Clinical and Biological Features of an Intranodal Palisaded Myofibroblastoma

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Abstract. Intranodal palisaded myofibroblastoma (IPM) is a rare benign tumor of the lymph nodes probably arising from smooth muscle-like cells. The tumor is characterized by intranodal proliferation of spindle cells. Neoplastic spindle-cell proliferation is most often of metastatic repetition which is very important in the recognition of IPM, because it may be mistaken for metastasis or other tumors such as Kaposi’s sarcoma. We report a novel case of IPM that confirms the myofibroblastic differentiation of the tumor. The onset of IPM has been associated with Epstein-Barr virus (EBV). In addition, recently reported cases of IPM have been seen with cyclin 1 overexpression and also with human herpes virus (HHV)-8 and EBV DNA sequences. In our case, there was no evidence of HHV-8 and EBV DNA sequences and we were not able to find cyclin 1 overexpression.

Intranodal palisaded myofibroblastoma (IPM) was first described in 1989 by three different groups. They termed the tumor: palisaded myofibroblastoma, intranodal hemorrhagic spindle-cell tumor with amianthoid fibers and myofibroblastoma (1, 2).

IPM is a rare benign mesenchymal neoplasm with myofibroblastic smooth muscle differentiation, often accompanied by the formation of amianthoid fibers (3). IPM probably arises from smooth muscle-like cells that are usually present in the capsules or stroma of lymph nodes (1). The tumor is characterized by intranodal proliferation of spindle cells that have little pleomorphism and a low mitotic rate, prominent interstitial hemorrhage, focal palisading of nuclei and stellate-shaped areas of collagen reminiscent of the "amianthoid fibers" found in other tumors. Spindle-cell proliferation to lymph nodes is frequently seen as metastases from malignant melanoma, carcinoma with a pseudosarcoma feature or sarcoma. It is very important to distinguish this entity from nodal involvement caused by Kaposi’s sarcoma, a lesion closely resembling IPM.

Immunohistochemically, the tumor cells are positive for smooth muscle actin and vimentin. Electron microscopic examination demonstrated features indicative of myofibroblastic and smooth muscle differentiation (4).

Forty-eight cases of IPM have been reported in the literature since its first description. Usually, IPM arises within inguinal lymph nodes, but two cases have been described in submandibular and cervical lymph nodes (5-8). In this study, a new case of IPM is reported.

Case Report

A 62-year-old woman presented at the Department of Surgery, University of Rome “La Sapienza”, Italy, with a slow-growing, tender mass in the groin. She was discharged without complication after simple local excision and has been free of disease to date.

Pathological findings. On gross examination, the lesion consisted of a well-circumscribed mass measuring 3 cm in greatest diameter. A transectional cut showed a gray-white, focally hemorrhagic, rubbery tissue occupying the central portion of the specimen, which was surrounded by a prominent rim of hemorrhage and a thin layer of tan-white soft tissue.

Histological findings. Low-power examination revealed an outer rim of compressed lymphoid tissue separated from the central portion of the tumor by a thick band of hyalinized, sclerotic tissue that formed a pseudo-capsule. The areas located directly beneath this pseudo-capsule showed
prominent fresh hemorrhage. The central portion of the tumor was composed of fascicles of elongated spindle cells that often appeared to cross at right angles. A suggestion of nuclear palisading was observed in some areas, resulting in a vaguely neuroid configuration (Figure 1).

The tumor cells were characterized by a scant, fibrillar, eosinophilic cytoplasm with indistinct cell margins and elongated nuclei with dispersed chromatin. Only rare mitoses were found. In some areas, the cells showed prominent perinuclear vacuolization, while in other zones the cells were smaller having round to oval nuclei, scant cytoplasm and indistinct cytoplasmic borders.

A striking feature was the presence of stellate areas of collagen deposits identical to those previously described as "amianthoid fibers". Many of these structures showed a concentration of tumor cells at their periphery. In other areas, the amianthoid fibers were centered by small vessels, lined by flattened endothelial cells and contained red blood cells in their lumens simulating perivascular rosettes. The transition between these early perivascular formations and fully formed amianthoid fibers were also evident, suggesting that the latter resulted from obliteration of pre-existing vascular structures.

Small dilated vessels and foci of fresh and old hemorrhage, with deposits of abundant hemosiderin pigment, were prominent throughout the tumor. In many areas, red blood cells were seen scattered among individual tumor cells. Occasionally, inflammatory cells were admixed with the spindle cell elements, including neutrophils, small lymphocytes and plasma cells.

**Immunohistochemical findings.** The spindle cells were immunoreactive for vimentin, muscle-specific actin and myosin antibodies. The tumor cells were negative for desmin, factor VIII-related antigen, glial fibrillary acidic protein, S-100 protein and keratin antibodies.

The amianthoid fibers were immunoreactive for type I collagen. Their peripheral portions were also immuno-reactive for type III collagen and actin.

Ultrastructurally, the spindle cells had a discontinuous basal lamina. According to the literature, our case showed the same immunohistochemical findings as previously described.

**Discussion**

Intranodal palisaded myofibroblastoma is uncommon. Only 48 cases of IPM have been reported in the literature since its first description in 1989 (Table I). We report a novel case of this neoplasm within the inguinal lymph node. IPM is a benign mesenchymal neoplasm characterized by myofibroblastic smooth muscle differentiation and intranodal proliferation of spindle cells, often with the presence of amianthoid fibers (2, 9). IPM probably arises from myofibroblasts (1). This is supported by immunohistochemical analysis that showed non-cohesive, elongated cells with long nuclei and abundant filaments (4, 9, 10). Ultrastructural examination identified the tumor cells as myofibroblasts. In addition, many ultrastructural features of IPM suggested that the lesions might have originated from modified smooth muscle cells (4). Neoplastic spindle-cell proliferations are more often metastatic repetitions. It is of paramount importance to recognize IPM because it may be mistaken for metastatic or other tumors.

IPM has to be distinguished from other neoplasms (Table II) such as: Kaposi’s sarcoma, leiomyosarcoma, benign metastasizing leiomyoma, dendritic cell sarcoma, intranodal schwannoma, metastatic carcinoma, metastatic malignant melanoma, inflammatory myofibroblastic tumor.
intranodal schwannoma, metastatic spindle-cell tumor (for example, carcinoma or malignant melanoma) and inflammatory myofibroblastic tumor (3, 4, 11, 12). The presence of spindle cells and interstitial hemorrhage in a lymph node may raise concern for Kaposi’s sarcoma, which also frequently demonstrates some degree of nuclear pleomorphism and a higher mitotic rate. Sarcomas, particularly leiomyosarcoma, can simulate this benign neoplasm, but the former always exhibits some degree of nuclear pleomorphism and readily identifiable mitoses. The vague nuclear palisading and amianthoid fibers in hemorrhagic spindle-cell tumors may suggest intranodal schwannoma. However, in the former tumor the distinct Antoni A and Antoni B areas are lacking, nuclear palisading is never well developed, and there is no staining for S-100 protein. The evidence of crystalline extracellular deposits (amianthoid fibers) may help to make a differential diagnosis.

The lesions are typically presented as unilateral, solitary, painless inguinal lumps. Patients with IPM range in age from 19 to 70 years with a male predominance (5). The benign nature of these lesions is supported by IPM features and by follow-up. There was no evidence of metastases and local recurrence was reported in only two cases (5, 13).

Usually IPM arises within inguinal lymph nodes, but two cases have been described in the submandibular region and one case in the cervical region (Table I). The predilection of IPM for inguinal lymph nodes is interesting for its pathogenesis.

Bigotti et al. found a greater number of myofibroblasts in inguinal lymph nodes than in non-inguinal controls, a feature that could be due to proliferation of myofibroblasts secondary to the increased drainage function in inguinal lymph nodes (7). This study has stimulated discussion about the probable genetic or environmental etiology. The tumor has been associated with an overexpression of cyclin 1 and with infections from Epstein-Barr virus (EBV) and human herpes virus-8 (HHV-8) (9, 14). In our case, there was no evidence of HHV-8 and EBV DNA sequences, nor was cyclin-1 overexpression found.

References


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