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CASE REPORT

Intussusception of small intestine due to metastasis of large cell carcinoma of the lung with a rhabdoid phenotype

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ABSTRACT: Large cell carcinoma of the lung with a rhabdoid phenotype is a rare type of lung cancer, and does not commonly metastasize to the small intestine. Herein we describe a 63-yr-old Japanese male with ileus resulting from small intestinal metastasis from lung cancer. Tumour enlargement was rapid and could not be treated with chemotherapy.

KEYWORDS: Large cell carcinoma, lung cancer, rhabdoid phenotype, small intestinal intussusception

Pulmonary carcinoma is the most frequently diagnosed major cancer, although only a few cases of large cell carcinoma of the lung with rhabdoid phenotype (LCCRP) have been reported [1]. Metastases to the brain, bone, adrenal glands and liver are more common in patients with lung cancer, whereas small intestinal metastasis is rare [2]. To our knowledge, there are no previous case reports describing intussusception of the small intestine caused by small intestinal metastasis of LCCRP.

CASE REPORT

A 63-yr-old male was admitted to the emergency department with continuous abdominal pain. He reported a 2-week history of vomiting at various times after meals. The abdomen showed distension and bowel sounds were absent. The patient was a smoker.

A routine chest radiograph taken as part of the initial work up showed a mass in the right lung (fig. 1a). Computed tomography (CT) scans demonstrated a mass in the right lung (fig. 1b and c), intussusception of the small intestine (fig. 1d) and lymph node enlargement in the abdomen. Besides these findings, a significant abnormality was not detected.

During emergency surgery, the patient underwent segmental resection of the region of the small bowel showing intussusception along with the flexure of the small bowel. Furthermore, there were many small lesions in the small bowel that had not been detected on CT examination, indicating that the primary lesion was situated elsewhere. These additional lesions were not resected. We performed percutaneous needle biopsy of the lung lesion for diagnosis. On pathological examination of the resected small bowel tissue (fig. 2a) and the lung biopsy specimen (fig. 2b and c), the microscopic appearances of these specimens were identical. These lesions were characterised histologically by the presence of typical rhabdoid cells, which are large cells with abundant cytoplasm and eccentric nuclei with central macronucleoli. In addition, these cells show rounded eosinophilic cytoplasmic inclusions. Based on these clinical findings, we diagnosed the patient as having primary lung tumour with metastasis to the small bowel.

Following surgical treatment the symptoms transiently recovered. However, the lung tumour quickly enlarged and pleural effusion developed within 3 weeks. Furthermore, the abdominal mass also increased and became more palpable, while the abdominal lymph nodes increased in size from 38×43 mm to 71×71 mm. Chemotherapy was not administered due to the patients poor general condition and performance status. Moreover, radiation therapy was not applied to the abdominal lesions due to gastrointestinal bleeding. The patient did not receive any further treatment except pain control. He died 27 days after emergency surgery.

DISCUSSION

Lung cancer is the leading case of cancer death worldwide in both males and females. Common

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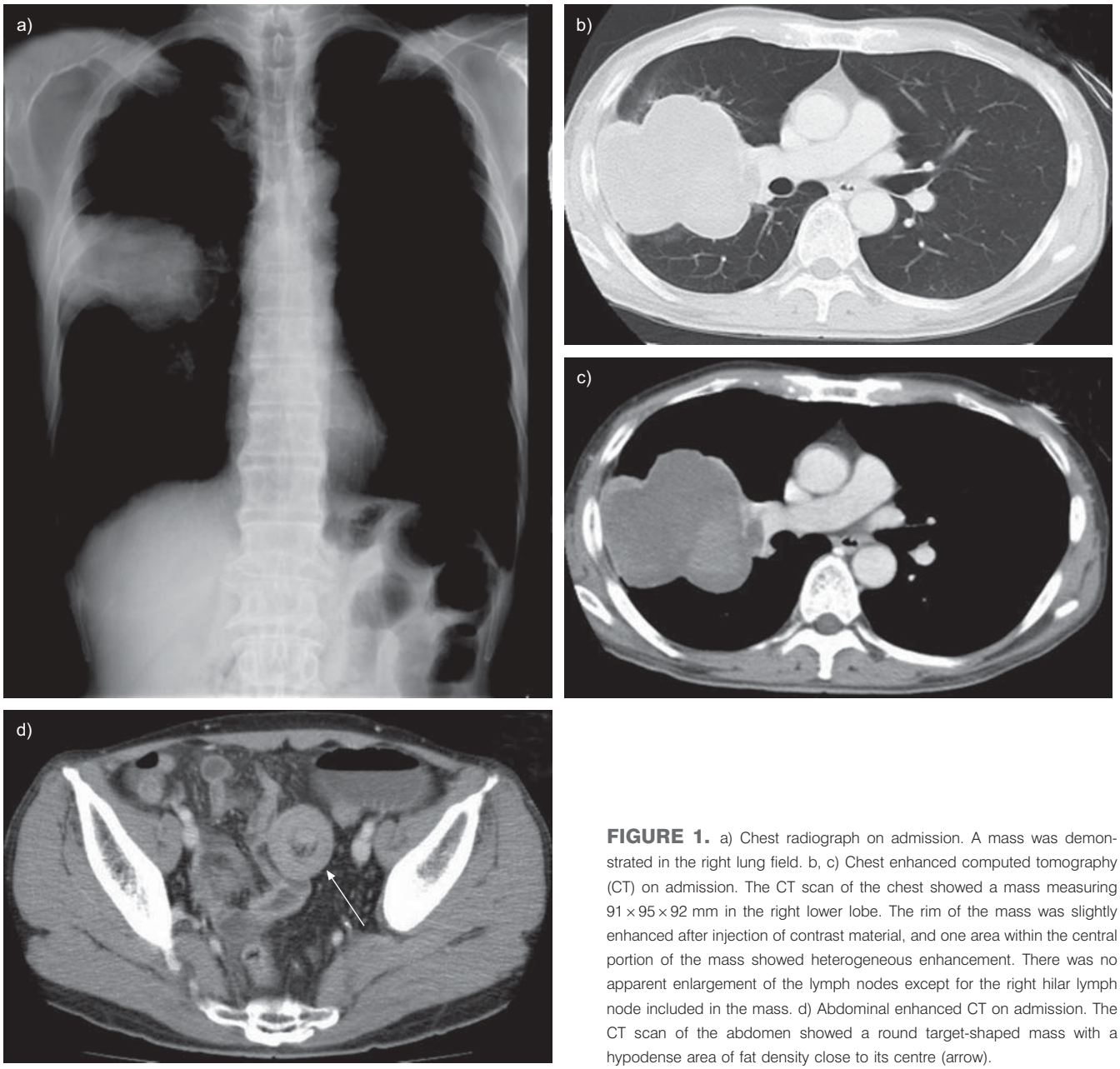

FIGURE 1. a) Chest radiograph on admission. A mass was demonstrated in the right lung field. b, c) Chest enhanced computed tomography (CT) on admission. The CT scan of the chest showed a mass measuring $91 \times 95 \times 92$ mm in the right lower lobe. The rim of the mass was slightly enhanced after injection of contrast material, and one area within the central portion of the mass showed heterogeneous enhancement. There was no apparent enlargement of the lymph nodes except for the right hilar lymph node included in the mass. d) Abdominal enhanced CT on admission. The CT scan of the abdomen showed a round target-shaped mass with a hypodense area of fat density close to its centre (arrow).

sites of metastasis from lung cancer include the brain, bone, adrenal glands and liver. However, metastases to the small intestine are rare.

Primary rhabdoid tumour of the lung is a rare histological and clinical entity, and intussusception of the small intestine due to small intestinal metastasis from this cancer has not been reported previously. In the 1999 World Health Organization classification of lung tumours, the rhabdoid phenotype was included as a variant of large cell carcinoma [3]. BECKWITH and PALMER [4] first described malignant rhabdoid tumours in the kidneys of children in 1978. In 1995, the first definitive case of lung tumour showing rhabdoid morphology was described as a neuroendocrine carcinoma with a rhabdoid phenotype [5]. The rhabdoid cells of these tumours are characterised by the presence of a rounded eosinophilic cytoplasmic inclusion within large cells containing abundant cytoplasm and by the presence of a large eccentric nucleus with a central macronucleolus. Rhabdoid cells show an appearance similar to the cells of rhabdomyosarcoma [1].

Tumours with such a rhabdoid appearance have been described in many extra-renal sites. However, a lack of pathological uniformity in these cases has caused many researchers to doubt the concept of extrarenal rhabdoid tumours as a pathological diagnostic entity [6]. Metastases to the lung from rhabdoid tumours of the kidney and extrarenal rhabdoid tumours are a relatively common finding. Therefore, establishing a diagnosis of primary rhabdoid tumour of the lung requires extensive clinical, pathological and radiological

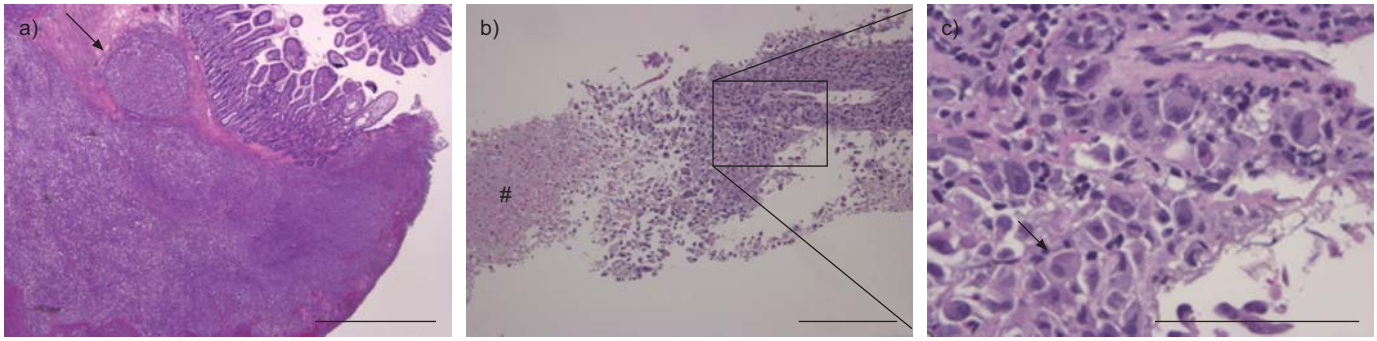

FIGURE 2. Histological findings. a) Small intestinal specimen obtained during surgery. Large tumour cells were mainly located in the submucosa and invaded the vessel (arrow) (haematoxylin and eosin stain.) b) Lung specimen obtained by percutaneous needle biopsy. Pathological findings of the specimen showed several large tumour cells similar to those in the intestinal lesion shown in (a). Abundant cytoplasm and eccentric nuclei and necrotic tissue (#) were also noted (haematoxylin and eosin stain). c) Enlarged view of boxed section in (b) showing several large tumour cells (arrow). a) Scale bar=2,000 μm, b) scale bar=200 μm, c) scale bar=100 μm.

investigations in order to exclude a primary tumour located elsewhere [7]. Microscopically, most LCCRP are high grade or poorly differentiated, and show a combined mixed morphology with a rhabdoid component and an epithelial or mesenchymal component. These tumours usually express epithelial markers, such as cytokeratin and epithelial membrane antigen (EMA), and non-muscle mesenchymal cell markers such as vimentin. However, thyroid transcription factor-1 (TTF-1) is not expressed [1, 7, 8], and is of limited value in determining the site of origin of an extrarenal rhabdoid tumour.

Histological findings of the resected small intestines in this case showed that the tumourous lesion was mainly located in the submucosa and invaded the vessels (fig. 2a). During surgery there were multiple tumour sites in the small intestine, suggesting metastases. Therefore, a primary tumour was suspected to be located elsewhere. The submucosal lesion at the flexure of the small intestine further supported this hypothesis. The larger metastatic lesion was considered to be the cause of intussusception. At present, positron emission tomography (PET) scanning is performed to determine malignancy, as well as staging of malignancy. SAINI *et al.* [1] reported the use of PET for LCCRP. However, we could not perform PET in this patient due to his poor condition. Based on the clinical findings, a diagnosis of primary lung tumour was accepted.

Intestinal metastasis from the lung is rare, whereas squamous cell carcinoma, poorly differentiated pulmonary adenocarcinomas and large cell carcinomas appear to be more common. However, digestive metastasis is more frequent than expected. As there are few symptoms of metastasis to the digestive tract, it is often difficult to diagnosis [2]. In our patient, the initial symptom was intussusception; therefore, we immediately selected surgical treatment.

CT is a useful pre-operative technique for adult intussusception [9]. Adult intussusception is considered a rare condition, accounting for 5% of all cases of intussusception and only 1–5% of intestinal obstructions. In contrast to those in children, cases of intussusception in adults are caused by secondary pathological conditions, such as tumours, metastatic neoplasms, *etc*. Therefore, surgical intervention is necessary [10]. We diagnosed intussusception in our patient based on CT findings. Surgical treatment was performed and his symptoms initially recovered. However, we found that the numerous other sites of small intestinal metastasis that had not been detected by pre-operative CT. Surgery was useful for the treatment and detection of the extent of small intestinal metastases. LCCRP may be considered aggressive, as the majority of patients present with advanced disease. Thus, staging and identification of other metastatic lesions is important, and rapid diagnosis is better.

Including the present case, there are reports of 42 cases in the literature, which are summarised in table 1. Lung tumours with rhabdoid morphology commonly occur in middle-aged to elderly adults presenting with haemoptysis and cough. Most of these cases presented at stage III or IV. Although patients with a short follow-up are included, this neoplasm is characterised by poor prognosis. Including our case, only six cases of digestive metastasis have been reported. Moreover, after expression of clinical symptoms of digestive metastasis, the prognosis is poor. In case 27, the patient died 5 months after gastrointestinal bleeding occurred. In case 39, the patient died 2 months after abdominal pain and tarry stool developed. In case 40, the patient died 4 months after rectal bleeding appeared. More than 10% rhabdoid cells indicate a significantly poorer prognosis, while <5% has a negligible effect on prognosis [8]. In the lung specimens from our patient, >10% of cancer cells were identified as rhabdoid cells, indicating a very poor prognosis. The lung tumour and abdominal lymph nodes rapidly enlarged during the brief post-operative course. Only two reports (cases 21 and 41) showed satisfactory results following timely surgery and adjuvant chemotherapy with rigorous follow-up, but such cases are rare.

In our case, chest CT showed a lobulated well-defined mass, which differed from that usually observed in ''standard'' nonsmall cell lung cancer. There is no consensus regarding the typical findings of LCCRP. The pathological findings of these reports characterised the tumours, the tumours had a clear border but were not encapsulated [7, 17, 19, 20]. The cut surface of these specimens showed haemorrhage [7, 11, 15] and necrosis [7, 11, 14–16, 18, 19]. Moreover, the tumour has a large area of central necrosis with a thin rim of variable tumour cells [11, 19]. Further accumulation of cases is necessary.

In conclusion, lung tumours containing rhabdoid cells are uncommon. We should consider that a patient with LCCRP

TABLE 1 Summary of reported cases of primary rhabdoid tumour of the lung

Case	Age yrs	Sex	Symptoms	Smoking history	TMN stage	Maximum dimension cm	Location	Site of metastases	Associated tumour type	Rhabdoid cells %	Follow-up status	[Ref.]
1	54	F	Haemoptysis	Yes	NA	8.0⩾, ⩽1.3	RUL	Lymph node	Large cell carcinoma	90	AWD, 6 months	11
2	36	M	Chest pain	NA	NA	8.0⩾, ⩽1.3	LUL	NA	Sarcoma	>10	AWD, 4 months	11
3	47	M	Haemoptysis	Yes	NA	8.0⩾, ⩽1.3	RLL	NA	Adenocarcinoma	>10	DOD, 4 months	11
4	71	F	NA	NA	NA	8.0⩾, ⩽1.3	LUL	Lymph node	Adenocarcinoma	10	AWD, 2 months	11
5	71	M	NA	NA	NA	8.0⩾, ⩽1.3	RLL	NA	Large cell carcinoma	>10	ASU, 5 months	11
6	25	F	Bazex's syndrome	No	NA	8.0⩾, ⩽1.3	LLL	Lung, lymph node	Large cell carcinoma	10	NA	11
7	74	M	None	Yes	NA	3.5	LLL	NA	NA	Most	ANED, 24 months	12
8	68	M	Cough, haemoptysis	NA	NA	3	RML	Lymph node	Adenocarcinoma	>10	DOD, 6 months	13
9	62	F	Cough, haemoptysis	Yes	NA	11	RUL	Present	LCNEC	25	DOD, 3 months	13
10	40	M	Cough, chest pain, dyspnoea	Yes	NA	6	RUL	NA	Adenocarcinoma	>10	LFU, 6 months	13
11	50	F	Cough, dyspnoea	Yes	NA	17	LLL	Lymph node	LCNEC, small cell carcinoma	15	DOD, 6 months	14
12	53	M	Cough, fatigue	Yes	NA	12	LUL	NA	LCNEC, small cell carcinoma, SCC	10	DOD, 12 months	14
13	51	M	Haemoptysis	Yes	IIIA	4	RUL	Lymph node	Adenocarcinoma	70	DOD, 36 months	15
14	72	M	Cough	No	IIIA	5.6	RUL	Lymph node	Adenocarcinoma	90	DOD, 4 months	15
15	50	F	Fatigue	No	IA	3.0	RML	NA	Adenocarcinoma	50	ASU, 41 months	15
16	69	M	None	NA	IIIA	4.5	RLL	None	Poorly differentiated tumour	18.2	DOD, 1.5 months	8
17	66	M	Haemoptysis	NA	IIIB	6.0	LUL	None	Poorly differentiated tumour	15.5	DOD, 0.6 months	8
18	82	M	Cough, chest pain, dyspnoea	Yes	IIIA	11.0	LUL	None	Poorly differentiated tumour	15	DOD, 1	8
19	47	F	Cough, haemoptysis	Yes	IIIA	8.5	RUL	None	Poorly differentiated tumour	60	DOD, 10 months	8
20	69	F	Chest pain	Yes	NA	2.0	RUL	Duodenum, skin adrenal gland	Pseudomesotheliomatous adenocarcinoma	NA	Death, other disease; 3 months	16
21	59	M	NA	Yes	NA	4.5	RUL	Adrenal gland	Large cell carcinoma	90	ANED, 60 months	17
22	70	F	None	NA	IB	9	LL	None	Large cell carcinoma	Pure LCCRP	ANED, 8 months	18
23	57	M	Respiratory	Yes	NA	23.0	LUL	Not known	Sarcomatoid carcinoma	25	LFU, NA	7
24	57	F	Haemoptysis	Yes	I	4.0	LUL	Brain, jejunum	Large cell undifferentiated carcinoma	90	DOD, 11 months	7
25	54	M	Haemoptysis	Yes	IV	4.0	RUL	Liver, bone, lymph, node	Large cell undifferentiated carcinoma	15	DOD, 4 months	7
26	48	F	Hoarseness	Yes	IIIB	5.1	RUL	Lung, lymph node	Adenocarcinoma	10	DOD, 19 months	7
27	54	M	Haemoptysis, GI bleeding	NA	IV	10	RUL	Bowel, soft tissue, lymph node	Large cell undifferentiated carcinoma	90	DOD, 5 months	7
28	70	M	None	Yes	IV	NA	Left lung	Bone, chest wall	Adenocarcinoma	90	DOD, 10 months	7
29	61	M	Haemoptysis	Yes	IIIA	4.7	RUL	Bone, lymph node	Sarcomatoid carcinoma	30	DOD, 1.5 months	7
30	59	F	Cough	Yes	IV	NA	RML	Brain, bone, lung, lymph node	Adenocarcinoma	50	DOD, 3 months	7
31	34	M	Haemoptysis	NA	IV	2.0	LLL	Lung	Sarcomatoid carcinoma	60	DOD, 3 months	7
32	65	M	None	NA	IV	NA	LLL	Chest wall	Sarcomatoid carcinoma	75	LFU, NA	7
33	59	F	Haemoptysis	Yes	IIIA	3.0	LLL	Lymph node	Adenocarcinoma	20	ANED, 20 months	7
34	55	M	Cough, haemoptysis	Yes	IV	3.5	LUL	Liver, spleen	Large cell carcinoma	Pure LCCRP	DOD, 2 months	19
35	50	M	Chest pain	NA	I	8	RUL	None	Large cell carcinoma	NGS	ASU, NA	20
36	58	M	Cough, chest pain, dyspnoea	NA	I	3	LLL	None	Large cell carcinoma	NGS	ASU, NA	20
37	56	M	Chest pain, weight loss	NA	I	4	RLL	None	Large cell carcinoma	NGS	LFU, NA	20
38	63	M	Cough, chest pain, weight loss	NA	I	4.8	RUL	None	Large cell carcinoma	NGS	AWD, NA	20
39	37	M	Cough, haemoptysis, back pain	Yes	IIIB	5	RUL	Small intestine, lymph node	Large cell carcinoma	NA	DOD, 6 months	21
40	73	F	Rectal bleeding	Yes	IV	NA	NA	Colon	NA	NA	DOD, 4 months	2
41	36	F	Haemoptysis	No	IB	6	LUL	Liver, lymph node	Large cell carcinoma	NA	AWD, 62 months	1
42	63	M	Intussusception	Yes	IV	9.5	RLL	Small intestine, lymph node	Poorly differentiated tumour	80	DOD, 1 months	Present case report

F: female; M: male; NA: not available; GI: gastrointestinal; RUL: right upper lobe; LUL: left upper lobe; RLL: right lower lobe; LLL: left lower lobe; RML: right middle lobe; LCNEC: large cell neuroendocrine carcinoma; SCC: squamous cell carcinoma; LCCRP: large cell carcinoma of the lung with rhabdoid phenotype; NGS: no evidence of glandular and/or squamous differentiation; AWD: alive with disease; DOD: dead of disease; ASU: alive, status unknown; ANED: alive with no evidence of disease; LFU: lost to follow-up.

will show dynamic progression and has the possibility of metastasis to the digestive tract, as well as the more common metastatic sites. Early diagnosis facilitates intensive chemotherapy, and timely surgical treatment is needed.

STATEMENT OF INTEREST

None declared.

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