Case Report

Journal of INTERNATIONAL MEDICAL RESEARCH

Journal of International Medical Research
48(11) 1–11

DOI: 10.1177/0300060520969473
journals.sagepub.com/home/imr
SAGE

Sarcomatoid intrahepatic cholangiocarcinoma in a patient with poor prognosis: a case report and literature review

Xu Li[1,*], Jian Li[1,*], Kai Liu[2], Ludong Tan[2] and Yahui Liu[2]

Abstract

Sarcomatoid intrahepatic cholangiocarcinoma (S-iCCA) is a rare histological variant of intrahepatic cholangiocarcinoma (iCCA). The diagnosis of S-iCCA is based on histopathological and immunohistochemical examinations, and S-iCCA often has a poorer prognosis than that of ordinary iCCA. In this article, we present the case of a 64-year-old man with S-iCCA who presented with intermittent right upper abdominal pain. The aim of this case report and literature review is to strengthen the understanding of S-iCCA among clinicians and reduce the incidence of missed clinical diagnoses.

Keywords

Sarcomatoid change, sarcomatoid intrahepatic cholangiocarcinoma, poor prognosis, case report, hepatobiliary malignancy, abdominal pain

Date received: 17 August 2020; accepted: 28 September 2020

Introduction

Epithelial tumors with sarcomatoid changes are rare neoplasms that have been reported in various sites, including the upper digestive tract, lung, pancreas, skin, breast, thyroid, uterus, urinary tract, and gallbladder.[1,2] Sarcomatoid intrahepatic cholangiocarcinoma (S-iCCA) is defined by the World Health Organization as

[1]Department of Hepatology, The First Hospital of Jilin University, Changchun, China, Jilin Province, China
[2]Department II of Hepatobiliary and Pancreatic Surgery, The First Hospital of Jilin University, Changchun, China, Jilin Province, China

*The authors contributed equally to this work.

Corresponding author:
Yahui Liu, Department II of Hepatobiliary and Pancreatic Surgery, the First Hospital of Jilin University, No. 71, Xinmin Street, Changchun, 130021, China.
Email: liuyahui2008@yeah.net

intrahepatic cholangiocarcinoma (iCCA) with sarcomatoid changes.[3] The mechanism of its pathogenesis is still unknown. It has been reported that sarcomatoid hepatocellular carcinoma (HCC) may be associated with preoperative anticancer treatments, such as transcatheter arterial chemoembolization, radiofrequency ablation, or percutaneous ethanol injection.[4–6] However, there is no relevant study demonstrating the association between anticancer treatments and S-iCCA. Confirmation by biopsy and immunohistochemical staining is critical to establish a definitive diagnosis of S-iCCA. Histological and immunohistochemical examinations of S-iCCA typically show a malignant neoplasm with both carcinomatous and sarcomatous components and positive expression of epithelial and mesenchymal molecular markers.[7] At present, the main treatment option is surgical resection; however, patients still have a relatively poor prognosis. There are a few reports on S-iCCA worldwide, and most of them are individual cases. This case report and review of relevant literature provide useful information to improve the awareness of S-iCCA among clinicians, reduce missed diagnoses, and achieve an accurate diagnosis and treatment.

Case report

The patient was a 64-year-old Chinese man admitted to our hospital for intermittent right upper abdominal pain. Physical examination showed tenderness and rebound pain in the right upper abdomen. Serological examinations revealed a slightly elevated gamma-glutamyl transpeptidase level at 119.3 U/L (normal range: 10–60 U/L), an elevated serum carbohydrate antigen 19–9 (CA19-9) level at 351.74 U/mL (normal range: 0–37.0 U/mL), and an elevated 24–2 level at 86.55 U/mL (normal range: 0–20.0 U/mL). His serum carcinoembryonic antigen and alpha-fetoprotein levels were within normal limits. A computed tomography scan of the entire abdomen plus three-stage enhancement (Figure 1) showed multiple calcifications in the left inner lobe of the liver and multiple calculi of intrahepatic and extrahepatic bile ducts, followed by dilatation of intrahepatic and extrahepatic bile ducts (part of the left outer lobe of the liver had expanded to the thickness of the bile ducts, and the contrast enhancement was obvious).

The preoperative diagnoses were space-occupying lesions in the left lateral lobe of the liver and extrahepatic and intrahepatic cholangiolithiasis. No related lymph node or distant metastasis was found. Hepatic left lateral lobectomy, cholecystectomy, exploration and lithotomy of the biliary tract, and T tube drainage were performed. Histological examination (Figure 2) of the liver tissue demonstrated the presence of an S-iCCA ($2.0 \times 1.8 \times 1.7$ cm) confined to the hepatic capsule; no evidence of cancer infiltration was found at the margin of hepatectomy, in vessels, or in nerves. A separate inspection (embolus) revealed the presence of cancer tissue. The tumor stage was determined to be T2aN0M0 based on the 7th edition of the American Joint Committee on Cancer TNM staging system. Immunohistochemical examination of the neoplasm showed positive staining for cytokeratin (CK)8, CK-pan, and vimentin and negative staining for CK7, CK20, and hepatocyte paraffin 1 (HepPar-1). The Ki-67 proliferation index was approximately 60%. Based on these histopathological and immunohistochemical findings, a definitive diagnosis of S-iCCA was determined. The overall follow-up duration was 3 months, and the patient died 3 months after surgery.

Discussion

S-iCCA is a rare histological subtype of iCCA and has only been reported in the

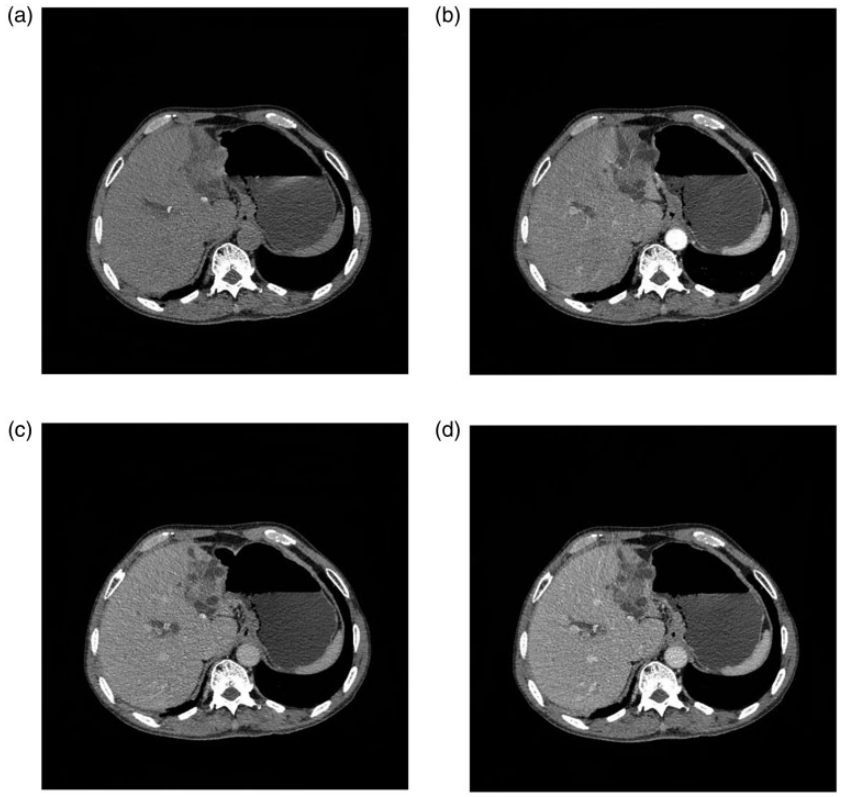

Figure 1. Axial computed tomography findings in the described patient with sarcomatoid intrahepatic cholangiocarcinoma. (a) and (b) Arterial phase reveals part of the left outer lobe of the liver that has expanded to the thickness of the bile ducts, and strengthening is obvious. (c) and (d) The lesion shows no contrast enhancement in venous or late phase reveals.

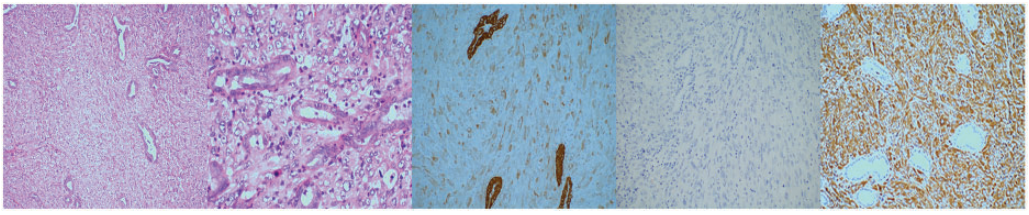

Figure 2. Histologic findings of liver biopsy in the described patient with sarcomatoid intrahepatic cholangiocarcinoma. (a) and (b) Hematoxylin and eosin staining, 10× and 40×, respectively. Immunohistochemistry (20×) for CK-pan (c), HepPar1 (d), and vimentin (e).
CK-pan, pan-cytokeratin; HepPar1: hepatocyte paraffin 1.

English literature in case reports or series. Its prevalence is unknown, but it is reported to account for 4.5% of cholangiocarcinoma cases[8] and less than 1% of hepatobiliary system malignancies.[9] As for the pathogenesis, researchers have proposed that anticancer treatments might lead to the development of sarcomatoid changes or

accelerate the transformation of epithelial-derived cells into sarcoma cells.[10] However, there are no reports regarding the relationship between S-iCCA and anticancer therapy.[8]

Abdominal pain is the most common clinical symptom of S-iCCA. According to previous studies,[9,11,12] serum CA19-9 and carcinoembryonic antigen (CEA) may not be sensitive enough for the diagnosis of S-iCCA. Of note, a low echogenic liver mass on ultrasound showing hypoattenuation and peripheral region enhancement after contrast injection on a computed tomography scan[9] are common characteristics of S-iCCA, similar to ordinary iCCA.[7] Therefore, it may be difficult to distinguish S-iCCA from ordinary iCCA using radiological imaging. A definitive diagnosis of S-iCCA can be determined by biopsy.

To review the known characteristics of S-iCCA, we searched PubMed using the keywords "liver", "sarcomatous", "sarcomatoid", and "cholangiocarcinoma". The clinical features of 46 cases are summarized in Table 1 (including the present case). Among these patients, 32 (69.6%) were male, 14 (30.4%) were female, and the mean age was 62 (range: 37–87) years. The mean tumor size was 8.0 (range: 2.0–22.0) cm. The tumor location was recorded in 21 patients, most frequently in the left lobe of the liver (13 patients; 61.9%), followed by the right lobe (7 patients; 33.3%) and the hepatic hilum (1 patient; 4.8%). Overall, 36 (78.3%) patients had one tumor, and 10 (21.7%) patients had multiple tumors. The initial radiologic impressions were recorded in 25 patients, with 18 (72.0%) characterized as HCC, 1 (4.0%) as lymphoma, 3 (12.0%) as hepatic abscesses, and 3 (12.0%) as hepatic space-occupying lesions. The laboratory findings are shown in Table 2. CA19-9 levels were elevated in 15 patients and normal in 20 patients; CEA was elevated in 3 patients and normal in 19 patients. Detailed immunohistochemistry results were available for 33 patients, which are shown in Table 3. Overall, tumors were positive for CK in 30 (90.9%) patients and positive for vimentin in 27 (81.8%) patients, which was considerably helpful to arrive at the final diagnosis of S-iCCA. Among the 46 S-iCCA patients who had treatment information available (Table 4), 22 (47.8%) underwent surgical treatment, 13 (28.3%) underwent chemotherapy or radiotherapy, 2 (4.3%) underwent transcatheter arterial embolization, and 9 (19.6%) remained untreated.

The diagnosis of the tumor is established by histopathological and immunohistochemical examinations. S-iCCA does have some unique histopathological and molecular cytogenetic patterns. Histopathological analyses of S-iCCA show the coexistence of adenocarcinoma cells with differentiated and sarcomatoid cells, which are spindle-shaped and arranged in bundles or weaves. Immunohistochemistry reveals that S-iCCA tumors are positive for both epithelial cholangiogenic tumor markers (CK7, CK8) and the mesenchymal tumor marker vimentin and negative for HepPar-1.[4,8,11,12] As a marker of hepatocytes, HepPar-1 provides useful diagnostic information in distinguishing HCC from cholangiocarcinoma and metastatic carcinoma in the liver. In addition, our patient (described above) was positive for CK-pan, CK-8, and vimentin and negative for CK20 and HepPar-1, which are consistent with our diagnosis.

At present, there are no relevant guidelines for determining the prognosis and survival of patients with S-iCCA, and radical liver resection is the only available treatment. In published case reports, the median survival of patients with S-iCCA who underwent surgical resection was 11 months, which is comparable with that in patients with ordinary iCCA who did not undergo surgery (8 months).[4] The median survival of patients with S-iCCA who did

Table 1. Clinical characteristics of S-iCCA reported in the English-language literature.

	Case no.	Age (y)	Sex	Location	Tumor size (cm)	Number of tumors	Initial radiologic impression
Kim et al[9]	1	45	M	N/A	7.5	Multiple	HCC
	2	67	M	N/A	2.5	Single	HCC
	3	55	M	N/A	6.5	Multiple	IHCC
	4	66	M	N/A	10	Single	Hepatic abscess
	5	56	M	N/A	8	Single	HCC
	6	66	F	N/A	7.5	Single	IHCC
	7	68	M	N/A	6	Single	HCC
	8	55	F	N/A	8.5	Multiple	IHCC
	9	49	M	N/A	9.5	Multiple	Lymphoma
	10	65	M	N/A	9.5	Multiple	IHCC
	11	61	M	N/A	5	Single	IHCC
Sintra et al[18]	12	N/A	M	Right	10	Single	Hepatic carcinoma
Sasaki et al[19]	13	79	M	Left	8	Multiple	Hepatic mass
Haratake et al[20]	14	59	M	Right	Fist-sized	Multiple	Hepatic abscess
Nakajima et al[21]	15	84	F	Hepatic hilum	3.5	Single	N/A
	16	43	F	Right	14	Single	N/A
	17	73	F	Left	7	Single	N/A
	18	37	M	Left	10	Single	N/A
	19	64	M	Left	7.5	Single	N/A
	20	52	M	Right	7.5	Single	N/A
	21	69	M	Left	10	Single	N/A
Imazu et al[22]	22	77	M	Left	6	Single	Cholangiocarcinoma
Honda et al[23]	23	61	F	N/A	N/A	Multiple	IHCC
Itamoto et al[24]	24	70	M	Right	8	Single	HCC
Matsuo et al[25]	25	77	F	Left	7.7	Single	Hepatic abscess
Kaibori et al[1]	26	69	F	Left	22	Single	Hepatic carcinoma
Lim et al[26]	27	41	F	Left	17	Single	Hepatic mass
Sato et al[8]	28	87	M	Left	4	Single	IHCC
Malhotra et al[2]	29	60	F	Right	20	Single	Hepatic carcinoma
Bilgin et al[27]	30	48	M	Left	13	Single	Hepatic carcinoma
Watanabe et al[12]	31	62	M	Right	5	Multiple	IHCC
Gu et al[13]	32	65	M	N/A	N/A	Single	N/A
	33	70	M	N/A	N/A	Single	N/A
	34	48	F	N/A	N/A	Single	N/A
	35	45	M	N/A	N/A	Single	N/A
	36	46	F	N/A	N/A	Single	N/A
	37	69	M	N/A	N/A	Single	N/A
	38	54	F	N/A	N/A	Single	N/A
	39	74	M	N/A	N/A	Single	N/A
	40	57	M	N/A	N/A	Single	N/A
	41	51	M	N/A	N/A	Single	N/A
	42	69	M	N/A	N/A	Single	N/A
	43	61	F	N/A	N/A	Single	N/A
	44	53	M	N/A	N/A	Single	N/A
Ning et al[4]	45	63	M	Left	8	Multiple	Cholangiocarcinoma
Our case	46	64	M	Left	2	Single	Hepatic mass

N/A: not available; S-iCCA: sarcomatoid intrahepatic cholangiocarcinoma; HCC: hepatocellular carcinoma; IHCC: intrahepatic cholangiocarcinoma; F: female; M: male.

Table 2. Laboratory findings reported in the English language literature.

Case no.	AST (U/L)	ALT (U/L)	GGT (U/L)	CEA (ng/mL)	CA19-9 (U/mL)
1	25	19	115	0.74	>1200.00
2	31	10	32	1.45	3.38
3	54	96	137	0.1	3
4	42	30	N/A	2.35	1809.57
5	43	57	203	1.81	2.33
6	23	39	253	12.7	710.38
7	23	16	224	1.18	12.59
8	30	31	323	3.15	>1200.00
9	80	30	98	1.08	<2.00
10	37	47	N/A	3.56	599.14
11	34	36	35	1.81	5.77
12	20	15	N/A	N/A	normal
13	34	N/A	77	normal	normal
14	75	46	356	N/A	N/A
15	N/A	N/A	N/A	N/A	N/A
16	N/A	N/A	N/A	N/A	N/A
17	N/A	N/A	N/A	N/A	N/A
18	N/A	N/A	N/A	N/A	N/A
19	N/A	N/A	N/A	N/A	N/A
20	N/A	N/A	N/A	N/A	N/A
21	N/A	N/A	N/A	N/A	N/A
22	33	27	99	<0.5	17
23	22	30	175	9	13394
24	normal	348	normal	normal	2634
25	N/A	N/A	N/A	normal	normal
26	N/A	N/A	N/A	normal	3665
27	N/A	N/A	N/A	normal	normal
28	N/A	N/A	N/A	16.2	2894
29	N/A	N/A	N/A	N/A	N/A
30	152	45	297	N/A	39
31	174	356	405	1.4	1109.9
32	N/A	N/A	N/A	N/A	11.25
33	N/A	N/A	N/A	N/A	22.44
34	N/A	N/A	N/A	N/A	7.28
35	N/A	N/A	N/A	N/A	10384
36	N/A	N/A	N/A	N/A	N/A
37	N/A	N/A	N/A	N/A	25.81
38	N/A	N/A	N/A	N/A	11.34
39	N/A	N/A	N/A	N/A	6.07
40	N/A	N/A	N/A	N/A	2
41	N/A	N/A	N/A	N/A	11.71
42	N/A	N/A	N/A	N/A	N/A
43	N/A	N/A	N/A	N/A	886.51
44	N/A	N/A	N/A	N/A	10.55
45	25	19	125	2.17	100.5
46	normal	normal	119.3	normal	351.74

AST: aspartate aminotransferase; ALT: alanine aminotransferase; GGT: gamma-glutamyl transferase; CEA: carcinoembryonic antigen; CA19-9: carbohydrate antigen 19-9; N/A: not available.

Table 3. Immunohistochemistry of S-iCCA reported in the English language literature.

Case no.	Positive results	Negative results
1	CK19, vimentin	HSA, CD10
2	CK, vimentin, CEA, AFP	CK7, CK19, HSA, c-kit, CD117
3	CK, CK19, vimentin	CK8, desmin, EMA, CEA, c-kit, S-100
4	CK, CK8, CK19, vimentin, CEA, EMA	HSA, AFP, TTF-1
5	CK, CK8, CK19, vimentin, SMA	HSA, CD5, CD68, HMW-CK
6	CK7, CK8, CK19, vimentin, CEA	HSA
7	CK7, CK8, CK19, vimentin, CD34	HSA, CEA, HMW-CK
8	CK19, vimentin, CEA, p53	CD31, CD34
9	CK19, vimentin, CEA	CK7, desmin, HSA, SMA, c-kit, S-100
10	CK, CK19, vimentin, CEA	HSA, CD31
11	CK7, CK19, vimentin, MUC1	HSA, CD10
12	CK7, vimentin	CK20, HepPar1
13	KER, EMA, vimentin, CEA	AFP, S-100, AAT
14	low molecular cytokeratin, vimentin	UEA-1, desmin
15	KER, EMA, CA19-9	PAS, CEA, AFP, vimentin, actin, desmin, S-100, NSE
16	KER, EMA, vimentin	PAS, CEA, AFP, CA199, actin, desmin, S-100, NSE
17	/	PAS, CEA, AFP, CA199, actin, desmin, S-100, NSE, KER, EMA, vimentin
18	PAS, KER, EMA, vimentin	CEA, CA199, AFP, actin, desmin, S-100, NSE
19	KER, EMA	PAS, CEA, AFP, CA199, actin, desmin, S-100, NSE, vimentin
20	PAS, KER, EMA, CEA	vimentin, CA199, AFP, actin, desmin, S-100, NSE
21	/	PAS, CEA, AFP, CA199, actin, desmin, S-100, NSE, KER, EMA, vimentin
22	wide-spectrum keratin, vimentin, CEA	muscle actin, AAT, S-100, AFP
23	vimentin	S-100, desmin, AFP, albumin, myoglobin
24	KER, EMA, vimentin	AFP, CEA, CA199, actin, desmin, S-100
25	AAT, vimentin, F13a	desmin, EMA, CYT, SMA, CEA, AFP
26	vimentin, EMA, CK	S-100, CEA, AFP
27	CK-pan, vimentin, CEA	CK7, CK20, S-100, HMB-45, AMA, CD34, AFP, c-kit
28	CK19, vimentin, CD44s	β-catenin
29	EMA, AE1/AE3, CK7, CK19, CEA	HepPar-1
30	N/A	N/A
31	CK, vimentin	N/A
32	N/A	N/A
33	N/A	N/A
34	N/A	N/A
35	N/A	N/A
36	N/A	N/A
37	N/A	N/A
38	N/A	N/A
39	N/A	N/A

(continued)

Table 3. Continued.

Case no.	Positive results	Negative results
40	N/A	N/A
41	N/A	N/A
42	N/A	N/A
43	N/A	N/A
44	N/A	N/A
45	AE1/AE3, STAT6, SOX10, CD34, CK19, desmin, MUC1, vimentin, SMA, S-100	N/A
46	CK-pan, CK8, vimentin	CK7, CK20, HepPar-1

CA19-9: carbohydrate antigen 19-9; CEA: carcinoembryonic antigen; CD10: cluster of differentiation 10; NSE: neuron-specific enolase; AFP: a-fetoprotein; N/A: not available; PAS: periodic acid–Schiff; KER: keratin; EMA: epithelial membrane antigen; CK: cytokeratin; MUC1: mucin-1; HSA: human serum albumin; AAT: α-1-antitrypsin; SMA: smooth muscle actin; AMA: anti-mitochondria autoantibodies; UEA-1: ulex europaeus agglutinin-1; TTF-1: thyroid transcription factor-1; HMW-CK: high molecular weight cytokeratin; c-kit: receptor tyrosine kinase; CYT: cytochrome; HMB-45: human melanoma black 45; SOX-10: SRY-related HMG-BOX Gene 10; STAT-6: signal transducer and activator of transcription 6; F13a: factor XIIIa; AE1/AE3=CK-pan; HepPar1: hepatocyte paraffin 1.

Table 4. Prognosis of S-iCCA patients reported in the English language literature.

Case no.	Survival (months)	Treatment	Outcome
1	1.7	Chemotherapy	Died
2	4.9	Chemotherapy	Died
3	4.3	Chemotherapy	Died
4	0.7	Supportive	Died
5	2.4	Chemotherapy	Died
6	4.2	Chemotherapy	Died
7	0.6	Supportive	Died
8	1.0	Chemotherapy	Died
9	1.4	Chemotherapy	N/A
10	0.5	Supportive	Died
11	12.8	Viscum album	Alive
12	1.5	Operation	Died
13	N/A	None	N/A
14	1.0	None	Died
15	3.0	None	Died
16	4.5	Operation	Died
17	5.0	Chemotherapy	Died
18	2.5	None	Died
19	1.0	TAE	Died
20	2.0	TAE	Died
21	36.0	Operation	Alive
22	11.0	Operation	Alive
23	3.8	None	Died
24	9.0	Operation	Alive

(continued)

Table 4. Continued.

Case no.	Survival (months)	Treatment	Outcome
25	5.0	Operation	Died
26	3.0	Operation	Died
27	2.0	Operation	Alive
28	3.0	None	Died
29	29.0	Operation	Alive
30	12.0	Operation	Alive
31	11.0	Operation	Died
32	3.0	Chemotherapy/radiotherapy	Progression
33	3.0	Operation	Recurrence
34	35.0	Operation	Recurrence
35	5.0	Chemotherapy/radiotherapy	Progression
36	2.0	Chemotherapy/radiotherapy	Progression
37	1.0	Operation	Recurrence
38	26.0	Operation	Recurrence
39	12.0	Operation	Recurrence
40	2.0	Chemotherapy/radiotherapy	Progression
41	3.0	Operation	Recurrence
42	2.0	Chemotherapy/radiotherapy	Progression
43	4.0	Operation	Recurrence
44	3.0	Operation	Recurrence
45	1.0	Operation	Alive
46	3.0	Operation	Died

S-iCCA: sarcomatoid intrahepatic cholangiocarcinoma; TAE: transcatheter arterial embolization; N/A: not available.

not undergo surgery was 3 months, which is considerably shorter than that of patients with ordinary iCCA.[12,13] Several studies have shown that cisplatin, doxorubicin, cyclophosphamide, and taxol as adjuvant chemotherapy after surgery may prolong survival in patients with sarcomatoid carcinomas.[14–17] In the patient described here, we learned by telephone follow-up that tumor metastasis and recurrence precluded further chemotherapy, and the patient died 3 months after surgery. To help improve the prognosis of patients with S-iCCA, early detection and radical surgery with careful follow-up are necessary, and this case demonstrates the importance of closer follow-up in patients with these malignancies. Furthermore, more comprehensive treatment options need to be explored.

In summary, S-iCCA is a rare malignancy. Diagnosis is only possible using pathology and immunohistochemical analyses because clinicomorphological findings and serologic and radiologic examinations are not disease-specific. Because of its high invasiveness, the prognosis of S-iCCA is poor, and closer follow-up is critical.

Ethics statement

Ethics permission was not obtained because our paper is a case report. The patient provided consent to publish this paper.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

This study was supported by the Science and Technology Development Program of Jilin Province under Grant no 20190103079JH, the Youth Development Foundation of the First

Hospital of Jilin University under Grant no JDYY102019004, and the National Science and Technology Major Project under Grant no 2018ZX10723203.

ORCID iDs

Xu Li https://orcid.org/0000-0002-0929-1428
Jian Li https://orcid.org/0000-0003-1665-9443

References

1. Kaibori M, Kawaguchi Y, Yokoigawa N, et al. Intrahepatic sarcomatoid cholangiocarcinoma. *J Gastroenterol* 2003; 38: 1097–1101.
2. Malhotra S, Wood J, Mansy T, et al. Intrahepatic sarcomatoid cholangiocarcinoma. *J Oncol* 2010; 2010: 701476.
3. Nagtegaal ID, Odze RD, Klimstra D, et al. The 2019 WHO classification of tumours of the digestive system. *Histopathology* 2020; 76: 182–188.
4. Zhang N, Li Y, Zhao M, et al. Sarcomatous intrahepatic cholangiocarcinoma: Case report and literature review. *Medicine (Baltimore)* 2018; 97: e12549.
5. Tsou YK, Wu RC, Hung CF, et al. Intrahepatic sarcomatoid cholangiocarcinoma: clinical analysis of seven cases during a 15-year period. *Chang Gung Med J* 2008; 31: 599–605.
6. Kim HM, Kim H and Park YN. Sarcomatoid cholangiocarcinoma with osteoclast-like giant cells associated with hepatolithiasis: a case report. *Clin Mol Hepatol* 2015; 21: 309–313.
7. Bridgewater J, Galle PR, Khan SA, et al. Guidelines for the diagnosis and management of intrahepatic cholangiocarcinoma. *J Hepatol* 2014; 60: 1268–1289.
8. Sato K, Murai H, Ueda Y, et al. Intrahepatic sarcomatoid cholangiocarcinoma of round cell variant: a case report and immunohistochemical studies. *Virchows Arch* 2006; 449: 585–590.
9. Kim DK, Kim BR, Jeong JS, et al. Analysis of intrahepatic sarcomatoid cholangiocarcinoma: Experience from 11 cases within 17 years. *World J Gastroenterol* 2019; 25: 608–621.
10. Kojiro M, Sugihara S, Kakizoe S, et al. Hepatocellular carcinoma with sarcomatous change: a special reference to the relationship with anticancer therapy. *Cancer Chemother Pharmacol* 1989; 23: 4–8.
11. Wang T, Kong J, Yang X, et al. Clinical features of sarcomatoid change in patients with intrahepatic cholangiocarcinoma and prognosis after surgical liver resection: A Propensity Score Matching analysis. *J Surg Oncol* 2020; 121: 524–537.
12. Watanabe G, Uchinami H, Yoshioka M, et al. Prognosis analysis of sarcomatous intrahepatic cholangiocarcinoma from a review of the literature. *Int J Clin Oncol* 2014; 19: 490–496.
13. Okabayashi T, Shima Y, Iwata J, et al. Surgical outcomes for 131 cases of carcinosarcoma of the hepatobiliary tract. *J Gastroenterol* 2014; 49: 982–991.
14. Higa T, Oshiro K, Kinjyo T, et al. Sarcomatoid carcinoma of the bladder in a child: case report of a successful treatment including gemcitabine and cisplatin. *Urology* 2016; 97: 200–203.
15. Galaal K, Van Der Heijden E, Godfrey K, et al. Adjuvant radiotherapy and/or chemotherapy after surgery for uterine carcinosarcoma. *Cochrane Database Syst Rev* 2013; 2013: Cd006812.
16. Chaft JE, Sima CS, Ginsberg MS, et al. Clinical outcomes with perioperative chemotherapy in sarcomatoid carcinomas of the lung. *J Thor Oncol* 2012; 7: 1400–1405.
17. Gu KW, Kim YK, Min JH, et al. Imaging features of hepatic sarcomatous carcinoma on computed tomography and gadoxetic acid-enhanced magnetic resonance imaging. *Abdom Radiol (NY)* 2017; 42: 1424–1433.
18. Sintra S, Costa R, Filipe C, et al. Intrahepatic sarcomatoid cholangiocarcinoma. *BMJ Case Rep* 2018; 2018: bcr-2018-225017.
19. Sasaki M, Nakanuma Y, Nagai Y, et al. Intrahepatic cholangiocarcinoma with sarcomatous transformation: an autopsy case. *J Clin Gastroenterol* 1991; 13: 220–225.
20. Haratake J, Yamada H, Horie A, et al. Giant cell tumor-like cholangiocarcinoma associated with systemic cholelithiasis. *Cancer* 1992; 69: 2444–2448.

21. Nakajima T, Tajima Y, Sugano I, et al. Intrahepatic cholangiocarcinoma with sarcomatous change: clinicopathologic and immunohistochemical evaluation of seven cases. *Cancer* 1993; 72: 1872–1877.
22. Imazu H, Ochiai M and Funabiki T. Intrahepatic sarcomatous cholangiocarcinoma. *J Gastroenterol* 1995; 30: 677–682.
23. Honda M, Enjoji M, Sakai H, et al. Case report: intrahepatic cholangiocarcinoma with rhabdoid transformation. *J Gastroenterol Hepatol* 1996; 11: 771–774.
24. Itamoto T, Asahara T, Katayama K, et al. Double cancer—hepatocellular carcinoma and intrahepatic cholangiocarcinoma with a spindle-cell variant. *J Hepatobiliary Pancreat Surg* 1999; 6: 422–426.
25. Matsuo S, Shinozaki T, Yamaguchi S, et al. Intrahepatic cholangiocarcinoma with extensive sarcomatous changes: report of a case. *Surg Today* 1999; 29: 560–563.
26. Lim BJ, Kim KS, Lim JS, et al. Rhabdoid cholangiocarcinoma: a variant of cholangiocarcinoma with aggressive behavior. *Yonsei Med J* 2004; 45: 543–546.
27. Bilgin M, Toprak H, Bilgin SS, et al. CT and MRI findings of sarcomatoid cholangiocarcinoma. *Cancer Imaging* 2012; 12: 447–451.