Abstract:
Intranodal Palisaded Myofibroblastoma (IPM) also called Intranodal hemorrhagic spindle cell tumor with amianthoid fibres is a rare benign mesenchymal tumor of lymphnode. It was misdiagnosed as schwannoma in the past. The lesion typically presents as a painless, solitary swelling with a great predilection to inguinal nodes. Approximately 55 cases have been reported in the literature since its characterization in 1989. We present a 45 year old female case with painless right inguinal mass since 5 years, who underwent surgical excision and diagnosed as IPM on histopathological examination and confirmed by histochemical stains and Immunohistochemistry.

Key words: Amianthoid; Intranodal; Mesenchymal; Palisaded myofibroblastoma; Spindle cell tumor

Introduction:
Intranodal palisaded myofibroblastoma (IPM) also called Intranodal hemorrhagic spindle cell tumor with amianthoid fibres is a very rare benign neoplasm of lymphnode arising from myofibroblasts. This lesion was described as malignant neurilemmoma in the past. The lesion was well characterized as a distinctive entity in 1989 simultaneously by Weiss et al, Suster and Rosai [1,2]. Incidence is very rare comprising of only 55 cases in the literature since 1989. IPM has a predilection for inguinal nodes, but it was even reported in submandibular and cervical nodes [3,4]. Men are more commonly affected, sixth decade being more common in the cases reported. In this study, we present a new case of IPM.

Case Report
A 45 year old female presented with a painless, slow-growing swelling in the groin which she incidentally noticed 5 years back, complained of recent increase in size since 2 months.

General examination revealed no respiratory illness, no generalized lymphadenopathy and no organomegaly. On Local examination, 4x4 cms solitary, non tender, mobile lymphnode swelling was noted in the right inguinal region. Routine laboratory investigations were within normal limits and she was
Discussion

IPM is a rare benign mesenchymal lymph node tumor. Initially, it was defined as a malignant neurilemmoma, first by Deligdish et al in 1968, and later by Katz et al in 1974. In 1989 simultaneously Weiss et al, Suster and Rosai described it as a distinctive benign spindle cell tumor, arising exclusively from the lymph nodes and named Intranodal palisaded myofibroblastoma [1,2]. The histogenesis is incompletely understood. The cell of origin is likely the myofibroblast. This is supported by positive immunostaining for vimentin, SMA and negative for desmin, as well as ultra structurally by the evidence of elongated cells with long nuclei and abundant filaments. Vascular or capsular smooth muscle cells undergo myofibroblastic differentiation and give rise to tumor as evidenced by immunohistochemistry. The overexpression of cyclin D1 presented in the study of Kleist et al points to the proliferation regulatory pathway as one of the factors involved in the etiologic pathogenesis of IPM [5].

Men are more commonly affected than women with an age predilection between 19 to 80 years, 6th decade being more common age group. Though inguinal area is a common site, in rare cases it was even reported in submandibular and cervical lymph nodes [3,4].

Bigotti et al found a greater number of myofibroblasts in inguinal lymph nodes than in noninguinal controls, a feature that could be due to proliferation of myofibroblasts secondary to the increased drainage function in inguinal lymph nodes [6].

Schwannoma, kaposi sarcoma, inflammatory myofibroblastoma, hemangioendothelioma, dendritic cell sarcoma, spindle cell melanoma are considered in the differential diagnosis of IPM [7].

The lesions typically present as unilateral, solitary, slow-growing, painless inguinal lumps. Surgical excision is the treatment of choice. Excellent prognosis has been reported after surgical excision with recurrence in only 2 cases so far and no malignant transformation in the literature [8,9].

Conclusion

IPM, very rare tumor arising in the lymphnodes, has distinct histological and immunohistochemical features. It should be included in the differential diagnosis of spindle cell neoplasms in lymphnode though metastatic deposits are common than the primary spindle cell neoplasms.
References

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Figure 3A: Microscopic examination showing stellate arrangement of amianthoid fibres, nuclear palisading. (H&E, 400X).
Figure 3B: Microscopic examination, special stain Masson’s trichrome demonstrating intense blue staining of amianthoid fibres (100X).

Figure 4A: Microscopic examination showing diffuse strong positivity of tumor cells for Vimentin (Immunohistochemistry, 400X).
Figure 4B: Microscopic examination showing diffuse strong positivity of tumor cells for Smooth muscle actin (Immunohistochemistry, 400X).

Figure 5A: Microscopic examination showing Cyclin D1 positivity in more than 50% tumor cells (Immunohistochemistry, 400X).
Figure 5B: Microscopic examination showing low Ki-67 index <1% (Immunohistochemistry, 400X).