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Intranodal Palisaded Myofibroblastoma: Radiological and Cytological Overview

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Summary

Background:

Intranodal palisaded myofibroblastoma is a benign and very rare mesenchymal neoplasm of the lymph nodes originating from differentiated smooth muscle cells and myofibroblasts.

Case Report:

We report a case of intranodal palisaded myofibroblastoma in an 84-year-old woman with Parkinson's disease that presented as a left inguinal mass. The diagnosis was made using ultrasound-guided fine needle aspiration biopsy and consequent cytopathological examination that included immunohistochemical analysis. Herein, we discuss the presentation of a rare intranodal palisaded myofibroblastoma with emphasis on its ultrasonographic and cytopathologic features.

Conclusions:

Intranodal palisaded myofibroblastoma should be considered in the differential diagnosis of inguinal lymphadenopathy and the diagnosis is possible with cytopathologic exam and immunohistochemical analysis using ultrasound-guided FNA biopsy, guiding the clinician to nodal excision rather than aggressive measures.

MeSH Keywords:

Biopsy, Fine-Needle • Histocytological Preparation Techniques • Lymphatic Abnormalities • Ultrasonography

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Background

Intranodal palisaded myofibroblastoma (IPM) is a rare benign mesenchymal tumor of the lymph nodes. These benign neoplasms arise from myofibroblasts or smooth muscle fibers [1]. IPM often presents as lymphadenopathy of the groin, but a few cases involving the axillary, mediastinal, retroperitoneal and submandibular lymph nodes have also been reported [2].

IPM is often diagnosed clinically as mass-like lymphadenopathy that is unilateral, painless, solitary, firm, and mobile. Various symptoms may ensue as the size increases, such as discomfort, pain, and compression of local surrounding structures [3].

Ultrasound (US) is a useful diagnostic tool for examination of superficial masses and lymph nodes of the inguinal

region. Ultrasound may reveal the nodal origin of IPM as a solid structure with homogeneous or heterogeneous echogenicity. Doppler imaging may reveal its vascularity and the change in elasticity can be detected and imaged using ultrasound elastography. Strain elastography, which depicts the stiffness of soft tissue by measurement of the tissue strain induced by manual compression may be used. Computed tomography (CT) can be performed to exclude the probability of a different origin or coming from nearby structures [3]. Fine needle aspiration (FNA) of the mass may be helpful for identifying a specific pattern of cells.

Herein, we discussed diagnostic findings regarding the sonographic pattern and cytological analysis of this rare tumor that had not been discussed previously.

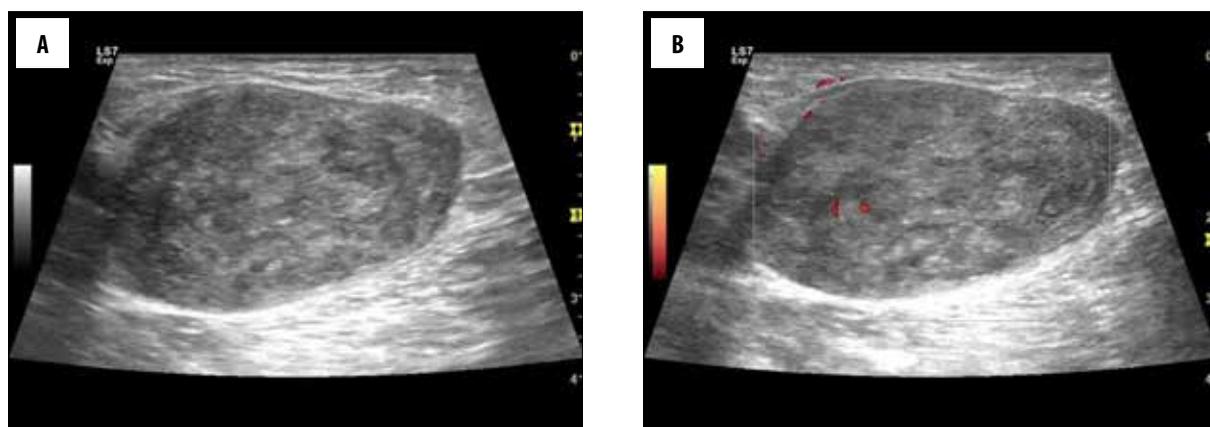


Figure 1. A well-demarcated ovoid-shaped mass with mixed echogenicity in the left inguinal area on ultrasound (A). Power Doppler imaging revealed minimal vascularity within the mass (B).

Case Report

An 84-year-old female patient, who had complaints of discomfort and a gradually increasing palpable mass in the left groin, underwent a US examination to investigate the cause of the swelling.

The patient had a medical history of Parkinson's disease, asthma and osteoporosis with no evidence of another mass elsewhere. Because of those diseases, she was on relevant medications.

An ultrasound examination was performed with a Logiq S7 Expert (GE Healthcare, Milwaukee, WI) equipped with a 9L-D linear-array probe. In the sonographic examination, a well-demarcated hypoechoic ovoid-shaped mass (46x28 mm) in the left inguinal area of probable nodal origin was detected (Figure 1A). Power Doppler imaging revealed minimal vascularity within the mass with a relatively high resistive index of 0.82 (Figure 1B). As an evaluation method, we used strain elastography with a semi-quantitative method of a 6-point scoring system. The elastographic exam revealed characteristics of hard tissue with a 4.6 score coded in blue (measurement range: 0–6; high scores and blue color denote the hardness) (Figure 2A–2C). The initial differential diagnosis according to findings of the US and elastographic exam was lymph node metastasis or lymphoma.

A fine needle aspiration biopsy was then performed using ultrasound guidance and the aspirate was smeared for cytological examination. Cellblock material was also obtained for histological examination. The initial cytological examination was suggestive of schwannoma but cellblock histomorphology and consequent immunohistochemical studies revealed that the diagnosis was consistent with intranodal palisaded myofibroblastoma (Figure 3A, 3B). The patient then underwent surgery for removal, which was in accordance with the cytopathological diagnosis.

Discussion

IPM was first described in 1989 [6–8] but in the older literature this lesion was often diagnosed or described as an intranodal form of schwannoma or as leiomyoma [3,9].

It is characterized by the proliferation of hemosiderin-laden histiocytes, spindle cells, and amianthoid fibers in the lymph node [5,7,8]. Possible viral origins are strongly suggested [1]. The lesion can be confused with metastatic lesions from malignant melanoma, carcinoma with pseudo-sarcomatous features, spindle cell sarcoma, schwannoma, or Kaposi sarcoma on cytopathological examination [3,5].

In immunohistochemical studies, spindle cells are found to be positive for smooth muscle actin, muscle-specific actin, vimentin, and negative for desmin, c-kit, carcinoembryonic antigen, keratins, CD34, S-100, HMB-45, and Epstein-Barr virus latent membrane protein 1 [10]. Spindle cells are characterized by continuous expression of actin and vimentin and a lack of desmin [3]. The histological features and immunohistochemical profile are effective in making a diagnosis of IPM [3]. Electron microscopy can be used to confirm the diagnosis [3]. When practicing cytologists encounter IPM in fine-needle aspiration, such a lesion may be reported as a low-grade spindle cell tumor and surgical excision may be recommended [3].

Although generalized lymphadenopathy is usually a manifestation of systemic diseases, localized adenopathy primarily suggests local causes. Thus, localized lymphadenopathy should prompt a search for pathology in the area of node drainage. Our patient had localized lymphadenopathy in her left side of the inguinal region. Inguinal region is a sort of peripheral lymph node groups. These lymph nodes drain lower extremity, genitalia, buttock, and abdominal wall below the umbilicus. In an adult patient, inguinal lymphadenopathy is usually caused by lower extremity infection, sexually transmitted diseases, or cancer. The patient with unexplained lymphadenopathy has a possibility of malignancy. In a study on patients with inguinal lymph node metastases, the primary site of malignancy was skin of the lower extremities, cervix, vulva, skin of the trunk, rectum and anus, ovary, and penis, respectively [11].

B-mode ultrasound is the most frequently applied imaging method for the evaluation of surface masses, such as in lymph nodes. IPM is seen on B-mode ultrasound as a solid structure with homogeneous or heterogeneous echogenicity. Doppler imaging may reveal its vascular structure. Elastography of the mass shows its elastic properties. The

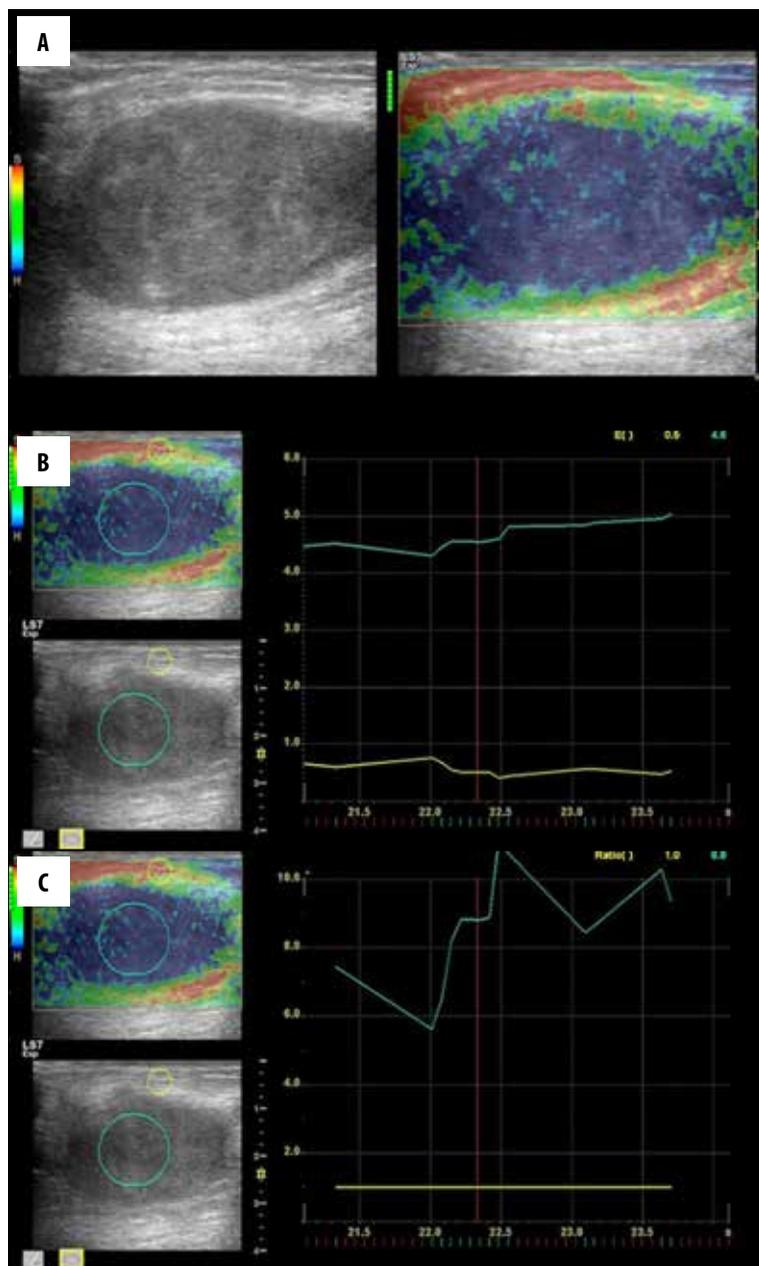


Figure 2. B-mode sonographic image showing a hypoechoic mass with a regular shape (left). Sonoelastographic image showing a predominantly blue lesion (blue color denotes the hardness) (A). Elastographic exam [Q-Analysis by GE Healthcare machine, E-Index (Elasticity Index)] revealed features of hard tissue characteristics with a 4.6 score (measurement range: 0–6; high scores denote the hardness) (B). The mass has a hardness of 8.8 fold compared with subcutaneous fat tissue on elastographic measurement (E-Ratio) (C).

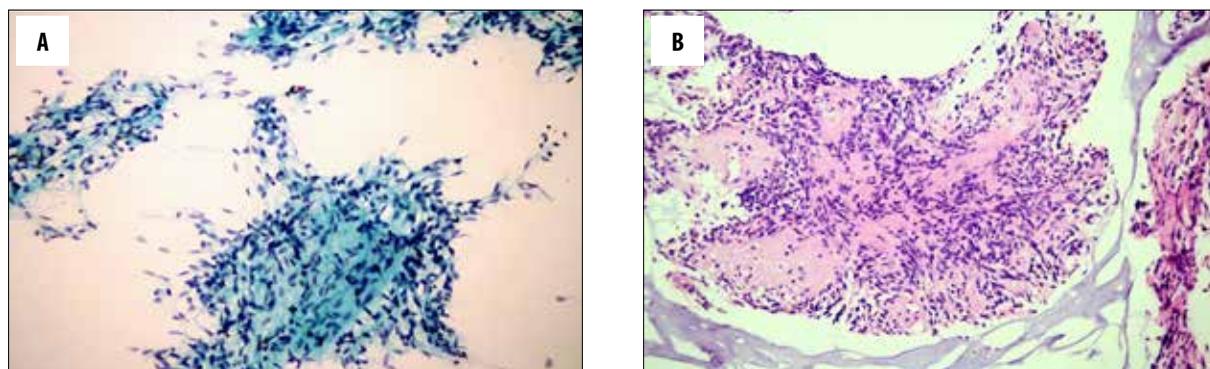


Figure 3. Spindle tumor cells showing a vaguely palisaded pattern in the collagenous matrix (cytologic feature) (A). Spindle cells showing a palisading arrangement in the collagenous background (cellblock histology) (B).

change in elasticity is detected and imaged using elastography. Sonoelastography is a new sonographic method that directly discloses the elasticity features of the tissue and allows examination of changes in tissue hardness [12]. Sonoelastography has been proposed with promising results for differentiating benign from malignant lesions in some tissues, such as breast, thyroid, salivary gland, and lymph node [13–16]. With increasing stiffness of a mass, the risk of malignancy is higher and more sensitive elasticity evaluation than palpation might be obtained with recent sonographic devices [15]. Our case with hardness features is an example of neoplasia even if it was benign. It should be noted that sonographic features, including spectral Doppler analysis and elastography, are useless for a certain diagnosis of this rare tumor. CT can be performed to exclude the probability of a different origin or coming from nearby structures. IPM is seen on CT as a well-demarcated tumor [3].

The radiological differential diagnosis is based on the origin of the tumor. With groin or axillary origin, it includes lymphadenopathy, such as metastasis, lymphoma and mass from a nearby structure, such as schwannoma or sarcoma. With retroperitoneal origin, it includes carcinoid lesions and teratoma of an undescended testis when present [2].

A single vascular pole with linear and regular branches is a sign of benignity according to the evaluation with color Doppler imaging of the lymph nodes, whereas multiple peripheral poles with distortion and displacement of the internal vessels generally indicate malignancy [17–24]. There are 2 main classes of neoplastic nodes, lymphomatous and metastatic. The presence of peripheral subcapsular vessels is a typical sign of metastasis and possible of

high-grade lymphomas, and this situation is definitely rare in lymphomas. In metastasis, neoplastic cells reach the lymph node from outside, whereas in lymphomas, the disease frequently arises inside the lymph node [17,25,26].

Most peripheral nerve sheath tumors indicate the common features of being hypoechoic and homogeneous, with posterior acoustic enhancement and peripheral nerve continuity. When peripheral nerve sheath tumors have undergone degenerative changes as calcification, posterior acoustic shadowing can also be present [27,28]. The finding of peripheral nerve continuity indicates peripheral nerve sheath tumor as the cause. But, schwannomas have also been described as eccentric [28,29]. The target appearance on sonography (hyperechoic center with a hypoechoic periphery) have been described by Lin et al. [28,30]. In addition, an internal echogenic ring, which has been described as rare but virtually pathognomonic of nerve tumors [28,29] may be seen.

Surgical excision is sufficient for treatment of IPM. No malignant transformation or metastasis has been reported. In the literature, local recurrence has been reported in six percent of cases at a low rate [1,4,5].

Conclusions

Intranodal palisaded myofibroblastoma should be included in the differential diagnosis of inguinal lymphadenopathy and the diagnosis is possible with cytopathologic exam and immunohistochemical analysis using ultrasound-guided FNA biopsy, guiding the clinician to nodal excision rather than aggressive measures.

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