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Case Report

Primary myxoid liposarcoma of the pelvis: An unusual location

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ABSTRACT

Myxoid liposarcoma is the second most common type of liposarcoma, normally located in deep tissues of the lower extremities and rarely in the mesenchyma of abdomen and pelvis We present a patient who, incidentally, showed a large pelvis mass. CT and MR revealed a loculated lesión with hypodense areas and very high signal in T2 respectively as well as heterogeneous contrast enhancement. The imaging findings of pelvic myxoid liposarcoma are nonspecific, but nevertheless a painless mesenchymal mass should be considered when we see lesions of myxoid aspect in the pelvic area without a clear relationship with defined anatomic structures.

Case report

A 57-year-old woman with a history of hiatus hernia surgery and cholecystectomy complains of persistent low back pain for 2 years. When a lumbar spine resonance was performed, a pelvic mass was incidentally discovered. The patient reported no digestive symptoms or weight loss at any time. In addition to performing colonoscopy and gynecological evaluation, a study of pelvic MRI and abdominal chest CT was performed.

The MRI showed a right parauterine mass of 6.6 × 7.1 × 7 cm in its transverse, anteroposterior, and craniocaudal axes respectively, presenting lobulated and relatively well-defined contours.

Its signal was high in the T2 Turbo Spin Echo (TSE) sequence, with diffusion restriction and hyperintense septa in the T1 TSE (Fig. 1). After administration of contrast media, a very heterogeneous enhancement was visualized.

CT images revealed a large, well-defined, and hypodense polylobulated mass with irregular enhancement and no intralesional fat areas (Fig. 2). No distant metastases were identified with total body CT imaging.

After excluding lesions in the rectum, uterus and ovaries, surgery for excision of the lesion was decided. No adjuvant therapy was performed.

Histologic examination reveals a high-grade myxoid liposarcoma with hypercellular foci of undifferentiated cells in 8% of tumor volume and rearrangement of the DDIT 3 gene

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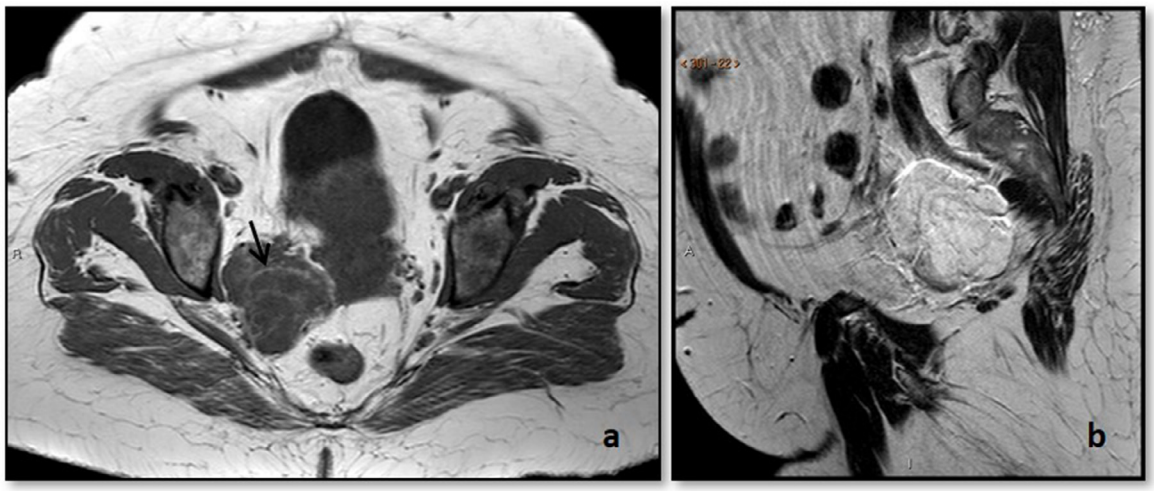

Fig. 1 – Axial T1-weighted (a) and sagittal T2-weighted (b) MR images reveal a mass predominantly high-water content (low signal intensity with T1-weighting and very high signal intensity with T2-weighting) and with fat septas in a linear pattern (arrows).

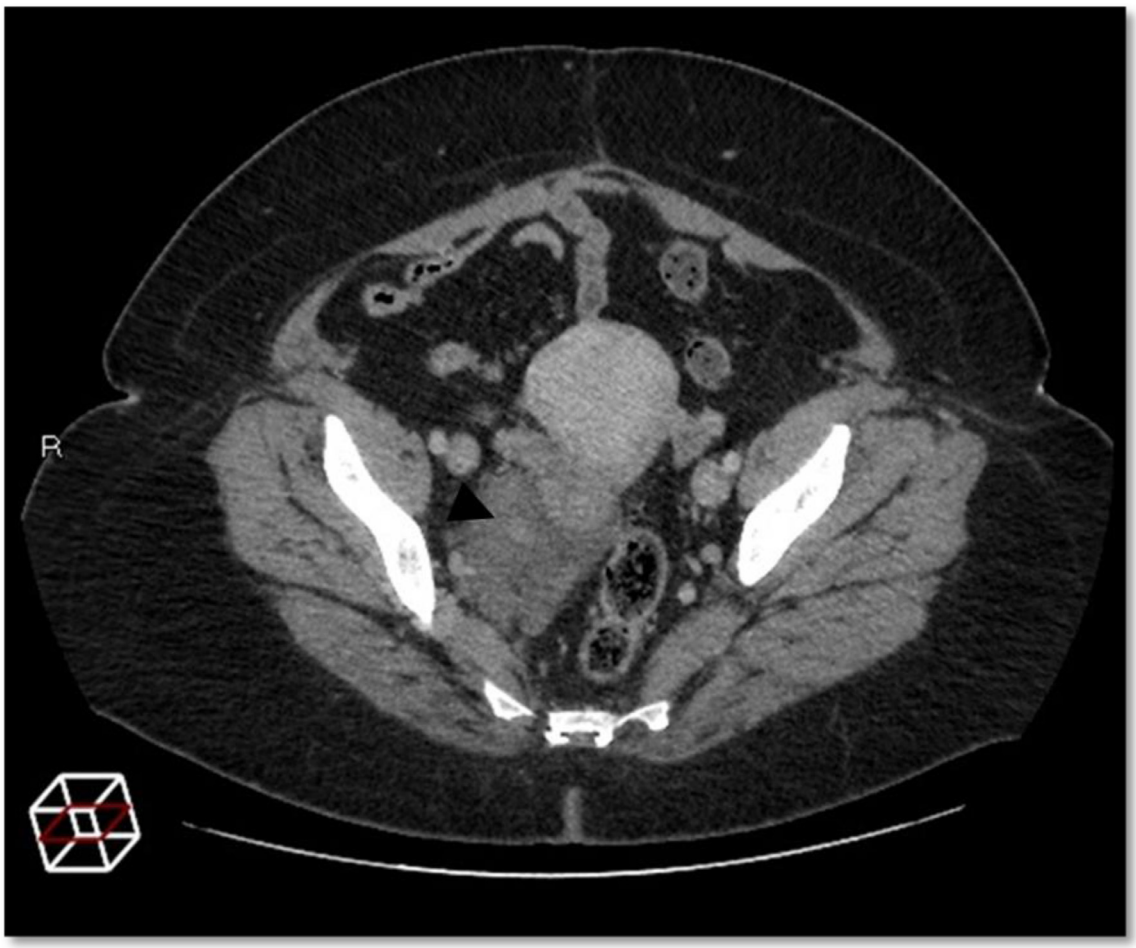

Fig. 2 – CT scan reveals a hypodense mass in pelvis of relatively well-defined margins and a nodular focus (arrowhead) of soft-tissue attenuation.

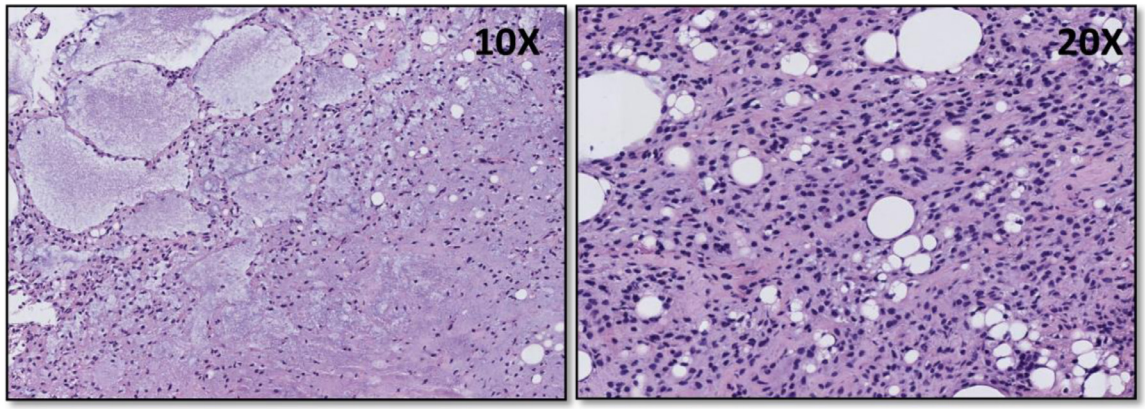

Fig. 3 – Malignant tumor composed of nonlipogenic, round or oval and uniform mesenchymal cells, interspersed with a variable number of lipoblasts, within a myxoid stroma with plexiform vascularization. In the myxoid matrix we can see lakes or cavities with mucin (H-E, 10 x -20 x).

(CHOP) in 90% of the cells evaluated, with positive resection margins (Fig. 3).

Discussion

The liposarcomas are fat-line mesenchymal malignant tumors and the most frequent soft-tissue sarcomas. The World Health Organization (WHO) Classification of Tumors differentiates four subtypes: Well differentiated, dedifferentiated, myxoid and pleomorphic.

Myxoid liposarcoma is composed of nonlipogenic mesenchymal cells, signet ring lipoblast, and myxoid stroma with a characteristic vascular pattern (chicken wire vasculature) [1]. Genetically they are associated with chromosomal translocations t (12;16)(q13-14,p-11), resulting in the formation of the TLS/CHOP fusion oncogene.

These types of lipoid tumors present as a spectrum of disease: low, intermediate, and high grade, based on the degree of cellularity (grade I <5%, grade II 5%-25% and grade III >25%, according to WHO). The last grade is associated with a high metastasis probability.

Myxoid liposarcoma tends to metastasize where fat tissue is present as the trunk or extremities, with a special tendency to retroperitoneal sites (frequently in paraspinal space) or serosal membranes, with a hematogenous spread that avoid the lung [2,3]. Patients with high-grade tumors should undergo imaging studies of chest and abdomen-pelvis MR or CT for staging and monitoring [4]. They can even occur as several tumor foci synchronously (metastatic seeding or multifocal liposarcoma) [5].

Usually, myxoid liposarcoma affects younger patients, between the 4th and 5th decade, and normally arises from the intermuscular fascial planes of the lower extremity, particularly the thigh or gluteus. Myxoid liposarcoma is extremely rare in abdomen and they appear usually as a metastasic tumor at retroperitoneal space [6].

Liposarcomas often exhibit different radiological forms which reflect a histopathological spectrum. The radiological appearance of these tumors depends fundamentally on the glycoprotein content of the mucoid material located in the extracellular matrix, but also on their fat content and cellular areas [7].

Habitually, the myxoid areas show hyperintense signal on T2- and hypointense on T1-weight images, but it depends on the concentration of proteins conditioning an increase of signal in T1 as their concentration increases. The round cell component incrementation will result in low-to-intermediate T1 and T2 signal [8,9]. On CT, the mucoid content appears as hypodense content although with higher value than water. On ultrasound they are hypoechogenic lesions with fine echoes, and posterior acoustic enhancement.

Fatty fractions are hyperintense on T1 images but in general myxoid liposarcomas do not contain substantial amount of fatty areas 15 (constitutes only a 10% of the overall mass size). Normally, these fractions are seen in septa with a linear pattern or as intralesional small nodules which some authors consider as a pathognomonic find [10].

The cellular components have nonspecific features with an intermediate echogenicity on ultrasound, attenuation similar to muscle on CT and hypointense on T1 and intermediate signal on T2 on resonance images. A high-contrast enhancement is identified in cellular component, resulting in worse prognosis. Moreover, the presence of >5% of cellular areas can be used as indicator for high-grade tumor [11]. It has been described a gradual reticular enhancement after contrast administration, resulting in a more solid appearance [12].

In general, myxoid liposarcomas are tumors with sharply defined borders or even with pseudocapsule in low-grade tumors. Bone infiltration or circular encasement of large vessels is found in high-grade tumor. Tumor size can be considerate as the most important predictor of long-term disease control. Other characteristics such as thick septa, pronounced enhancement or absence of lobulation support the diagnosis of high-grade tumors [13].

Our case presents the features of a liposarcoma with a high myxoid component and a linear pattern in its fatty content as well as cellular areas with heterogeneous enhancement. However, the atypical location for this type of tumors allowed poor diagnostic accuracy.

The differential diagnosis in our case should be made with pelvic tumors with a myxoid matrix but not related to anatomical structures, such as pseudomyxoma peritonei [7] (it usually presents as loculated collections along peritoneal surfaces of pouch of Douglas and rectovesical space, with a scalloped appearance.), myxofibrosarcoma (more frequent in extremities and with greater infiltration of neighboring anatomical structures), ancient or rearranged schwanomas [14] (more frequent in presacral space and normally oval or sphere-shaped) or angiomyxomas (mesenchymal tumor that shows involvement of both pelvic and perineal compartments with swirling appearance).

Surgical excision with or without radiotherapy is the choice treatment in located myxoid liposarcoma. Chemotherapy with trabectedin is reserved for patients at greater risk: High-grade, deep-site tumors, tumor size >5 cm or positive surgical margins. The trabectedin gets a replacement of the cellular tumor component by acellular stroma and a decrease in tissue density [15]. The systemic treatment will improve by means of a targeted therapy after the knowledge of the genomic basis of this tumor.

In conclusion, the imaging findings of pelvic myxoid liposarcoma are nonspecific, especially when it has poor fat component, nevertheless we must include it in our differential diagnosis when myxoid nature is suspected by CT or MR images and we cannot prove a clear origin in pelvic anatomical structures.

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