapeutic challenge to the concerned pathologist and clinician respectively. Myositis ossificans occur most commonly in adolescents and adults, children less than 10 years has very rare incidence.^{2,3} The exact pathophysiology is not clear. However the necrosis of the tissue with fibroblastic, vascular proliferation and ulceration in ground substance of connective tissue leads to heterotopic benign bone formation.^{3,4}

Myositis ossificans classified into three types. Myositis ossificans progressiva (severe generalized and hereditary), MO without history of trauma (with burns, hemophilia and neurological), MO circumscripta/ traumatic (related to direct blow or direct trauma).¹ The third type MO traumatica represents 60-75% of all the cases and the most common type.⁵ Most commonly affects the adolescent and young adults with the most common site is extremities, mainly anterior thigh. Common presentation of MO is painful soft tissue mass with no sign of inflammation. The diagnosis often requires radiological and histopathological details.⁵

Hoffa's disease/Lipoma arborescens (LA) is a rare slow growing, chronic, benign intraarticular lesion characterized by villous lipomatous proliferation of synovial membrane with unclear etiology.^{6,7} It was first described by Hoffa in 1904 and in detail by Arzimanoglu in 1957.⁸ Due to presence of mature fat cells in synovium it misleads as lipoma and arborescens (tree in latin) means treelike lipomatous villous synovial proliferation.⁹ LA most commonly occur in sites like knee followed by hip, wrist, elbow and shoulder.⁹ Less than 100 cases of lipoma arborescens has been reported in the literature. Main

age group of LA is 3rd to 5th decade and only handful of cases are reported in children.

We present a case of myositis ossificans with Hoffa's disease in 14 year old male due to its clinical suspicions of synovial sarcoma and unusual combination (not found in Indian literature to till date) and rare gross and micro findings. We enlighten the role of histopathology in such challenging cases.

Case History

A 14 year old male patient presented to the surgical OPD with chief complaints of swelling of left thigh near knee joint with restricted movements. Swelling is associated with progressive pain since last month. Personal and family history is not contributory. Past history of fall from bicycle and blunt trauma to left thigh was found 1.6 years back. On physical examination showed restricted movements with single, large oval mass of about 12x10x6 cms in the anterior aspect of left thigh near knee joint. The mass was hard to firm in consistency. All the hematological, biochemical and serological investigations were within normal limits. USG of left knee joint mass showed lobulated, solid, hypoechoic lesion with irregular central calcification in suprapatellar bursa with surrounding fatty proliferation. Moderate vascularity was noted on color Doppler. MRI scan of knee joint lobulated mass of 7x6x4 cms with heterogeneous, hyperintense on T1 and T2 images with fatty intensity with joint effusion. Clinically suspected as soft tissue sarcoma and wide local excision was performed. We received excised soft tissue mass measuring 10x9x5 cms.

Gross Examination

Received excised soft tissue mass measuring 10x9x5 cms. E/S- rounded, circumscribed and multinodular with encapsulated. On C/S- yellowish white multiple nodules are

noted of varying sizes, larger measures 3x3 cms. In the center of the excised mass a large rounded grey chalky white calcified mass with glistening white surface noted measuring 4x3 cms (Fig. 1) and on c/s gives gritty sound. Surrounding grey yellowish soft tissue mass shows congestion and focal areas of hemorrhages. Also received excised synovial membrane measuring 7x1cms and 3x1 cms. E/S and on cutting seen yellowish fibro adipose tissue with congested blood vessels.

Light Microscopy

Multiple sections studied show well circumscribed lobules of fibroadipose tissue with characteristic 'ZONAL' pattern of fibroadipose with fibroblastic tissue with richly vascular granulation tissue like areas. Few of the fibroblastic cells show mild atypia. Peripheral zonal areas showed large areas of immature cartilage formation with adjacent ill defined trabeculae of lamellar bone or osteoid formation thinly rimmed by osteoblasts and occasional osteocytes. (Fig. 2,3) Characteristic zonal pattern of cartilage interspread with bone with fibroblastic proliferation was evident (Fig. 4). Sections through synovial membrane showed multiple and elongated 'Villous proliferation' of synovium. The villous proliferation of the synovium contain abundant amount of fat with many congested blood vessels and sparse mononuclear cells infiltration (Fig. 5). Final histopathological diagnosis given as Myositis ossificans with Hoffa's disease.

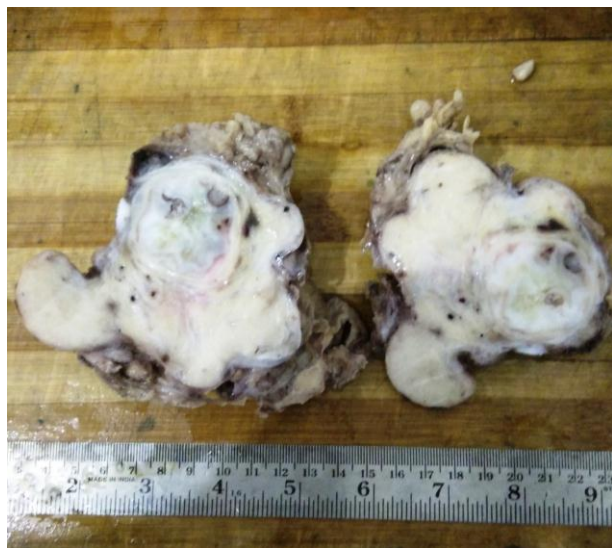


Fig. 1: Cut section of the excised mass showed large rounded grey chalky white calcified mass with glistening white surface in the center with peripheral fatty proliferation.

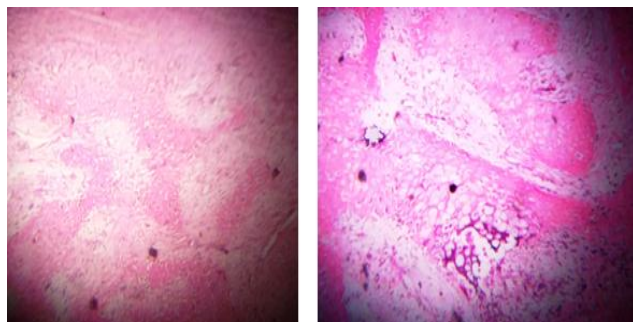


Fig. 2,3: Photomicrograph showed characteristic zonal pattern of cartilage interspread with bone with fibroblastic proliferation (H &E stain, x100)

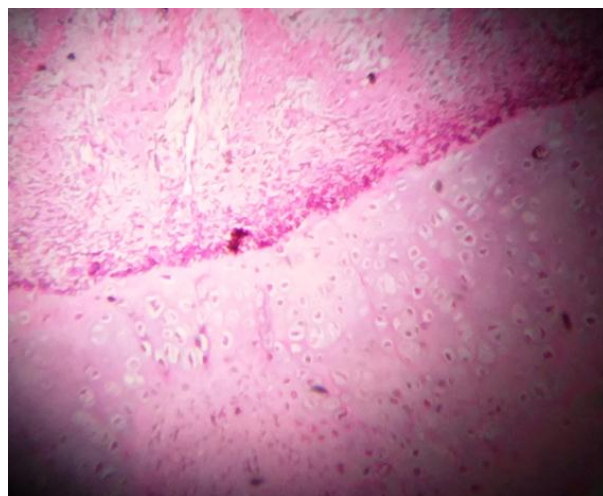


Fig. 4: Photomicrograph showed early trabeculation of ossifying areas and a peripheral zone of osteoid trabeculae rimmed by osteoblasts (H &E stain, x400)

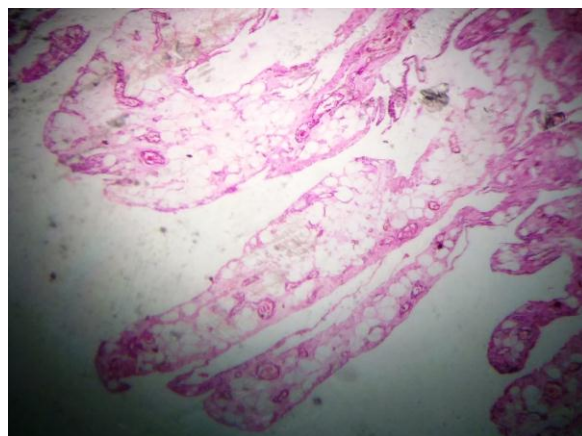


Fig. 5: The villous proliferation of the synovium contain abundant amount of fat with many congested blood vessels and sparse mononuclear cells infiltration (H &E stain, x100)

Discussion

Myositis ossificans is a benign, ossifying soft-tissue lesion typically occurring in adolescents and young adults with usual affinity to the large muscles of extremities.^{4,5} There are various terminologies to describe MO as 'pseudomalignant osseous tumor of soft tissue' or 'non-neoplastic heterotopic ossification' in the muscles and soft tissue.^{4,5} Out of three subtypes of MO, myositis ossificans traumatica constitutes the major (70%) type.⁵ The same is true in our case that is blunt trauma to knee joint after fall from bicycle. However, the exact etiology is unknown but damage to muscles with subsequent proliferation of connective tissue and differentiating into mature bone may be suggested. The remaining of tissue leads to dystrophic, heterotopic fibroblastic and vascular proliferation leading to ossification.³

Routinely on gross –MO is well circumscribed. On C/S- it is white, soft, gelatinous (or hemorrhagic) in the center with yellow-gray and firm with a rough granular surface at the periphery.^{3,4} We have match these findings in our case.

Microscopically, myositis ossificans is characterized by a distinct zonal pattern. There are three distinct zones: a central portion in which there is varied fibroblastic cellular proliferation, pleomorphic characteristics and numbers of mitotic figures; an intermediate portion where there is collagen and osteoid deposition among proliferating spindle cells, with early trabeculation of ossifying areas; and a peripheral zone of osteoid trabeculae rimmed by osteoblasts, with bone surrounded by loose fibrous tissue and atrophic fat. Characteristically, in myositis ossificans the bone formation is most prominent at the periphery of the lesion.^{3,4}

The main differential diagnosis of myositis ossificans includes extraskeletal bone-forming lesions such as fibrodysplasia (myositis) ossificans progressiva and osteosarcoma. Histopathologically and radiologically we can differentiate these entities. Fibrodysplasia (myositis) ossificans progressiva is a rare, slowly progressive hereditary disease that principally affects children under the age of ten years. It is associated with symmetrical malformations of the digits (especially microdactyly) and other skeletal malformations. The disease is characterized by progressive fibroblastic proliferation and subsequent calcification and ossification of subcutaneous fat, muscles. Unlike localized myositis ossificans, the ossification occurs in the center of the nodules. Differentiating between MO and extraskeletal osteosarcoma is critical. The zoning pattern of growth in myositis ossificans is an important diagnostic point. Osteosarcoma displays disorderly growth of atypical hyperchromatic and pleomorphic tumor cells. Bone deposition in osteosarcoma is haphazard and disorganized, sometimes with a "reverse zoning effect", with bone formation in the center of the lesion and immature spindle cells toward the periphery. Mitotic figures are present in both the immature, central portion of myositis ossificans and osteosarcoma, but myositis ossificans lacks the atypical or tripolar mitotic figures characteristic of osteosarcoma.^{3,4} An adequate open biopsy will display the

zoned pattern and exclude malignant entities like osteosarcoma. Because myositis ossificans is a benign self-limiting process, the prognosis is excellent. Local excision is curative.^{5,6}

Lipoma arborescens (LA) is a rare benign intra-articular lesion characterized by villous proliferation of the synovial membrane also named as diffuse lipoma of joint or synovial lipoma.⁷ The etiology is still remains unclear but it is a rare cause of chronic monoarticular arthritis, with only a few cases reported in the literature.^{7,10} It is most commonly seen in the knee, but cases in other joints also.⁷ *Lipoma arborescens* typically affects adults. It most commonly involves the knee, but other locations have also been described. People present with joint pain, swelling, and effusion. The diagnosis is based on the typical appearance on MRI, and the recommended treatment is open or arthroscopic synovectomy. Since its nomenclature in 1957 by Hoofa/Arzimanoglu, less than 100 cases have been reported in the literature.¹¹ The peak incidence is between the third and fifth decades, with a predilection for male.¹¹ There are only a few cases described in literature about children. The exact etiology is unknown, although it can occur without antecedents, it is associated in the literature with trauma, inflammation, or inflammatory joint diseases, neoplasm, and degenerative conditions (osteoarthritis).^{6,11} The hypothesis of lipoma arborescens being a reaction to chronic inflammation is supported by the histological finding of a villi filled with mature fat cells and hyperemic capillaries with mononuclear cell infiltrate in the underlying synovial membrane.^{12,13} However, the combination of lipoma arborescence and myositis ossificans not found in the present literature though we encounter several separate entities of both. Hence we published this case with unusual combination and rarity.

Conclusion

Due to tumor mimics and varied presentation as hard mass, myositis ossificans pose a diagnostic challenge to clinicians and pathologists. Histopathology has a key role in diagnosis of both MO and LA due to its characteristic morphology of zonal distribution of osteoid formation and villous lipomatous proliferation of synovial membrane respectively.

We report this with aim to highlight the unusual combination of Hoffa's disease with Myositis ossificans as not described in the literature.

Conflict of Interest: None.

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How to cite this article: Nikumbh DB, Mujawar P. Myositis ossificans with Hoffa's disease (Lipoma Arborescens): Usual mimicker of soft tissue sarcoma. *Arch Cytol Histopathol Res* 2019;4(2):173-6.