

Proliferative fasciitis, a rare pseudosarcoma

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Abstract

Of the many benign mimickers of soft tissue sarcoma, proliferative fasciitis is a rare entity which is a diagnostic challenge for pathologist. Recognition of this pseudo sarcoma is very essential as it aids in the appropriate management of the same. Other members of the pseudo sarcoma family are proliferative myositis, nodular fasciitis, inflammatory pseudotumor and proliferative peribursitis. We present 3 cases of proliferative fasciitis which was reported in our hospital in the last 10 years, between 2007 and 2017.

Keywords: Pseudosarcoma, Proliferative fasciitis, Rare soft tissue pseudo sarcoma.

Introduction

Proliferative fasciitis (PF) is a benign, reparative process that manifests as an acute onset-rapidly growing, tender mass, common site of occurrence being in the upper and lower extremities especially in the forearm and thigh and rarely involving shoulders, chest and face. Due to its rapidly progressive nature, clinically it raises the suspicion of sarcoma. It is commonly seen in middle age and older adults between 40-70 years (mean age being 54 years). There is no gender or race predilection. Clinically patients present with acute onset, painful, palpable, firm, tender, subcutaneous nodule(s) which is freely mobile and unattached to the overlying skin. There is usually a history of trauma prior to the development of the subcutaneous nodule.

Case Presentation

We present three cases of proliferative fasciitis reported in our Institution between 2007 and 2017. All the three cases had in common the clinical suspicion of sarcomas due to the rapidly progressive, painful and tender nature of the swelling. Two cases were located in the arm and one in the thigh. Two cases were male/smokers and one female. All were above the age of 50 years. 2 out of 3 patients gave history of Type 2 DM.

Radiology of two cases were normal and non-contributory. In one of the cases, ultrasound showed well-defined heterogenous, hypoechoic lesion that raised the possibility of a soft tissue tumor. FNA of one of the cases suggested a spindle cell lesion. Excision was performed on all the cases.

Histopathological examination of all the cases showed spindle cells which are the proliferating fibroblasts and myofibroblasts interspersed with ganglion-like cells which are specific for proliferative

fasciitis. One case showed superficial infiltration of the deeper muscles. One case showed few typical mitosis.

Immunohistochemistry on all cases showed positivity of the lesional cells to vimentin and SMA. Variable positivity for CD68 and negative for S100, beta catenine, desmin and CD34 which favoured the diagnosis of Proliferative fasciitis.

A close follow up of patients were carried out with no recurrence till date.



Fig. 1: Gross picture –proliferative fasciitis- grey white lesion with in the muscle fibres

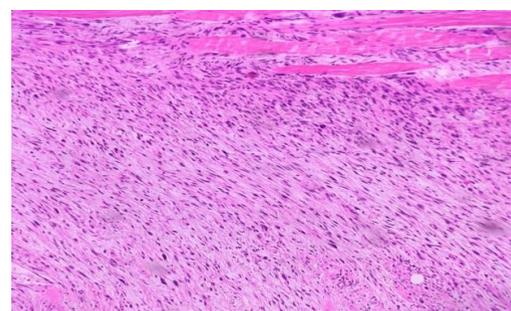


Fig. 2: H&E x100x proliferating bipolar spindle cells with few ganglion-like cells

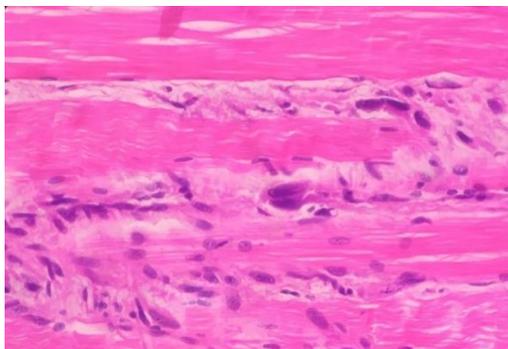


Fig. 3: H&E.x200x. Ganglion-like cells and spindle cells in between muscle fibres

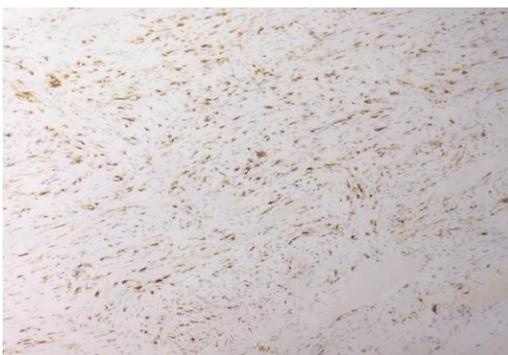


Fig. 4: IHC.CD68 x40x. Positive nuclear stain in the spindle cells

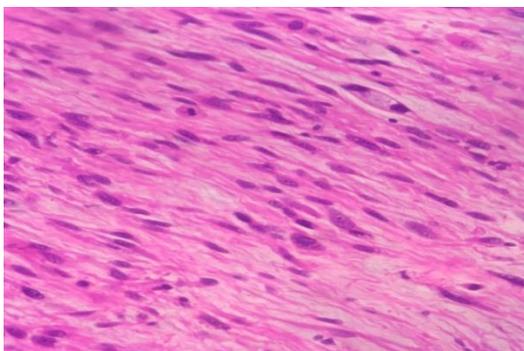


Fig. 5: H&E.100x Proliferating spindle cells showing mitosis

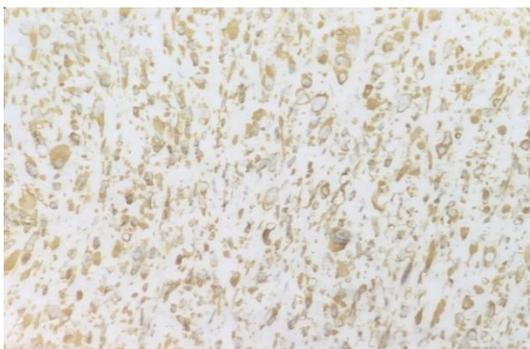


Fig. 6: IHC for vimentin x100x Picture shows positive staining for vimentin in the cytoplasm and membrane of the spindle cells

Discussion

The most common conditions to be considered under the family of 'Pseudo sarcomas' apart from proliferative fasciitis, showing fibroblasts and myofibroblasts proliferations are proliferative myositis, nodular fasciitis, inflammatory pseudotumor and proliferative peribursitis.

Nodular fasciitis is the most common condition mistaken for a sarcoma, which is a myofibroblastic proliferation characterised by extremely rapid growth (2 to 3 cm in weeks time). It occurs between the age of 20 and 50 years, affecting men and women equally. The usual locations are forearm, arm, face, shoulder etc., clinically it appears as a well circumscribed nodular lesion. Grossly, appears as a grey white nodule with myxoid areas unlike proliferative fasciitis.

Microscopically, nearly all lesions have the characteristic low power architecture called 'Zonation effect'. The centre of the lesion is hypocellular with eosinophilic fibrinous area with a more hypercellular periphery with small vessels in lobular array abutting a collagenous zone. The fibroblasts appear as bipolar spindle cells and the myofibroblasts appear as tripolar or stellate shaped cells with no atypia, arranged in a loose "tissue culture"- like manner. There are 5 subtypes, namely usual type, reactive type, cellular type with microcysts, metaplastic type with focal osteoid or chondroid metaplasia and the proliferative type which is same as proliferative fasciitis. Inflammatory cells composed of lymphocytes and macrophages may be present which will be positive for CD68. Plasma cells and neutrophils are unusual in nodular fasciitis. If they are present, a possibility of inflammatory fibrous histiocytoma should be raised. Molecular studies of this condition has showed rearrangement of UPS6 locus in most of the cases, resulting in an MYH-UPS6 gene fusion. This recent finding has now disproved the idea that Nodular fasciitis being a reactive process, as believed by many in the past.

By Immunohistochemistry, the lesional cells stains positive for SMA and MSA, but often negative for desmin. Most cases contain 1-5 mitosis/HPF. This makes it a classic example of a non-malignant reactive lesion with high mitotic activity.

Proliferative fasciitis is considered as a variant of nodular fasciitis, as it shares some common histological features like location with in deep tissue, small size, zonation pattern and loose quality of cell growth. Striking feature of proliferative fasciitis is the presence of polygonal shaped cells, ganglion-like cells having abundant eosinophilic cytoplasm surrounding a large but oval to vesicular nuclei with prominent nucleoli, dispersed in a slightly myxoid or collagenous stroma. Mitosis variably present.

Proliferative Myositis: This lesion is histologically similar to proliferative fasciitis, but in proliferative myositis the proliferation of ganglion like cells takes

place between muscle fibres (intramuscularly) that separates them, giving the appearance of “checker board” pattern at low power as against proliferative fasciitis that is confined to the subcutaneous plane.

Inflammatory Pseudotumor: Also known as inflammatory myofibroblastic tumor is a benign lesion of unknown origin (consisting of variable mix of inflammatory cells composed of plasma cells, lymphocytes, mast cells, eosinophils along with fibroblasts and myofibroblasts.) It is recognised as a neoplastic lesion associated with ALK gene rearrangement which is seen in younger age group. It usually occurs in lung parenchyma or rarely intrabronchial, skin and subcutaneous plane. Although clinically and grossly this tumor may resemble proliferative fasciitis, histology of inflammatory myofibroblastic tumor is characteristically composed of inflammatory cells along with fibroblasts and myofibroblasts which is absent in proliferative fasciitis. By Immunohistochemistry, Inflammatory myofibroblastic tumor shows spindle cells positive for vimentin, SMA, ALK (40%). Plasma cells positive for Kappa and Lambda.

Proliferative Peribursitis: This is a reactive angiomyxoid tumor-like mass occurring near joints and ligaments. It can occur at any age. Prior history of joint disease, trauma is usually present. Clinically and grossly resembles proliferative fasciitis. Rarely it can be cystic with or without synovial lining.

Microscopically there is No zonation pattern. Instead it exhibits organization in to vascular and non-vascular region which is nothing but clusters of vessels with nodular configuration in the myxoid substance with bipolar and stellate-shaped cells. These spindle cells are dispersed evenly, throughout with few lymphocytes and histiocytes. Ganglion-like cells seen in proliferative fasciitis is absent in proliferative peribursitis.

By immunohistochemistry, the spindle cells are positive for actin.

Conclusion

This rare rapidly growing pseudotumor not only misleads the clinician to think in terms of sarcoma, but also creates dilemma for a pathologist because of the presence of variably pleomorphic spindle cells, mitosis and necrosis.

In spite of the above mentioned clinical behaviour and histologically aggressive features, this condition is still a benign one that carries an excellent prognosis with surgical excision and rarely recurs. The ultimate benefit of identifying this condition is favoured to the patient as the branding of a ‘Malignant’ diagnosis and amputation is prevented.

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