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A retroperitoneal primary undifferentiated pleomorphic sarcoma

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ABSTRACT

A 52-year-old male had pain in the right back and right hypochondrium, and an abdominal CT scan revealed a 49-mm tumor in the right upper perirenal space. Additional MRI and PET-CT suggested that the tumor may be a primary adrenal carcinoma and could invade the liver and diaphragmatic leg. The tumor was completely removed by laparotomy and histopathologically diagnosed as retroperitoneal primary undifferentiated pleomorphic sarcoma. The patient has remained recurrence-free for 1.5 years after the surgery.

1. Introduction

Undifferentiated pleomorphic sarcoma (UPS), a type of soft-tissue sarcoma, predominantly affects the extremities and trunk. However, it is relatively rare in the retroperitoneum. Differentiation using imaging findings alone is often difficult; thus, histological diagnosis based on surgical results is often required. Here, we describe a case of retroperitoneal primary UPS in a 52-year-old male with pain in the right side of the back and right hypochondrium.

2. Case presentation

A 52-year-old male presented with pain in the right back and right hypochondrium and underwent abdominal CT scan, which revealed a 49-mm tumor in the fat tissue in the upper right perirenal space (Fig. 1a). Contrast-enhanced CT suspected a large right adrenal tumor with a predominantly marginal contrast effect (Fig. 1b). MRI showed a heterogeneous signal in the enlarged right adrenal gland on T2-weighted images and a high signal with marginal predominance on diffusion-weighted images, suggesting that the tumor had invaded the liver and diaphragmatic leg (Fig. 1c and d). PET-CT revealed fluorine-18-deoxyglucose accumulation in the right adrenal gland (Fig. 2). Adrenal hormone levels were within the normal range. Accordingly, we diagnosed the tumor as a non-functioning right adrenal carcinoma (cT4N0M0).

An open right adrenalectomy through a chevron incision was performed. The tumor had invaded the liver and diaphragmatic leg; therefore, we conducted resection with sufficient margins to include a segment of each organ. A tumor embolus was found in the adrenal vein. The total operative time was 525 min, and the blood loss was 1350 ml. Histopathologically, the irregularly shaped nuclei of unequal size showed irregular convoluted arrangements and bundle-like growth infiltrating the surrounding liver, adipose tissue, and diaphragmatic crura (Fig. 3). Cellular atypia, including nuclear fission images, multinucleated cells, and cells with indistinguishable differentiation tendencies, were conspicuous, and various immunohistochemical stains were performed, all of which were negative for CKAE1/AE3, Caldesmon, desmin, SMA, CD34, ERG, EMA, S100, MDM2, CDK4, SOX10, Chromogranin A, Synaptophysin, STAT6, and ALK. Based on the aforementioned findings, we diagnosed the tumor as a UPS and FNCLCC grade 3, which indicates the histological grade of soft-tissue sarcoma. Anatomically, we strongly suspected UPS of primary adrenal gland origin; however, since no adrenal gland could be histopathologically confirmed within the excised tumor, we made a diagnosis of UPS (pT2N0M0, Stage IIIA) with a retroperitoneal primary tumor. Complete tumor resection was achieved both clinically and pathologically, and we opted for a follow-up observation approach without additional therapy. At the current follow-up, 1.5 years after surgery, no recurrence has been detected.

3. Discussion

UPS, formerly called malignant fibrous histiocytoma, is a malignant soft-tissue tumor of unknown origin and was defined in the 2002 WHO

Abbreviations: UPS, Undifferentiated pleomorphic sarcoma; T2WI, T2-weighted images; DWI, diffusion-weighted images.

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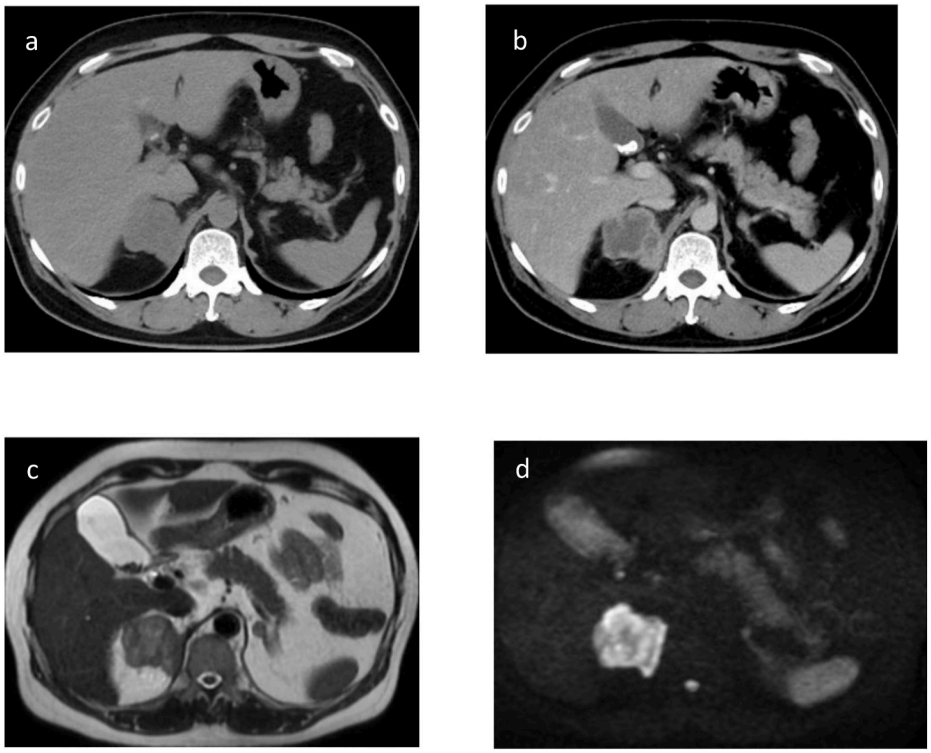

Fig. 1. Contrast-enhanced CT showing a right adrenal tumor with a predominant marginal contrast effect (a: non-contrast CT; b: contrast-enhanced CT). MRI shows a heterogeneous signal in the enlarged right adrenal gland on T2-weighted images (T2WI) and high signal with marginal predominance on diffusion-weighted images (DWI). (c: T2WI, d: DWI).

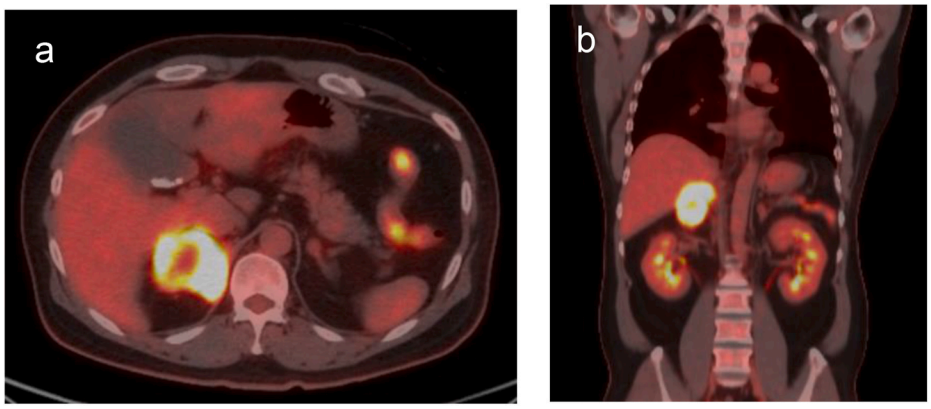

Fig. 2. PET-CT shows fluorine-18-deoxyglucose accumulation in the right adrenal gland (a: axial, b: coronal).

classification as "an undifferentiated sarcoma without obvious differentiation into histiocytes" and pathologically classified in 2013 as "undifferentiated/unclassified sarcomas." Currently, the 2020 classification is a generic term for "many unclassifiable malignant soft tissue tumors for which no specific genetic characteristics or propensity to differentiate can be demonstrated by genetic or histopathological methods."[1] The tumor accounts for 17.1 % of all soft tissue sarcomas and is the second most common after liposarcoma, with an incidence of 1–2 per 100,000 persons. The limbs and trunk are the predominant sites of occurrence, with retroperitoneal primary sites reported in 9 % of the cases. This disease is most common in adults in their 50s–70s, and has a poor prognosis, with 5- and 10-year survival rates of 60 % and 48 %, respectively.[2]

The pathological findings revealed cellular atypia, including nuclear fission, multinucleated cells, and undifferentiated cells. To make a definitive pathological diagnosis, multiple immunohistochemical stains are required to exclude other diseases[3]; however, since all of them were negative in this case, UPS was diagnosed. Anatomically, this case was suspected to be a UPS arising from the adrenal gland; however, grossly and pathologically, no evidence of adrenal tissue was observed within the tumor. Although we considered the possibility that all normal adrenal tissues had been replaced by UPS, we diagnosed the patient with primary retroperitoneal UPS. Few previous reports have described primary adrenal UPS. TNM classification of UPS depends on their site of origin. The T classification of primary retroperitoneal tumors was defined by tumor diameter: T1, 0–5 cm; T2, 5–10 cm; T3, 10–15 cm; and T4, >15 cm. Tumors with a diameter larger than 5 cm and smaller than 10 cm were classified as T2, and the pathological classification was determined by the TNM classification plus histological grade; therefore, in the present case, the tumor was Stage IIIA.

The first choice of treatment was complete surgical resection, which is crucial to ensure adequate resection margins. The 5-year survival rates

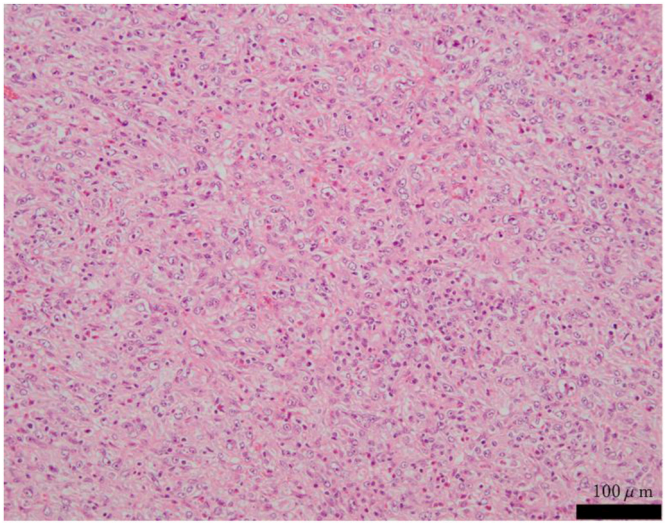

Fig. 3. Hematoxylin–eosin stain.

for patients with positive and negative margins were significantly different (33 % and 63 %, respectively).[4] Postoperative radiation therapy is an option in cases where sufficient resection margins cannot be achieved. However, in this case, we were able to achieve sufficient margins during tumor excision, and considering the potential difficulty of re-resection in the event of future recurrence, postoperative radiation therapy was not administered.

Recently, a nomogram predicting overall survival was reported, and the disease was calculated to have a 3-year survival rate of 61 % and 5-year survival rate of 48 %, suggesting that the prognosis is poor and that close follow-up is warranted.[5] In accordance with the NCCN guideline's follow-up plan for Stage II–IV patients,[6] follow-up thoracoabdominal CT has been performed every 3 months for 2 years after surgery, and no recurrence has been observed until now, 1.5 years after surgery. We plan to follow-up with CT scans every 6 months for the first 3–4 years postoperatively and annually from the fifth year postoperatively.

4. Conclusion

Herein, we report a case of UPS arising in the retroperitoneum. Complete surgical resection was performed, and no recurrence or metastasis was observed 1.5 years postoperatively.

Consent for publication

Informed consent was obtained from the patient for publication of the case details.

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Declarations of interest

None.

CRediT authorship contribution statement

Makishi Nakamura: Writing – original draft, Visualization, Investigation, Formal analysis, Data curation. **Kazuaki Yamanaka:** Writing – review & editing, Writing – original draft, Validation, Supervision, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Taigo Kato:** Writing – review & editing. **Koji Hatano:** Writing – review & editing. **Atsunari Kawashima:** Writing – review & editing. **Shinichiro Fukuhara:** Writing – review & editing. **Norio Nonomura:** Writing – review & editing. **Yoichi Kakuta:** Writing – review & editing.

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