

Case report

Open Access

## A unique dedifferentiated tumor of the retroperitoneum

Shahzeer Karmali<sup>1</sup>, Halligrimur Benediktson<sup>2</sup>, Walley Temple<sup>3</sup> and Oliver F Bathe\*<sup>3</sup>

Address: <sup>1</sup>Department of Surgery, University of Calgary, Calgary, AB, Canada, <sup>2</sup>Department of Pathology, University of Calgary, Calgary, AB, Canada and <sup>3</sup>Departments of Surgery and Oncology, University of Calgary, Calgary, AB, Canada

Email: Shahzeer Karmali - karmalis@ucalgary.ca; Halligrimur Benediktson - benedikt@ucalgary.ca; Walley Temple - walleyte@cancerboard.ab.ca; Oliver F Bathe\* - bathe@ucalgary.ca

\* Corresponding author

Published: 23 July 2004

Received: 30 April 2004

World Journal of Surgical Oncology 2004, 2:25 doi:10.1186/1477-7819-2-25

Accepted: 23 July 2004

This article is available from: <http://www.wjso.com/content/2/1/25>

© 2004 Karmali et al; licensee BioMed Central Ltd. This is an open-access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/2.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

### Abstract

**Background:** Dedifferentiated liposarcomas represent heterogeneous tumors with lipomatous and nonlipomatous elements starkly juxtaposed. It is thought that the high grade nonlipomatous elements of the tumor portend a worse prognosis.

**Case Presentation:** A 19.8 kg heterogeneous retroperitoneal tumor was successfully and completely resected. Because of its extent, no additional treatment modalities were practicable. The tumor soon recurred. The recurrent tumor differed from the primary tumor in that it was more homogeneous, consisting mainly of nonlipogenic, calcific tissue.

**Conclusions:** Dedifferentiated liposarcomas are known to have a very high recurrence rate. The biological behavior of dedifferentiated liposarcomas is likely dictated by the most aggressive element of these heterogeneous tumors.

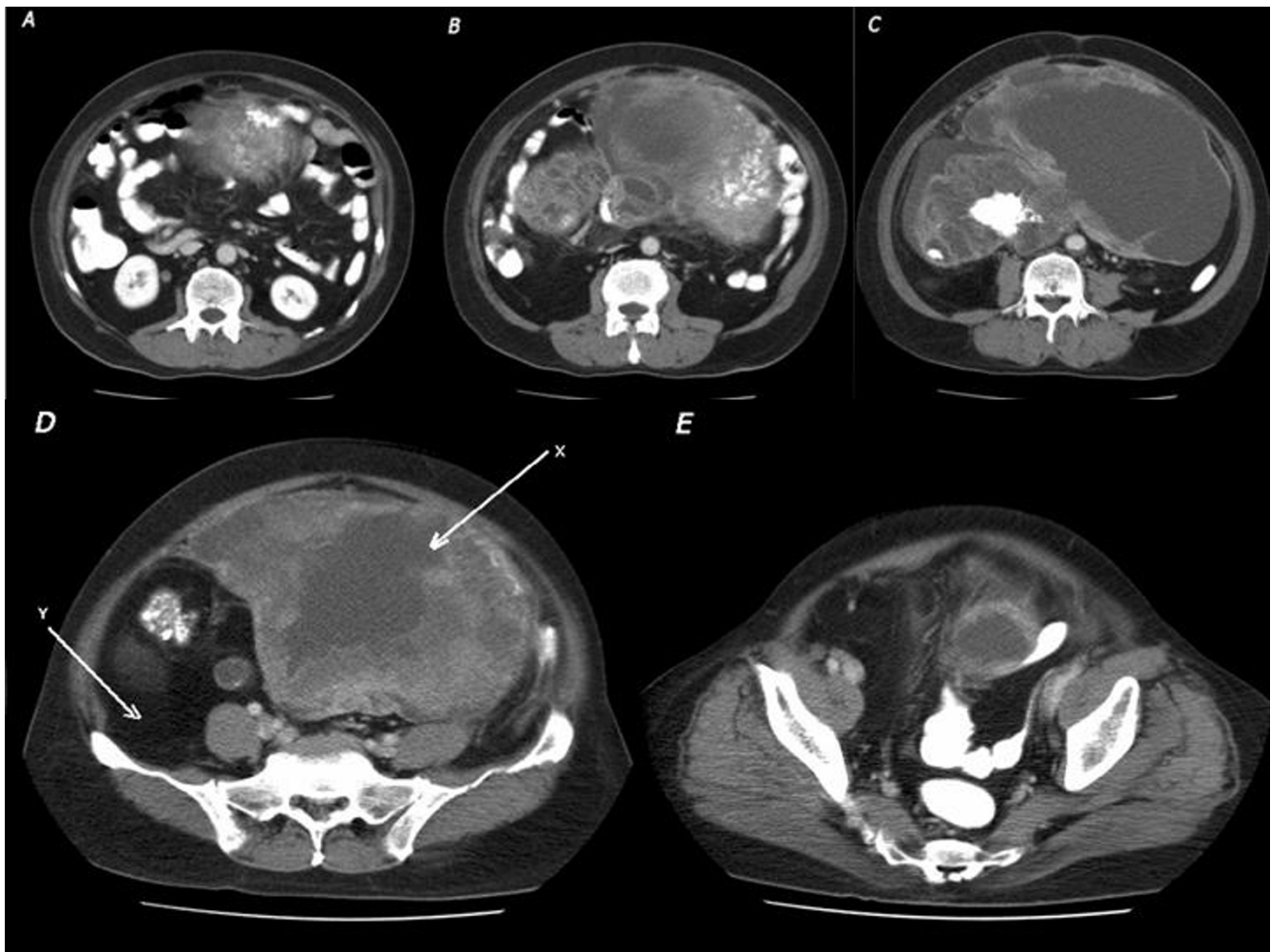
### Background

Sarcomas arising from the retroperitoneum are rare tumors, accounting for 10–15% of all soft tissue sarcomas [1]. Liposarcoma is the single most common soft tissue sarcoma and accounts for at least 20% of all sarcomas in adults [1]. Classification of liposarcoma into four types, based on morphologic features and cytogenic aberrations, is now widely accepted [2]. These four types are (a) well differentiated; (b) dedifferentiated; (c) myxoid/round cell and (d) pleomorphic. The extent of differentiation, as reflected by histological grade, remains the most important determinant of clinical course and of ultimate prognosis for patients with liposarcoma after resection. The following case illustrates the great morphological and biological heterogeneity of these tumors. A very rapid recurrence was observed, and this recurrence was considerably

less heterogeneous than the primary tumor, consisting mainly of the calcific, nonlipomatous component.

### Case presentation

A 65-year-old male presented with a three-week history of progressively worsening abdominal distension. He denied any abdominal pain but stated that he noticed an increased frequency of bowel movements. His past medical history was unremarkable. On examination, he was afebrile and had a hugely distended abdomen with an immobile, nontender mass occupying all four quadrants of the abdomen. Computed tomographic (CT) scan revealed a large, heterogeneous lobulated mass occupying most of the abdomen (Figure 1). The peripheral component appeared lipomatous and the margins of this component were difficult to estimate accurately. There was



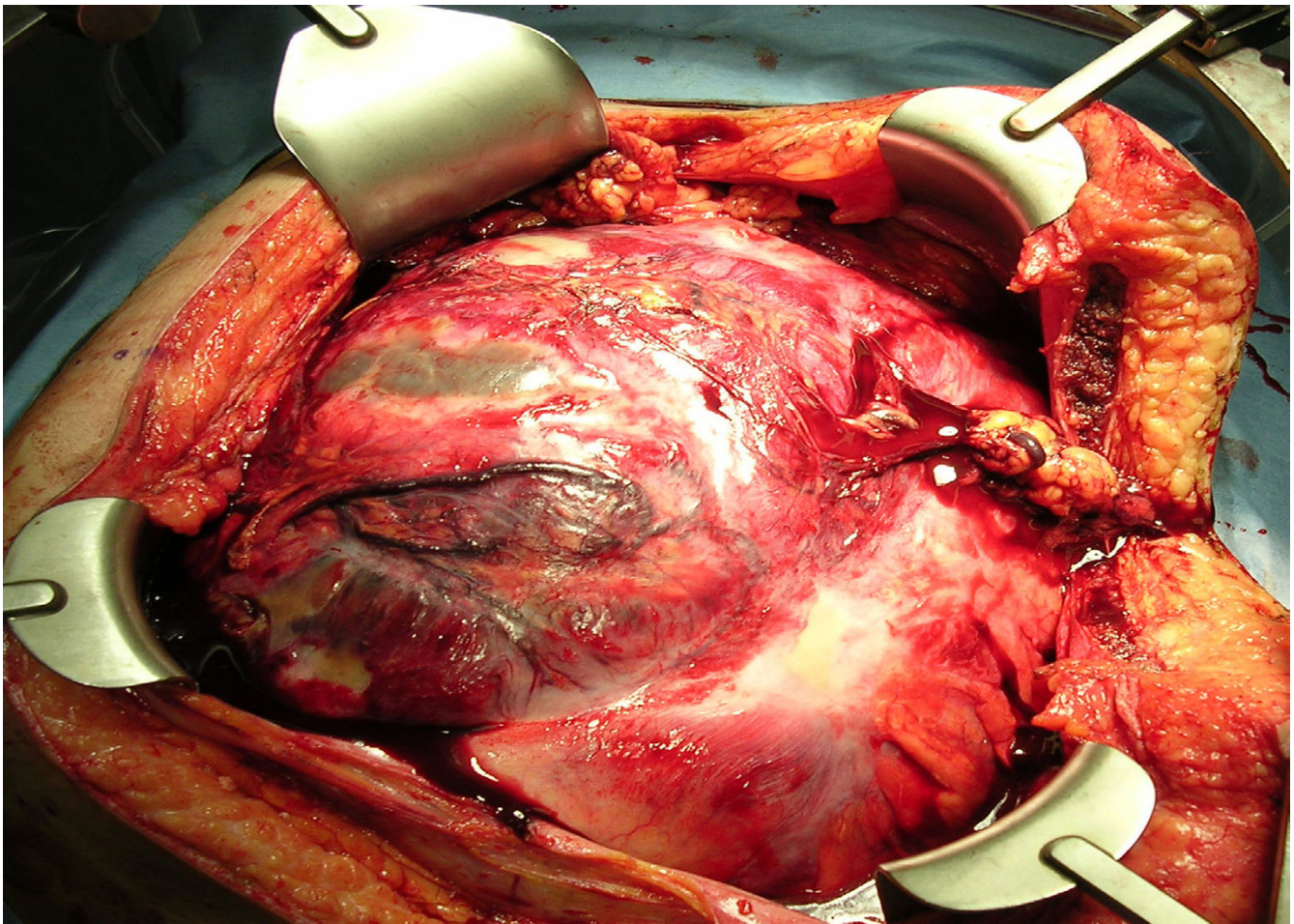
**Figure 1**

CT appearance at multiple cuts (A – E) of a huge retroperitoneal mass with lipomatous and nonlipomatous components. The nonlipomatous component (arrow X, Panel D) contains calcific elements. Note the posterior extension of the lipomatous component (arrow Y, Panel D), which extends superiorly (Panels A – C). The exact boundaries of this component are difficult to appreciate on CT.

also a heterogeneous nonlipomatous component that contained areas of lesser density, as well as a central stellate region of calcifications. The preoperative differential diagnosis included a retroperitoneal sarcoma (especially dedifferentiated liposarcoma), desmoid tumor, undifferentiated carcinoma, carcinoid or sclerosing mesenteritis; lymphoma was also considered.

While neoadjuvant chemotherapy and radiation comprise a frequent approach for retroperitoneal sarcomas at our institution, the extent of the tumor made this approach unfeasible. Resection was therefore planned unless an

intraoperative biopsy revealed lymphoma. Resection was accomplished through a T-type incision (Figure 2), and entailed removal of the right kidney, terminal ileum, ascending colon, sigmoid colon and the left spermatic cord structures, all of which were intimately attached to the mass. Encasement of the external iliac artery and vein was also encountered near the end of the procedure. This was not fully appreciated preoperatively, as that component of the tumor was so much less conspicuous on CT than the rest of the tumor, given its fatty consistency (Figure 1E). The mass was split in half to facilitate dissection from the iliac vessels. An anastomosis was constructed



**Figure 2**  
Operative exposure of the tumor through a T-type abdominal incision.

from descending colon to rectum. A transverse colon mucous fistula and an ileostomy were brought out, as the ileum was dusky at the end of the procedure.

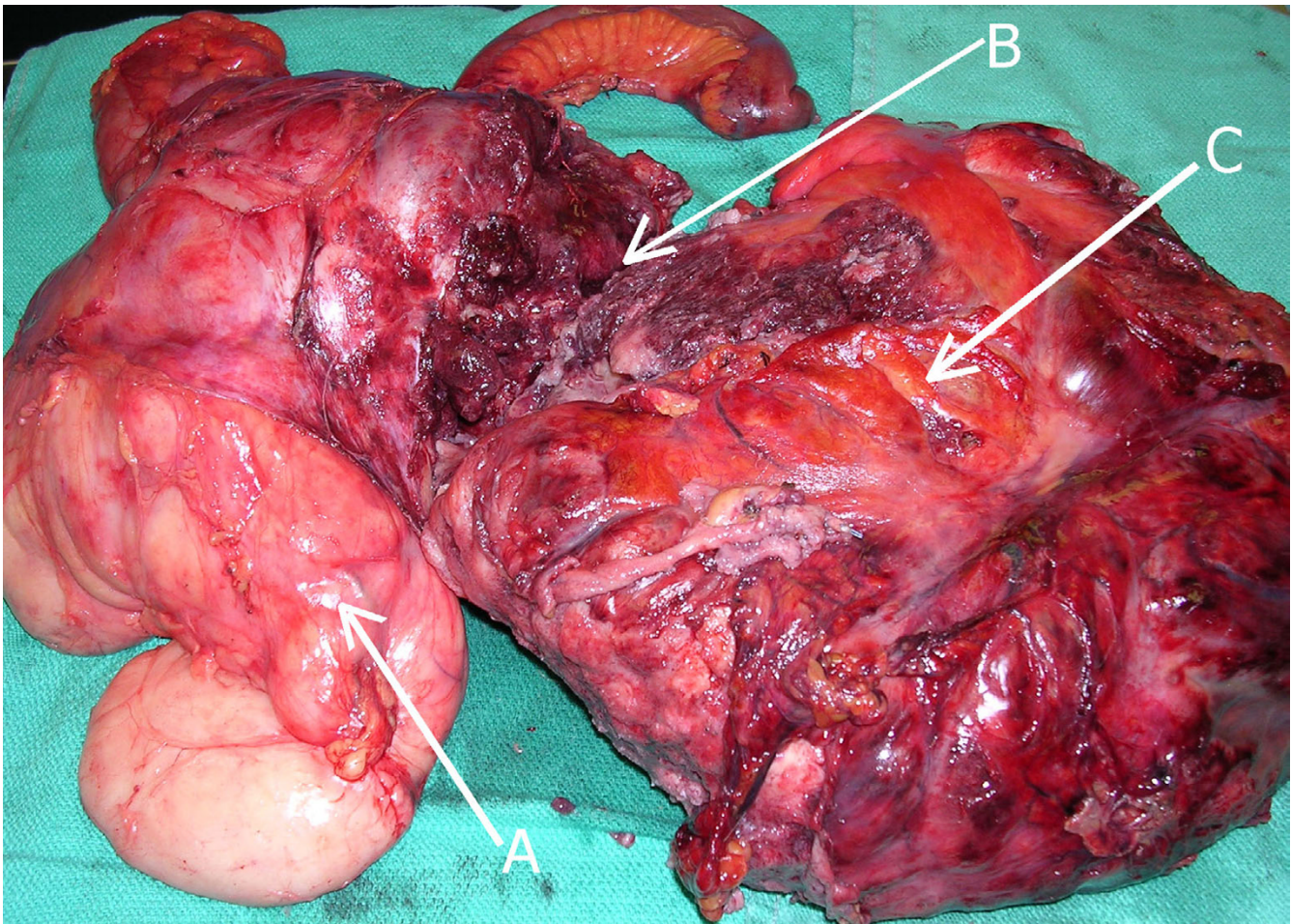
The tumor was submitted for histological examination in two parts measuring,  $30.0 \times 10.0 \times 6.5$  cm and  $32.0 \times 20.0 \times 10.0$  cm, weighing 19.8 kg in total (Figure 3). On gross examination the mass was variegated, with fleshy and solid cystic degeneration containing areas of osseous consistency. Microscopic examination revealed a juxtaposition of well differentiated liposarcoma and a spindle cell sarcoma with heterologous chondrosarcomatous elements consistent with a dedifferentiated liposarcoma (Figure 4).

The patient was discharged from hospital on the seventh postoperative day. He was followed in the surgical oncology outpatient clinic monthly. Four months after resec-

tion the patient had a follow-up CT scan, which demonstrated an intra-abdominal recurrence consisting almost completely of a calcified, nonlipomatous tumor (Figure 5). The patient died one month later.

### Discussion

The patient in our case manifested the dedifferentiated variant of liposarcoma. The term "dedifferentiated liposarcoma" refers to the development of a high grade nonlipogenic sarcoma juxtaposed to a well differentiated liposarcoma [3]. The majority (80 – 90%) occur primarily *de novo*, although secondary dedifferentiation can occur with multiple recurrences of a well differentiated liposarcoma [4]. CT and Magnetic resonance imaging scans typically reveal well defined nonlipomatous masses associated with fatty tumor; the transition between the two components is characteristically abrupt, although blended transitions are seen in about 20% of cases [5].

**Figure 3**

*En bloc* resection specimen of heterogeneous tumor with attached organs. Note the lipomatous regions (A), the calcified areas (B), and the remaining nonlipomatous component (C).

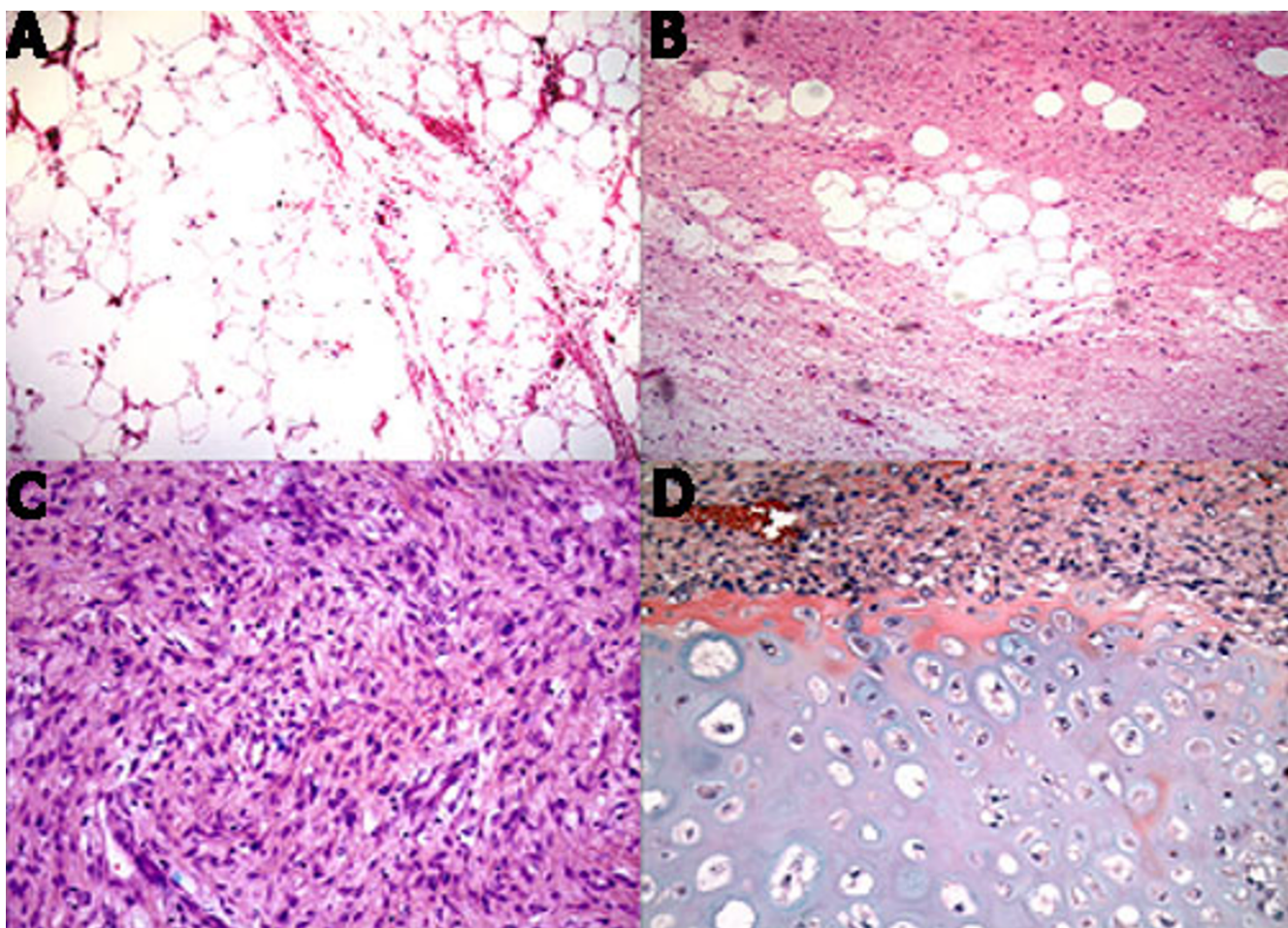
Calcifications appear in about 30% and usually correspond to osseous metaplasia, although they may represent osteosarcomatous or chondrosarcomatous elements. The most frequent phenotype of dedifferentiation is that of a high grade pleomorphic malignant fibrous histiocytoma-like sarcoma [4,6]. Other phenotypes observed include leiomyosarcomatous, rhabdomyosarcomatous, osteosarcomatous and angiosarcomatous elements, as well as other nonlipogenic elements [3,7]. A further distinctive pattern in some cases is the presence of micronodular spindle cell whorls, often associated with ossification [8].

Among liposarcomas, the presence of features of the dedifferentiated variant strongly portends a worse prognosis. The overall 5-year survival of dedifferentiated liposarcomas is 20%; the 5-year survival of well differentiated liposarcomas is 83% [9]. Dedifferentiated liposarcomas

recur locally in 40 – 83% and distant metastases appear in 15 – 30% [4,7,9]. Therefore, histomorphologic features impact outcomes related to retroperitoneal liposarcomas.

While generally the phenotype of the nonlipogenic component does not impact prognosis of dedifferentiated liposarcomas, the presence of calcifications has been identified as an adverse prognostic factor [5]. In the present situation, it is obvious that the biologically most aggressive component consisted of the calcified (chondrosarcomatous) component. That is, the recurrence was less heterogeneous than the primary tumor, as it had widespread and dense calcifications, but no obvious lipomatous elements.

Complete resection of the tumor is perhaps the most important factor determining long-term survival. Unfortunately, the rate of complete respectability is only about



**Figure 4**  
 Histologic appearance of various elements of the tumor as sampled in different regions. A. Well differentiated liposarcoma. B. Low grade spindle cell component. C. Cellular spindle cell component. D. Chondrosarcomatous component.

53% [10]. As illustrated in the present case, in addition to the limitations imposed by the retroperitoneal anatomy, another obstacle to successfully obtaining margins is the difficulty in distinguishing normal retroperitoneal fat from the lipogenic component of the tumor [9]. This was illustrated by the underestimation of the extent of the tumor around the iliac vessels. Moreover, the intraoperative decision to remove the kidney was made in view of the difficulty in distinguishing normal perinephric fat and neoplastic fat; kidney was not involved with tumor, once examined microscopically. Indeed, in a series of retroperitoneal liposarcomas from Memorial Sloan-Kettering Cancer Center, nephrectomy was performed in 38% of patients, although the number in which kidney was actually involved on pathology was not reported [9]. Thus, anatomical constraints and difficulty distinguishing more differentiated fatty tumor from normal fat limit the

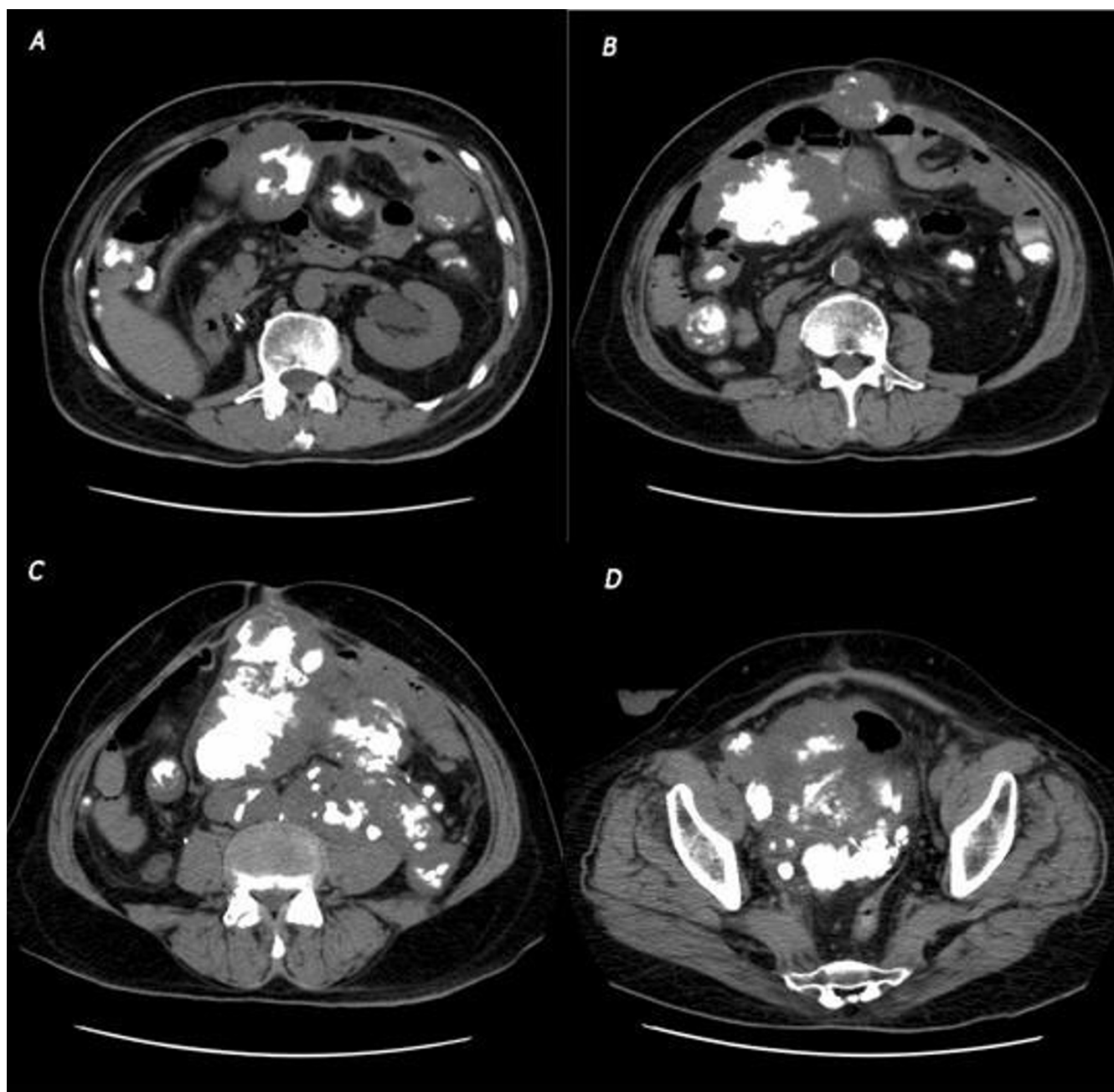
surgeon's ability to confidently and completely remove all neoplastic elements.

### Conclusions

Dedifferentiated liposarcomas represent aggressive variants of liposarcomas. Each morphological element of these heterogeneous tumors may manifest completely different biology. The overall biological behavior of dedifferentiated liposarcomas is likely dictated by the most aggressive element, which typically resides in the non-lipomatous portion of the tumor.

### Competing Interests

None declared.



**Figure 5**  
 CT appearance at multiple cuts (A – D) of the recurrent tumor. The recurrence was more homogeneous than the primary tumor, consisting almost completely of the calcific component of the nonlipomatous portion of the tumor.

**Authors Contributions**

SK, HB, WT, and OB made substantial contributions to the intellectual content of the paper, in the interpretation of data, and in drafting the manuscript. All authors read and approved the final manuscript.

**Acknowledgements**

Written consent was obtained from the patient's relatives for publication of this study.

**References**

1. Brady MS, Brennan MF: **Soft tissue sarcoma.** In: *Surgical Oncology* Edited by: allen-Mersh TG. Longon: Chapman and Hall; 1996:401-20.

2. Fletcher CD, Akerman M, Dal Cin P, de Wever I, Mandahl N, Mertens F, Mitelman F, Rosai J, Rydholm A, Sciort R, Tallini G, van den Berghe H, van de Ven W, Vanni R, Willen H: **Correlation between clinicopathological features and karyotypes in lipomatous tumors.** *Am J Pathol* 1996, **148**:623-630.
3. Nascimento AG: **Dedifferentiated liposarcoma.** *Semin Diagn Pathol* 2001, **18**:263-266.
4. Henricks WH, Chu YC, Goldblum JR, Weiss SW: **Dedifferentiated liposarcoma: A clinicopathological analysis of 155 cases with a proposal for an expanded definition of dedifferentiation.** *Am J Surg Pathol* 1997, **21**:271-281.
5. Tateishi U, Hasegawa T, Beppu Y, Satake M, Moriyama N: **Primary dedifferentiated liposarcoma of the retroperitoneum. Prognostic significance of computed tomography and magnetic resonance imaging features.** *J Comput Assist Tomogr* 2003, **27**:799-804.
6. Coindre JM, Mariani O, Chibon F, Mairal A, De Saint Aubain Somershausen N, Favre-Guillevin E, Bui NB, Stoeckle E, Hostein I, Aurias A: **Most malignant fibrous histiocytomas developed in the retroperitoneum are dedifferentiated liposarcomas: A review of 25 cases initially diagnosed as malignant fibrous histiocytoma.** *Mod Pathol* 2003, **16**:256-262.
7. McCormick D, Mentzel T, Beham A, Fletcher CD: **Dedifferentiated liposarcoma: clinicopathologic analysis of 32 cases suggesting a better prognostic subgroup among the pleomorphic sarcomas.** *Am J Surg Pathol* 1994, **18**:1213-1223.
8. Nascimento AG, Kurtin PJ, Guillou L, Fletcher CD: **Dedifferentiated liposarcoma: a report of nine cases showing a peculiar neural-like (whirling) pattern associated with metaplastic bone formation.** *Am J Surg Pathol* 1998, **22**:945-955.
9. Singer S, Antonescu CR, Riedel E, Brennan MF: **Histologic subtype and margin of resection predict pattern of recurrence and survival for retroperitoneal liposarcoma.** *Ann Surg* 2003, **238**:358-371.
10. Bevilacqua RG, Rogatko A, Hajdu SI, Brennan MF: **Prognostic factors in primary retroperitoneal soft tissue sarcomas.** *Arch Surg* 1991, **126**:328-334.

Publish with **BioMed Central** and every scientist can read your work free of charge

"BioMed Central will be the most significant development for disseminating the results of biomedical research in our lifetime."

Sir Paul Nurse, Cancer Research UK

Your research papers will be:

- available free of charge to the entire biomedical community
- peer reviewed and published immediately upon acceptance
- cited in PubMed and archived on PubMed Central
- yours — you keep the copyright

Submit your manuscript here:  
[http://www.biomedcentral.com/info/publishing\\_adv.asp](http://www.biomedcentral.com/info/publishing_adv.asp)

