

## Intranodal Palisaded Myofibroblastoma of Inguinal Region, a Rare Benign Lesion: Case Report

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### ABSTRACT:

Intranodal Palisaded Myofibroblastoma is a benign lesion characterized by intranodal proliferation of cells with myofibroblastic origin. It shows striking histopathological features characterized by spindle cells with nuclear palisading, intralesional haemorrhage, amianthoid fibers, intracellular and extracellular fuchsinophilic bodies, abundance of mast cells and compressed remnant lymphoid tissue at the periphery. Immunohistochemically the spindle cells are Vimentin, Beta catenin and SMA positive and are negative for S 100, with low ki-67 index. Palisaded Myofibroblastoma is a rare intranodal entity and it needs to be differentiated from Schwannoma which it closely resembles. Here we report a case of a 52 years old male patient who presented with mass in the left inguinal region.

**Keywords:** Intranodal Palisaded Myofibroblastoma (IPM), Amianthoid Fibers, SMA, Vimentin, Masson's Trichrome, Hematoxylin & Eosin (H&E), Immunohistochemistry (IHC)

### INTRODUCTION

Palisaded myofibroblastoma, also known as benign, lymph node based myofibroblastic tumor of unknown pathogenesis or intranodal hemorrhagic spindle cell tumor with amianthoid fibers<sup>1-4</sup>. The tumor is found almost exclusively in inguinal lymph nodes although few cases have been reported in mediastinal, pelvic and axillary lymph nodes. Tumor consist of cellular proliferation of cytologically bland spindle cells arranged in short fascicles and whorls, replacing the substance of lymph node and irregular knots of collagen bundles (the amianthoid fibres with a crystalline appearance). The spindle cells have elongated nuclei that often show nuclear palisading similar to that of nerve sheath tumor. The cells have eosinophilic and tapered cytoplasm giving them a myoid quality. There are perinuclear intracytoplasmic hyaline globules, extravasated red blood cells and proliferation of haemosiderin containing histiocytes in the lymph node<sup>1-4</sup>. Mast cells are abundant in this lesion.

It is postulated that the core of amianthoid fibers represents interstitial matrix degraded by the mast cells while the peripheral palisaded spokes are the result of Vimentin and Smooth muscle actin laid down by proliferating myofibroblasts<sup>2</sup>. There is a characteristic antigen profile which shows homogenous expression of Vimentin, Beta catenin and Smooth muscle actin as well as lack of Desmin.

### CASE REPORT

The physical examination of a 52 years old male patient who presented with a slow growing mass in the Left inguinal region, revealed a firm, non-tender and mobile swelling measuring 4x2 cm. Radiological findings were suggestive of a soft tissue mass in the inguinal region with subcentric and enlarged lymph nodes surrounding the lesion.

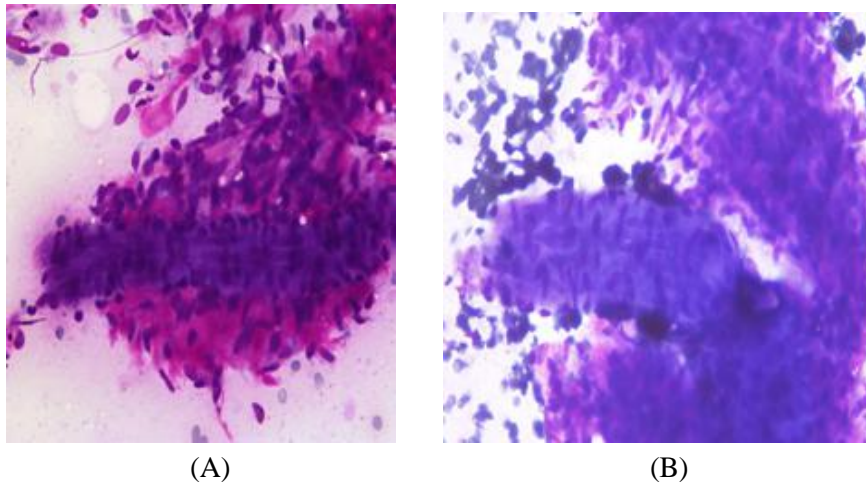
Fine needle aspiration cytology (FNAC) of the mass revealed features suggestive of stroma rich spindle cell lesion. Histopathological correlation was advised. The mass was excised and sent for histopathological examination.

Grossly, single globular encapsulated greyish white soft tissue mass was received measuring 6x4x3 cm. The cut section was solid, greyish-white with patchy red-brown areas. The microscopic examination showed spindle cell proliferation along with homogenous eosinophilic deposition, haemosiderin-laden macrophages and extravasated erythrocytes. Spindle cell nuclei displayed a patchy pattern of palisading. The collagen accumulation were recognised as "amianthoid fibers" exhibiting irregular pattern of distribution and forming stellate structure in some areas. No mitosis,

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necrosis or atypia was found in the lesion which was surrounded by compressed lymphoid tissue and a fibrous capsule.

On Masson’s Trichrome staining,the spindle cells were identified as smooth muscle and the amianthoidfibres (homogenous eosinophilic accumulation) were positive for collagen. On immunohistochemistry,the tumor cells displayed positive reaction for Vimentin, Smooth muscle actin (SMA)and Beta-catenin and negative reaction for S-100, CyclinD1,Epithelial Membrane Antigen (EMA), Cytokeratin,Caldesmon and Calponin.The ki-67 index of proliferation was below 1%.In view of the above findings the casewas diagnosed as Intranodal Palisaded Myofibroblastoma.



**Figure1.** *Microphotographs of Fine needle aspiration cytology*  
(A) *Amianthoid like structures with dense pink central stroma with central vessel,surrounded by ovoid to spindle cells in gentle swirls (H&E x 400).*  
(B) *Dense pink staining stroma with metachromatic hue with peripheral calcification (MGG x 400).*

## DISCUSSION AND REVIEW OF LITERATURE

There have been few case reports of Intranodal Palisaded Myofibroblastoma (IPM) in inguinal, pelvic, retroperitoneal, axillary and submandibular lymph nodes mainly, where it is described as a rare benign primary mesenchymal neoplasm originating from differentiated smooth muscle cells and myofibroblasts.

IPM was first described in English literature in 1968, although back then it was not classified as IPM. This current name was adopted because of its reflection of a myofibroblastic origin as well as prominent palisaded spindle cells histologically<sup>1-6</sup>.

It is essential to differentiate IPM from other soft tissue tumors such as Schwannoma, Intranodal leiomyoma, Kaposi’s sarcoma, inflammatory pseudotumor, solitary fibromastocytic tumor, angiomyomatous hamartoma and metastatic spindle cell lesions in the lymph node. The clinical history, examination, characteristic histological and immunohistochemical features aid in the correct diagnosis of IPM. Schwannoma is quite uncommon in the inguinal region and it is positive for S 100 which differentiates it from IPM. Absence of Epstein Barr Virus (EBV) DNA, negative immunostaining for HHV 8 and immunocompromised status favors the diagnosis of IPM rather than Kaposi’s sarcoma. Also, there are no slit-like vascular channels, extravasated RBCs and no hyaline globules as seen in Kaposi’s sarcoma<sup>2-9</sup>.

Although the previous studies have provided comprehensive analysis of the characteristic clinicopathologic features of IPM, its pathogenesis remains unknown. In order to decipher a key to its etiopathogenesis, Leskin et al did a clinicopathologic immunohistochemical and molecular genetic study in 18 cases of this rare entity, 15 from inguinal lymph nodes, one neck node and two undesignated lymph nodes. Screening was done for mutation in the beta-catenin gene glycogen synthase kinase-3(B) phosphorylation mutational hotspot region in Escon-3 using PCR amplification and sanger sequencing. The results demonstrated that mutational activation of the beta-catenin 1 (CTNNB1) gene is likely a pivotal event in the pathogenesis of IPM. Based on their study, IPM could be added to the list of mesenchymal neoplasms driven by gain of function CTNNB1 mutation<sup>5</sup>.

Most of the cases have been reported in Inguinal lymph node where the tumor cells were positive for SMA and Beta catenin but negative for Desmin, CD-34, S-100 and CD 117 by immune histo

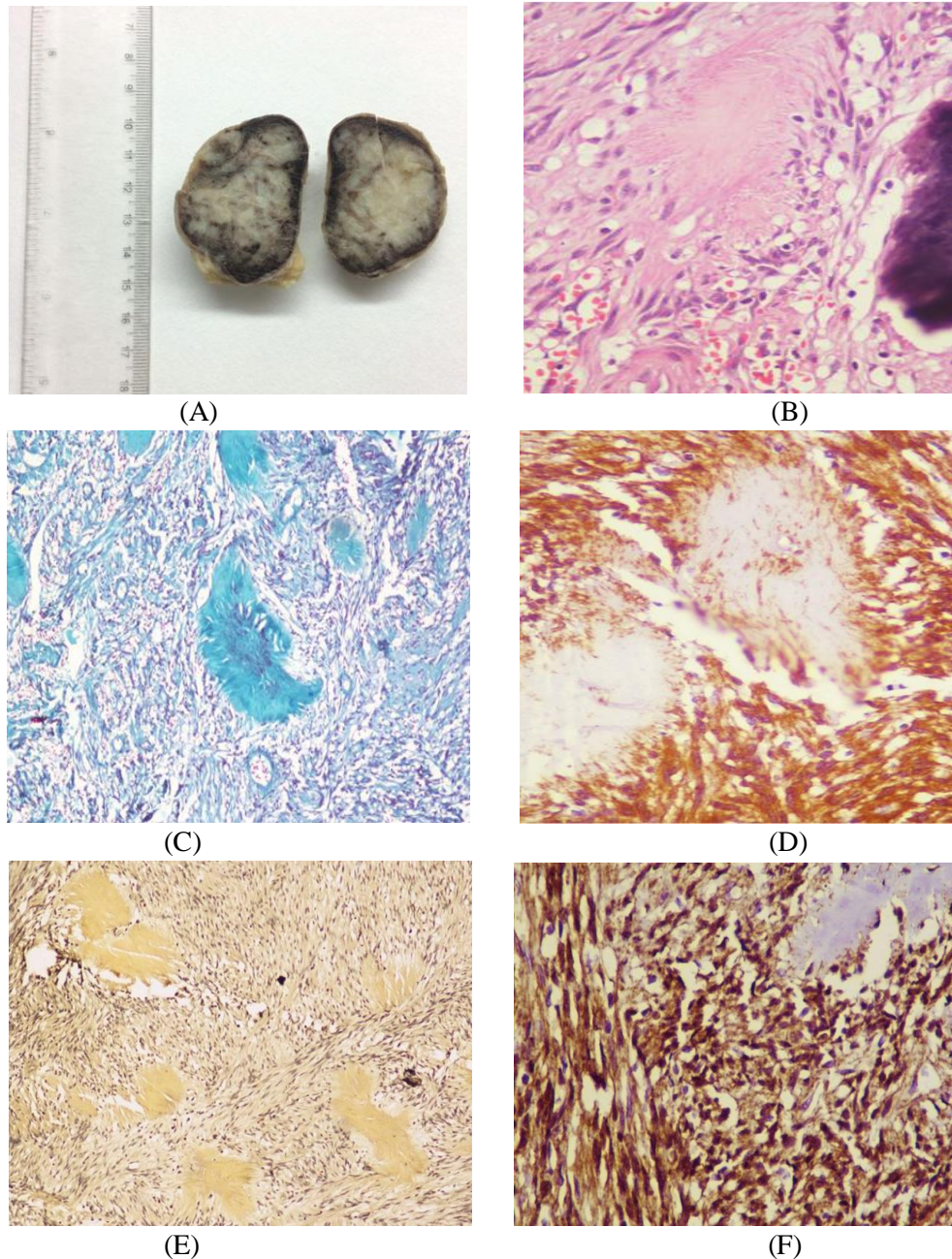


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chemical (IHC) staining. A higher concentration of myofibroblasts in the inguinal lymph nodes as compared to other lymph nodes in the body, due to increased drainage area and function, might explain its more common occurrence in the inguinal region.

Among the rare sites of occurrence, Sagar et al<sup>5</sup> reported a case with characteristic clinicopathological and IHC profile originating from retroperitoneum (an unusual origin)<sup>10</sup>. Creager reported a case of recurrent IPM in a 49yr old woman, who had cadaveric renal transplantation, the recurrent IPM presented with metaplastic bone formation<sup>11</sup>.

Hence in presence of intranodal spindle cell proliferative lesion IPM always needs to be considered and ruled out by employing full spectrum of IHC.



**Figure2.**

- (A)** Cut section of gross specimen of Inguinal lymph node.
- (B)** Microphotograph showing spindle cell proliferation in a palisading pattern around Amianthoidfibers(left) and peripheral calcification (right) (H&E x 400).
- (C)** Trichrome stain highlighting the differential staining pattern of amianthoidfibers (x400)
- (D)** Strong expression of Smooth muscle actin (SMA) in spindle cells (SMA.Biocare .IgG2a, RTUx400).
- (E)** Reticulin stain enhancing the peripheral stellate appearance of amianthoidfibers (x400).
- (F)** Strong nuclear expression of Beta catenin in spindle cells (x400).

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