

Intranodal Palisaded Myofibroblastoma with Desmin Expression

– A Brief Case Report –

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Received : August 14, 2008
Accepted : December 15, 2008

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Intranodal palisaded myofibroblastoma is a rare benign mesenchymal neoplasm of the lymph node. It is characterized by intranodal spindle cell proliferation along with amianthoid fibers and prominent hemorrhage. It has been rarely reported in South Korea. We report here on a case of palisaded myofibroblastoma that arose in the left inguinal lymph node. The tumor mass was well demarcated, and it was composed of a proliferation of benign-looking spindle cells. It showed focal hemorrhage and a fibrous pseudocapsule. The tumor cells displayed little pleomorphism, no mitotic count, and characteristic palisading nuclei and amianthoid fibers. The tumor cells were positive for smooth muscle actin, vimentin, and also for desmin, but they were negative for S-100 protein, supporting the diagnosis of myofibroblastoma.

Key Words : Lymph node; Mesenchyme; Myofibroblastoma

Primary mesenchymal tumors of the lymph nodes are very rare.^{1,2} Intranodal palisaded myofibroblastoma is a rare primary benign mesenchymal neoplasm of the lymph node. It was simultaneously described in 1989 by three independent groups and it was termed palisaded myofibroblastoma, intranodal hemorrhagic spindle cell tumor with amianthoid fibers and intranodal myofibroblastoma.¹⁻³

This tumor is histologically similar to Schwannoma, so in many past reported cases the patients were diagnosed with neuronal tumors, namely an intranodal form of schwannoma.^{1,4,5} Yet on immunohistochemical staining, the tumor cells were negative for S-100 protein, but were positive for smooth muscle actin and vimentin. In addition, immunohistochemical staining results for desmin, CD31, factor VIII, epithelial membrane antigen and keratins were negative. These findings support that intranodal palisaded myofibroblastoma probably arises from intranodal myofibroblasts or specialized smooth muscle cells.^{1,2,5-7}

These tumors have low grade histologic features and show benign clinical course, so they can be treated by simple excision.^{2,7} Only a few cases were reported with recurrences, which had

similar histologic features, compared to the original tumor, with no malignant changes.^{4,5}

Intranodal palisaded myofibroblastomas occur almost exclusively in the inguinal region, followed by the submandibular region and neck.⁷ These tumors have been found in adults ranging in age from 19-78 years, and there is a male predominance (1.5:1).^{6,7} They are usually solitary, painless, and round to oval nodular masses.

There have been about 50 reported cases of intranodal palisaded myofibroblastoma since 1989. We have not found any reported cases in the Korean literature.

CASE REPORT

A 49-year-old woman visited our hospital with a mass at the proximal region of the left leg. It had been detected about two years previously. The mass was hard with no tenderness. It was movable and slowly growing, and has rapidly enlarged recently. MRI demonstrated a well defined mass at the left inguinal

area. It was a $4.0 \times 3.0 \times 2.5$ cm sized mass with a rim and inner heterogenous enhancement. The mass was removed by total excision.

Macroscopically, the tumor was a well demarcated, soft mass with a thin fibrous capsule. The cut surface showed a grayish white color and some irregular brownish-tan areas with hemorrhage, but no necrosis.

Microscopic examination revealed a thin, collagenized fibrous capsule and rims of compressed lymphoid tissue with follicles at the periphery of the tumor (Fig. 1). The tumor cells were composed of variably intersecting fascicles of uniform, slender spindle cells. They had mildly eosinophilic cytoplasm and indistinct cell borders. The nuclei were elongated and occasionally wavy

with a tapered or blunt-ended outline. The chromatin was rather vesicular with an occasionally single eosinophilic nucleolus. There was no nuclear pleomorphism. Mitotic figures were not found. Marked nuclear palisadings reminiscent of Verocay bodies were present in some areas. A prominence of irregular collagenous hyaline stroma and characteristic acellular stellate areas of eosinophilic amorphous material (the so-called amianthoid fibers) were present (Fig. 2). There were focal interstitial hemorrhage and frequent hemosiderin deposits. A few inflammatory cells were scattered.

On immunohistochemical staining, the tumor cells were diffusely positive for vimentin, smooth muscle actin (Fig. 3A) and desmin (Fig. 3B). Immunoreactivities for actin and desmin were

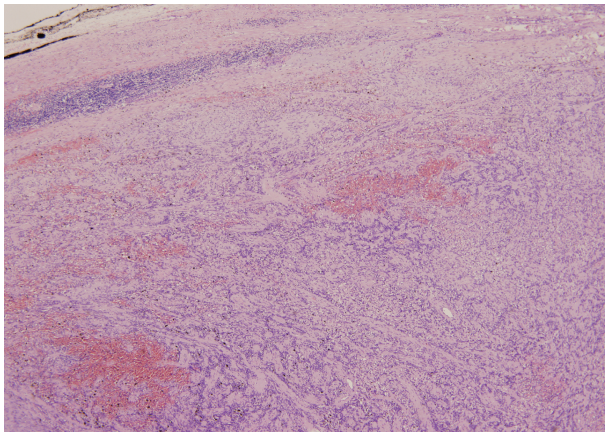


Fig. 1. The tumor shows nearly complete effacement of lymph node, and distinct palisadings.

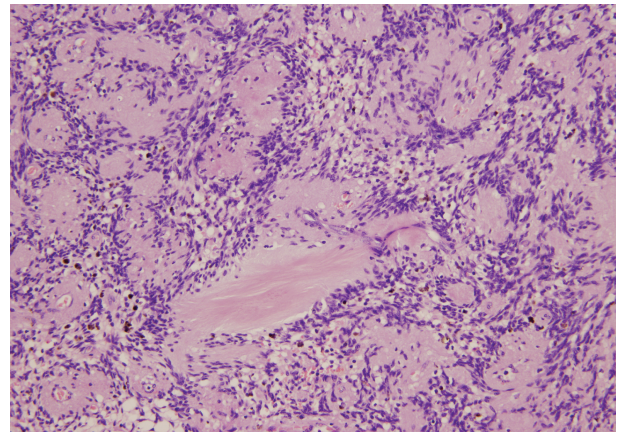


Fig. 2. The tumor has stellate areas of acellular amorphous material, the so called amianthoid fibers.

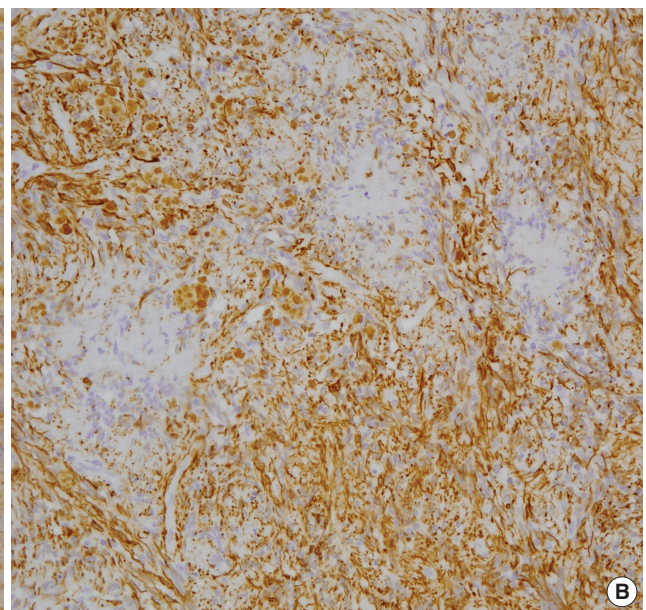
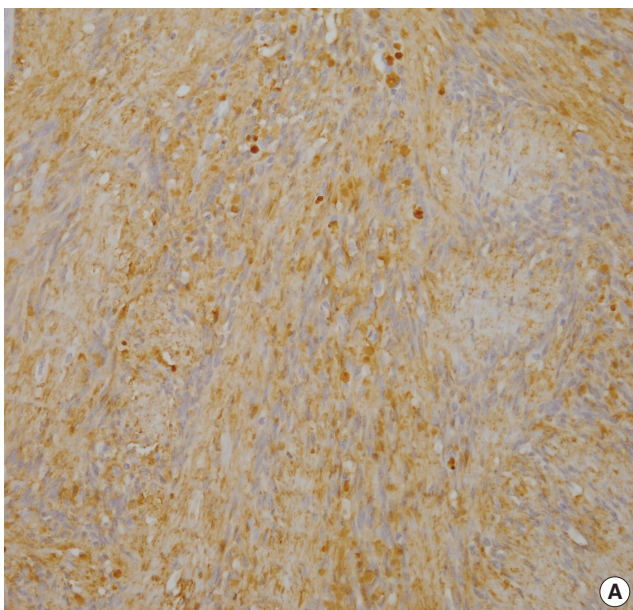


Fig. 3. On immunohistochemical staining, the tumor cells are positive for smooth muscle actin (A) and desmin (B).

stronger in spindle cell portion than in palisading areas and amianthoid fibers. The tumor cells were negative for S-100 protein, cytokeratin, EMA, GFAP, factor VIII, CD31 and CD34. The MIB1 labeling index of the tumor cells was less than 0.1%.

DISCUSSION

Intranodal palisaded myofibroblastomas are distinctive spindle cell tumors arising in the lymph nodes. Various lymph node lesions should be considered in the differential diagnosis, including intranodal schwannoma, intranodal leiomyoma, nodal involvement of Kaposi's sarcoma, inflammatory myofibroblastic tumor, angiomatous hamartoma, spindle cell vascular proliferating tumor and other metastatic tumor with spindle cell features.^{2,6,7}

The main feature that distinguishes intranodal palisaded myofibroblastoma from schwannoma is the lack of S-100 protein immunoreaction.² Intranodal palisaded myofibroblastomas usually show positive immunoreaction for actin and vimentin, and negative reaction for desmin, S-100, GFAP, CD31, CD34 and keratins. Particularly on immunohistochemical staining for desmin, most of previously reported cases were generally negative, and only one case showed focal positivity.⁷ These immunohistochemical features suggest that the spindle cells in intranodal palisaded myofibroblastomas may be derived from myofibroblasts or specialized smooth muscle cells that are present in the capsule or vascular walls of the lymph nodes.^{1,2,5-7} These immunohistochemical findings might be related to the selective occurrence of intranodal palisaded myofibroblastomas in the inguinal lymph nodes. The actin positive, desmin negative myofibroblasts or various smooth muscle cells are more prominent in the inguinal lymph nodes than in other sites.^{1,6} And yet it is expected to be much higher percentage of reactivity for desmin, because of variable percentage of desmin expression of smooth muscle cells or myofibroblasts, regardless of the previous reports. In the present case, the tumor cells showed a diffuse positive reaction for desmin, which was also expressed in some vascular walls and the stromal tissue of the pericapsular area. The tumor cells may be

derived from the smooth muscle of the vascular or capsular tissue with desmin, and so there was also positive reaction for desmin.

Immunoreactivities for actin and desmin were stronger in spindle cell portion than in palisading areas and amianthoid fibers. Amianthoid fibers are not specific, but very distinctive features of palisaded myofibroblastomas. Amianthoid fibers are a sheaf of collagen fibrils, measuring 25-1,000 nm, unlike the normal fibrils. In the present case, the stellate areas of amianthoid fibers were very weak positive for actin and desmin on immunohistochemical stain. The findings were similar to that of other reported cases.⁷ Some amianthoid fibers had small vascular lumen in the center. There is a possibility of degenerative process of perivascular collagen.⁷ But still, the mechanisms in formation of amianthoid fibers require further studies.

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