Clinics in Oncology

Case Report
Published: 28 Apr, 2021

Giant Perinephric Dedifferentiated Liposarcoma Masquerading Cystic Renal Tumor: A Case Report and Literature Review

*Yi Yan[1], Yong Jia[2], Mohammad Akra[4], Shady Attalla[5], Miao Lu[2] and Signy Holmes[1,3]**

[1]Department of Radiology, University of Manitoba, Canada

[2]Department of Pathology, University of Manitoba, Canada

[3]Department of Radiology & Diagnostic Imaging, Cancer Care Manitoba, Canada

[4]Cancer Care Manitoba, Canada

[5]Ain Shams University, Egypt

Abstract

Liposarcoma is one of the most common primary retroperitoneal neoplasms, and the perinephric space is a classic location. Liposarcoma displays a wide range of radiographic features depending on histological types and tumor size, and dedifferentiated liposarcoma can present with an even broader spectrum of imaging findings; therefore, the diagnosis of liposarcoma often requires tissue sampling. We present a unique case of a giant perinephric dedifferentiated liposarcoma mimicking a cystic renal tumor.

A 48-year-old man presented to the emergency department with worsening of 2 months of sciatica. Lumbar spine CT revealed disc herniation at L4-L5 level with an incidental finding of a large mass occupying the left retroperitoneum. Subsequent abdominal CT demonstrated a well-defined 20 cm cystic mass intimate to the lower pole of the left kidney that contained no solid components or foci of macroscopic fat. The favored diagnosis was cystic renal cell carcinoma. MRI was advised for further characterization, but unfortunately was terminated early due to the patient's claustrophobia. The patient was subsequently referred for urology consultation. Since the image findings suggested cystic renal malignancy, a right radical nephrectomy was performed. Grossly, the tumor showed a predominantly gelatinous and glistening cut surface with small pale yellow fleshy component. Occasional hemorrhagic areas were also present. The final diagnosis was dedifferentiated liposarcoma arising from the perinephric tissue.

Our case highlights that, when confronted with a large perinephric cystic mass on CT scan, both benign and malignant etiologies (i.e. extra-renal AML, cystic RCC and dedifferentiated perinephric liposarcoma) should be included in the differential diagnosis.

Keywords: Perinephric liposarcoma; Dedifferentiated liposarcoma cystic renal mass

OPEN ACCESS

Correspondence:
Signy Holmes, Department of Radiology, University of Manitoba, Winnipeg, MB, R3T 2N2, Canada, E-mail: sholmes@hsc.mb.ca

Received Date: *01 Apr 2021*
Accepted Date: *23 Apr 2021*
Published Date: *28 Apr 2021*

Citation:
Yan Y, Jia Y, Akra M, Attalla S, Lu M, Holmes S. Giant Perinephric Dedifferentiated Liposarcoma Masquerading Cystic Renal Tumor: A Case Report and Literature Review. Clin Oncol. 2021; 6: 1798.

Introduction

Soft Tissue Sarcomas (STS) are rare tumors arising from connective tissues, accounting for 1% of all cancers globally with an overall incidence of around five per 100,000 per year [1]. Liposarcoma is the second most common soft tissue sarcoma in adults after undifferentiated pleomorphic sarcoma. The World Health Organization classification of soft tissue tumors has classified liposarcoma into four main subgroups: Well-differentiated, myxoid/round cell, pleomorphic and dedifferentiated [2].

Liposarcoma represents up to 45% of sarcomas with retroperitoneal localization [3]. It is more frequent in men in the fifth decade of life and typically presents with non-specific symptoms induced by the tumor [4]. Well-Differentiated Liposarcoma (WDLPS) is the most common subtype, and is rarely metastatic [5]. Therefore, complete resection with a margin of uninvolved tissue is the mainstay of treatment [6]. The main adjuvant therapy is Radiation Therapy (RT), but the benefits remain controversial [2].

Dedifferentiated Liposarcoma (DDL) is an aggressive subtype of liposarcoma, which demonstrates the progression from low grade slow-growing tumor to high grade faster-growing tumor [7]. The diagnosis of dedifferentiated liposarcoma on imaging is challenging due to a

broad range of radiologic findings [8]. Herein, we present a case of perinephric dedifferentiated liposarcoma mimicking a cystic Renal Cell Carcinoma (RCC).

Case Presentation

A 48-year-old male presented with on and off low back pain radiating to the thigh for 3 months, with clinical diagnosis of sciatica. He experienced decreased appetite and mild weight loss for 6 months. Recent blood work showed hemoglobin of 71 g/L. He also had a history of non-Hodgkin Lymphoma at age 21, for which he was treated with chemotherapy and radiation to neck, chest and pelvis.

On presentation, he was referred for CT lumbar scan performed at peripheral hospital in January 2020, which showed small disc herniation at L4-L5. However, CT scan also incidentally noted a low density (HU: 20) mass to left retroperitoneum. The mass was not present on the previous scan from 2015. On exam, there was a firm, palpable mass on the left side of abdomen (Figure 1).

Further dedicated contrast enhanced CT of the abdomen and pelvis confirmed a large 18.7 cm × 21.3 cm × 18.8 cm water density mass which appeared to arise from the mid lower left kidney, compressing the upper pole with dilatation of the upper pole collecting system. It was reported as cystic lesion with no definite solid component. It did not contain foci of macroscopic fat. There were no signs of lymphadenopathy or of tumor thrombus in the renal vein. Although this lesion was felt incompletely characterized on routine portal venous phase study without pre/post contrast images, preliminary diagnosis of cystic Renal Cell Carcinoma (RCC) was suggested given the rapid growth. Patient was advised to have MRI for further characterization, but unfortunately the MRI was terminated early due to the patient's claustrophobia. Only two limited T1 and T2 weighted sequences were performed. T2-weighted images demonstrated somewhat heterogeneous hyper-intense signal which could represent a myxoid component (Figure 2, 3).

He was admitted to the hospital soon after due to increasing flank pain and dropping hemoglobin to 67 g/L. He was transfused with two units PRBCs during this admission. He underwent upper and lower GI endoscopy during admission and no pathologic findings were identified. The patient underwent a repeat CT scan after 2 weeks, which showed the lesion had increased in size to 24 cm × 22 cm but with no change in imaging characteristics.

Urology consultation was initiated. Given imaging features favoring a renal cyst or cystic neoplasm, arrangements were, made for the patient to come for surgery to either have the cyst decorticated or undergo partial nephrectomy. At surgery, it became very clear that the mass was not cystic. It was solid and could not be drained with an 18-gauge needle. The lesion was large and distended the patient's left abdomen. Exposure was challenging throughout the entire case as the mass occupied almost the entirety of the left abdomen. A radical left nephrectomy was performed. The mesentery was adherent to the mass, and sharp meticulous dissection was required for separation.

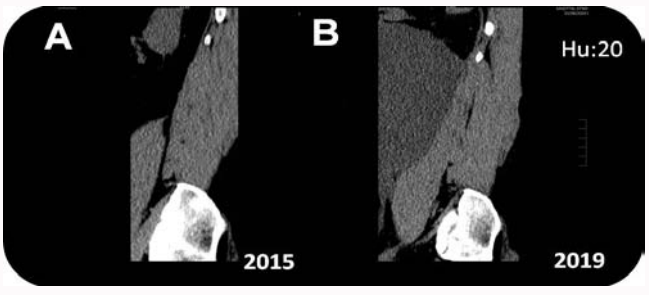

Figure 1: CT lumbar scan performed January 2020, which showed small disc herniation at L4-L5. However, CT scan also incidentally identified a low density (HU: 20) mass involving the left retroperitoneum. The mass was not present on the previous scan from 2015.

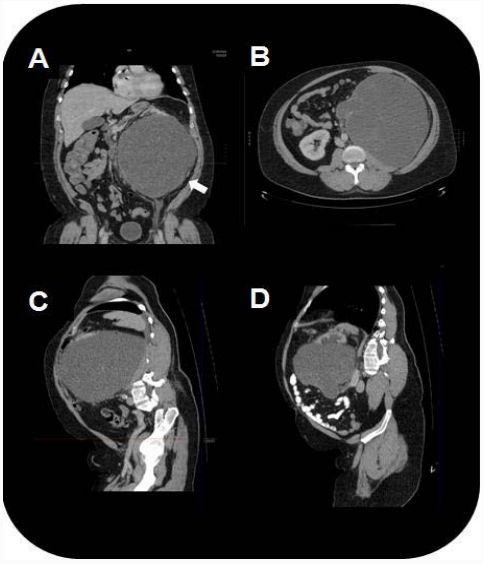

Figure 2: Further dedicated contrast enhanced CT of the abdomen and pelvis revealed a large 18.7 cm × 21.3 cm × 18.8 cm cystic mass which appeared to arise from the mid lower left kidney and compressed the upper pole with dilatation of the upper pole collecting system. The lesion appeared cystic with no definitive solid component.

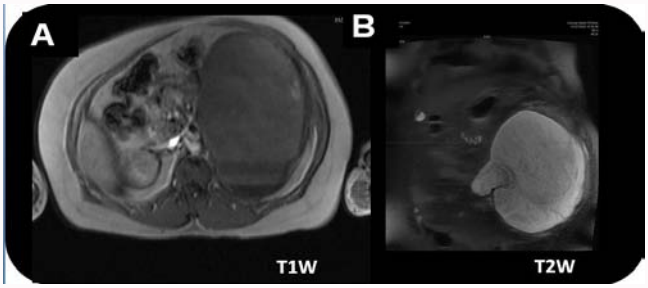

Figure 3: MRI was advised for further characterization, but unfortunately the examination was terminated early due to the patient's claustrophobia. Only two limited T1- and T2-weighted sequences were performed. T2 weighted images demonstrated somewhat heterogenous hyperintense signal. T1 weighted images demonstrated low signal intensity.

The surgical specimen weighed 5.9 kg. It was forwarded to pathology for evaluation. Grossly, the specimen measured 24 cm × 21 cm × 21 cm and was well-circumscribed and lobulated including kidney and surrounding perinephric tissue. In contrast to radiographic findings, the tumor was not felt to arise from kidney parenchyma but rather from kidney hilum, as kidney parenchyma was stretched and distorted around the side of the tumor. The majority of the tumor showed a gelatinous and glistening cut surface. There was a small white to pale yellow firm area at the periphery of the tumor (Figure 4A). Extensive necrosis and hemorrhage was identified.

Microscopically, the majority of the tumor was composed of round to spindled cells with low grade cytologic atypia in a prominent myxoid stroma with delicate blood vessels (Figure 4B). Large areas of

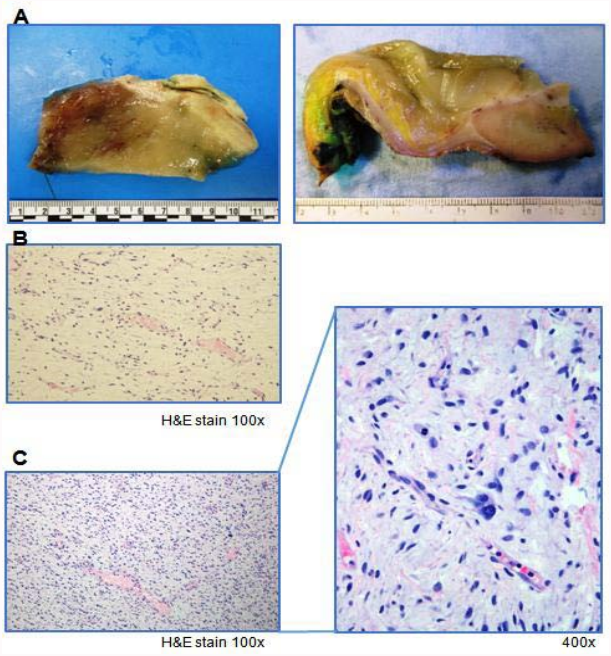

Figure 4: Gross and microscopic findings of perinephric tumor.
4A) Grossly, the tumor showed a mucinous and glistening cut surface mixed with white fleshy, pale yellow and hemorrhagic areas. 4B) Most of the viable tumor showed low cellularity with abundant myxoid stroma and delicate blood vessels. Tumor cells had low grade cytologic atypia (H&E stain, original magnification x100). 4C) There were areas demonstrating increased cellularity. Tumor cells had pronounced cytologist atypia with increased mitoses (H&E stain, original magnification x100 and x400).

tumor necrosis were present. In some areas, the tumor demonstrated increased cellularity and pronounced cytological atypia with increased mitoses (Figure 4C). A fat component was not identified either grossly or microscopically. Tumor cells showed strong diffuse positivity for CD34, and were negative for pancytokeratin, EMA, PAX8, actin, desmin, S100, SOX10, HMB45 and MART1. Ki67 index was approximately 10%. Fluorescence *in Situ* Hybridization (FISH) assessment was positive for amplification of the MDM2 (12q15) and CDK4 (12q13-14) loci. However, a fat component was absent. The histologic features and molecular study was consistent with dedifferentiated liposarcoma. The diagnosis of DDL, grade 3, was made according to the FNCLCC (French Fédération Nationale des Centres de Lutte Contre le Cancer) system.

The patient made a good recovery and no chemotherapy or radiotherapy was given. However, post-surgical CT abdomen and pelvis one month after surgery revealed a complex primarily cystic mass with small internal fat components at the left nephrectomy bed measuring 15.5 cm × 4.0 cm × 7.7 cm (Figure 5). Residual or recurrent liposarcoma could not be excluded. The mass was intimate to the left psoas muscle without a clearly defined intervening fat plane. The case was discussed during multi- disciplinary tumor board, and the decision was made to proceed with Radiation Therapy (RT) and

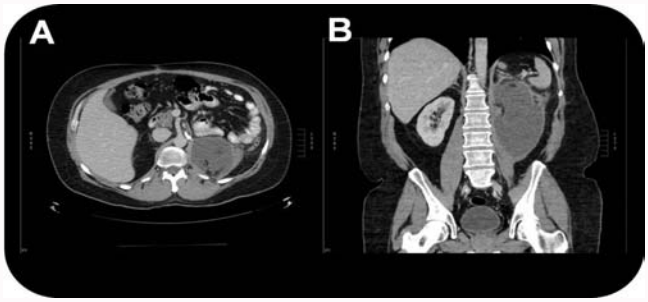

Figure 5: Post-surgical contrast enhanced CT of the abdomen and pelvis one month after surgery revealed a complex primarily cystic mass with small internal fat components at the left nephrectomy bed, intimate to the left psoas muscle without defined intervening fat plane. It measured 15.5 cm × 4.0 cm × 7.7 cm. Residual or recurrent liposarcoma could not be excluded.

imaging follow up.

Discussion

Most sarcomas cannot be characterized to cell type with imaging, with the exception of classic liposarcoma [9]. The presence of macroscopic fat on images ascertains the diagnosis of a liposarcoma. Septa and mild nodularity are often identified in well-differentiated liposarcoma. However, the presence of solid or fluid component suggests the dedifferentiation into a more aggressive tumor such as dedifferentiate subtype [6]. Nevertheless, in instances where these tumors are composed predominantly of soft tissue and fluid components, differentiating a liposarcoma from other types of perinephric tumor on images can be extremely challenging [8]. In particular, retroperitoneal dedifferentiated liposarcomas can present with a broad spectrum of imaging findings [8,10]. It has been reported that these tumors can mimic large cystic lesions such as malignant ovarian cancer on imaging [11].

Dedifferentiated liposarcoma usually contains fatty and non-fatty components [8,10]. Our case is unique in the literature as a growing dedifferentiated perinephric liposarcoma demonstrating neither microscopic nor macroscopic fat on CT or MRI. It was therefore favored to represent a cystic renal lesion such as renal cell carcinoma. This presentation is extremely rare with only three other similar cases in our knowledge of a liposarcoma mimicking a cystic Renal Cell Carcinoma (RCC) reported in the Asian population (Table 1) [12-14]. No such case is previously published in North America. Although differentiating dedifferentiated sarcoma from cystic RCC is challenging both clinically and with diagnostic imaging, the distinction is important, because prognoses for these two entities are drastically different.

Pathologic assessment is often required to reach the definitive diagnosis of dedifferentiated perinephric liposarcoma. In our case, FISH study demonstrated the amplification of both MDM2 and CDK2, confirming the diagnosis of dedifferentiated liposarcoma. According to current literature, WDLPS and Dedifferentiated

Table 1: Literatures of dedifferentiated perinephric liposarcoma.

Author	Year	Journal	CT findings	CT findings	Side	Size	Treatment	Outcome
Horiguchi et al. [12]	2002	Nihon Hinyokika Gakkai Zassh	Japan	Well-defined cystic mass of lower pole	Right kidney lower pole	12 × 12 × 8 cm	Open radical nephrectomy	No metastasis no recurrence
Kuratate et al. [13]	2010	J Med Invest	Japan	Well-defined low attenuating heterogeneous mass	Left anterior pararenal space	22 × 19 × 16 cm	Open radical nephrectomy	Recurred 10 years later
Howairis et al. [14]	2018	African Journal of Urology	UAE	N/A	N/A	14 × 9 × 3 cm	Open partial nephrectomy	N/A

Perinephric Liposarcoma (DDLPS) can share the same basic genetic abnormality characterized by a simple genomic profile with a 12q14-15 amplification involving MDM2 gene [7,15]. The majority of dedifferentiated liposarcomas arise de novo and a small percentage develops in local recurrence of WDLPS. Dedifferentiation also occurs in 10% to 15% of the WDLPS [16].

The classic microscopic appearance of dedifferentiated liposarcoma is transition from WDLPS to non-lipogenic sarcoma. However, it is not uncommon that a WDLPS component is absent [17]. It should also be noted that non-lipogenic component may demonstrate broad morphologic spectrum and many show a mixture of different patterns, including cellular pleomorphic pattern, myxoid stroma pattern, storiform pattern, inflammatory pattern, and epithelioid pattern [16,17]. In our case, the tumor shows a prominent myxoid stroma pattern. The differential diagnosis of this case includes renal cell carcinoma and soft tissue tumor with myxoid stroma [17]. Neither the histologic nor the IHC stains support the diagnosis of renal cell carcinoma. The differential diagnosis for soft tissue tumor with myxoid stroma may include myxoid liposarcoma, myxofibrosarcoma, low-grade fibromyxoid sarcoma and extraskeletal myxoid chondrosarcoma [18].

Clinical and radiology presentations can provide important clues to distinguish dedifferentiated liposarcoma from other entities, especially when WDLPS component is absent. Dedifferentiated liposarcoma is most often seen in middle aged to elderly adults and most commonly located the in retroperitoneum and abdominal cavity. Interestingly, cases of well-differentiated and dedifferentiated LPS with prominent myxoid stroma appear to show a predilection for such anatomical sites [17]. In contrast, myxoid liposarcoma most often occurs in young to middle-aged adults, arising in deep soft tissue of the extremities. The majority of myxofibrosarcomas affect patients in their 6th to 8th decades and arise in dermal/subcutaneous tissue. Low-grade fibromyxoid sarcoma is most common in deep soft tissue of the extremities and trunk. Extraskeletal myxoid chondrosarcoma is another tumor most common in the extremities. It is extremely rare that these tumors arise in the retroperitoneum or abdomen.

Microscopic findings such as vascular patterns are also helpful to point to the correct diagnosis. Myxoid liposarcoma may demonstrate classic thin-walled, branching capillaries ("chicken wire" pattern); myxofibrosarcoma usually has curvilinear vessels and low grade fibromyxoid sarcoma will show arcades of blood vessels. In our case, delicate blood vessels in the background do not demonstrate the above features. However, according to a review study done by Sioletic et al. [17]. 22 of 34 cases of DDL with prominent myxoid stroma showed a myxofibrosarcoma-like pattern, containing thin-walled elongated curvilinear blood vessels, which may represent a potential diagnostic pitfall.

Molecular studies are of great value in distinguishing dedifferentiated liposarcoma from other tumors with myxoid stroma. Myxoid liposarcoma has characteristic t (12;16) with FUS-DDIT3 fusion and extraskeletal myxoid chondrosarcoma commonly has t (9;22) with EWSR1-NR4A3 fusion [19]; none of those tumors will demonstrate MDM2 amplification.

The dedifferentiation is best defined as the presence of nonlipogenic components within the WDLPS. However, in our case, 100% nonlipogenic areas with myxoid background was demonstrated without transition zone. Interestingly, the CT image finding with lack of fatty component is consistent with nonlipogenic area, although it is technically hard to conclude no microscopic fat on imaging without opposed phase MRI. Full imaging workup could have aided in the diagnosis even without a fat component. The internal complexity and enhancing components would have been shown prior to surgery and would have impacted OR planning.

Conclusion

Our case highlights that, when confronted with a large perinephric/renal cystic mass on CT scan, both benign and malignant etiologies (i.e. extra-renal AML, cystic RCC and dedifferentiated perinephric liposarcoma) should be included in the differential diagnosis. In particular, dedifferentiated liposarcoma without WDLPS component may mimic the imaging findings of a cystic lesion with pathological features required for final diagnosis. In addition, this case demonstrates the value of a full imaging workup and multi-disciplinary approach to diagnosis and treatment of dedifferentiated perinephric liposarcoma because of potentially ambiguous clinical and radiological findings.

References

1. Hoang NT, Acevedo LA, Mann MJ, Tolani B. A review of soft-tissue sarcomas: Translation of biological advances into treatment measures. Cancer Manag Res. 2018;10:1089-114.
2. Vijay A, Ram L. Retroperitoneal liposarcoma: A comprehensive review. Am J Clin Oncol. 2015;38(2):213-9.
3. Pisani M, Al-Buheissi S, Whittlestone T. Retroperitoneal liposarcomas: A representative literature review occasioned by a rare case of laterelapse abdominal liposarcoma. Urologia. 2016;83(2):68-70.
4. Lauretti S, Cappa M, Emiliozzi P, Casareale P, Ianari A, Defidio L. Retroperitoneal liposarcoma. Minerva Chir. 1998;53(1-2):77-81.
5. Shibata D, Lewis JJ, Leung DH, Brennan MF. Is there a role for incomplete resection in the management of retroperitoneal liposarcomas? J Am Coll Surg. 2001;193(4):373-9.
6. Craig WD, Fanburg-Smith JC, Henry LR, Guerrero R, Barton JH. Fat-containing lesions of the retroperitoneum: Radiologic-pathologic correlation. Radiographics. 2009;29(1):261-90.
7. Coindre JM, Pedeutour F, Aurias A. Well-differentiated and dedifferentiated liposarcomas. Virchows Arch. 2010;456(2):167-79.
8. Hong SH, Kim KA, Woo OH, Park CM, Kim CH, Kim MJ, et al. Dedifferentiated liposarcoma of retroperitoneum: Spectrum of imaging findings in 15 patients. Clin Imaging. 2010;34(3):203-10.
9. Aga P, Singh R, Parihar A, Parashari U. Imaging spectrum in soft tissue sarcomas. Indian J Surg Oncol. 2011;2(4):271-9.
10. Sung MS, Kang HS, Suh JS, Lee JH, Park JM, Kim JY, et al. Myxoid liposarcoma: Appearance at MR imaging with histologic correlation. Radiographics. 2000;20(4):1007-19.
11. Kim H, Jeong T, Lee Y, Kim G, Hong S, Beck S, et al. A retroperitoneal dedifferentiated liposarcoma mimicking an ovarian tumor. Obstet Gynecol Sci. 2017;60(6):598-601.
12. Horiguchi A, Oyama M. Perinephric liposarcoma mimicking cystic renal tumor. Nihon Hinyokika Gakkai Zasshi. 2002;93(3):491-4.
13. Kuratate S, Chikakiyo M, Kaneda Y, Harino Y, Hirose T, Yagi T, et al. A case of perinephric liposarcoma which recurred ten years later from the initial operation. J Med Invest. 2011;58(1-2):154-8.
14. Howairis ME, Malliwal R, Bhanotc S, Buchholza N. Asymptomatic dedifferentiated liposarcoma mimicking renal cell carcinoma-A rare case report and review of literature. Afr J Urol. 2018;24(1):67-9.

15. Thway K, Flora R, Shah C, Olmos D, Fisher C. Diagnostic utility of p16, CDK4, and MDM2 as an immunohistochemical panel in distinguishing well-differentiated and dedifferentiated liposarcomas from other adipocytic tumors. Am J Surg Pathol. 2012;36(3):462-9.

16. Hasegawa T, Seki K, Hasegawa F, Matsuno Y, Shimodo T, Hirose T, et al. Dedifferentiated liposarcoma of retroperitoneum and mesentery: Varied growth patterns and histological grades--a clinicopathologic study of 32 cases. Hum Pathol. 2000;31(6):717-27.

17. Tajima S, Koda K. Paratesticular dedifferentiated liposarcoma with prominent myxoid stroma: Report of a case and review of the literature. Med Mol Morphol. 2017;50(2):112-6.

18. Sioletic S, Cin PD, Fletcher CDM, Hornick JL. Well-differentiated and dedifferentiated liposarcomas with prominent myxoid stroma: Analysis of 56 cases. Histopathology. 2013;62(2):287-93.

19. Nishio J, Iwasaki H, Nabeshima K, Naito M. Cytogenetics and molecular genetics of myxoid soft-tissue sarcomas. Genet Res Int. 2011;2011:497148.