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**Multiple clear-cell sarcomas of small intestine with** [**parotid gland**](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) **metastasis: A case report**

Hao Su *et al*. clear cell sarcoma of gastrointestinal tract

Hao Su, Wen-Hao Ren, Peng Wang, Lei Shi, Hai-Tao Zhou

**Hao Su, Peng Wang, Lei Shi, Hai-Tao Zhou,** Department of Colorectal Surgery, Cancer Hospital, Chinese Academy of Medical Science and Peking Union Medical College, Beijing 100021, China

**Wen-Hao Ren,** Department of Pathology, Cancer Hospital, Chinese Academy of Medical Science and Peking Union Medical College, Beijing 100021, China

**Author contributions:** Su H collected the data and drafted the manuscript; Zhou HT designed the study and helped revise the manuscript; Ren WH participated in the descriptions and detailed discussions of the postoperative pathology; Shi L conceived the study and participated in its coordination; Wang P participated in the data interpretation; All authors read and approved the final manuscript.

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**correspondence to:** **Hai-Tao Zhou, MD, Professor**, Department of Colorectal Surgery, Cancer Hospital, Chinese Academy of Medical Science and Peking Union Medical College, No. 17, Pan jia yuan Nan li, Chaoyang district, Beijing 100021, China. [zhouhaitao01745@163.com](mailto:zhouhaitao01745@163.com)

**Telephone:** +86-10-67787110

**Fax:** +86-10-67787110

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Clear-cell sarcoma is a rare, malignant soft tissue tumor that displays melanocytic dif­ferentiation with a distinct molecular profile. It is rarely localized in the gastrointestinal tract. Herein we reported a case of multiple synchronous clear-cell sarcomas of the gastrointestinal tract with [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) metastasis. A 51-year-old male patient presented with a growing painless mass lesion under the right ear. A preopera­tive positron emission tomography/computed tomography showed multiple intestinal masses and a mass in the right parotid with increased glucose uptake, and he subsequently underwent operative treatment with resection of three tumors in the jejunum and ileum and then received a right parotidectomy. Postoperative pathological examination showed cells in the intestinal tumor were consistent with clear-cell sarcoma of the gastrointestinal tract, and the malignant cells in the parotid gland were similar to the intestinal tumor. Immunohistochemical studies revealed positiv­e expression of HMB-45, Melan-A, and S-100. EWSR1 gene fusion transcripts were undetectable by fluorescence in situ hybridization.

**Key words:** Clear-cell sarcomas; Clear-cell sarcomas of the gastrointestinal tract; Parotid gland metastasis; Immunohistochemistry

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**Core tip:** Over the past 13 years, only 53 cases of clear-cell sarcomas of the gastrointestinal tract have been reported in the world. Most of the literature on clear-cell sarcoma of the gastrointestinal tract (CCS-GI) describes a single tumor in the diagnosis; our presentation is the third report of simultaneous tumors during the diagnosis to date and is the first case of CCS-GI with metastasis to the [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation). Our article reviewed all cases of CCS-GI in the literature to date. Because of the high rarity, more cases need to be accumulated for further analysis.

Su H, Ren WH, Wang P, Shi L, Zhou HT. Multiple clear cell sarcomas of small intestine with [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) metastasis: a case report. *World J Gastroenterol* 2017; In press

**INTRODUCTION**

Clear-cell sarcoma (CCS) is a rare tumor of unknown cell origin that was first described by Enzinger *et al*[1] in 1965. CCS shows a predilection for the tendons or aponeuroses in the extremities in young adults 20-40 years old [2]. Ekfors *et al*[3] described the first clear-cell sarcoma of the gastrointestinal tract (CCS-GI) in1993, which occurred in the duodenum. Only finger countable cases[4] of CCS-GI have been reported in the literature to date, but CCS-GI shows specific histopathological, immunohistochemical, and genetic features. Here, we presented a case of three synchronous clear-cell sarcomas in the jejunum and ileum with [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) metastasis.

**Case report**

***Patient details***

A 51-year-old male presented with a two-year history of a growing painless mass lesion under the right ear, initially the size of a soybean. The mass grew noticeably in the last six months. There was a one-year history of night sweats and frequent stool (three to four times a day). There was no history of fever, weakness, dysphagia, [dyspnea](http://dict.youdao.com/w/dyspnea/#keyfrom=E2Ctranslation), cough, hoarseness, jaundice, vomiting, [melena](http://dict.youdao.com/w/melena/#keyfrom=E2Ctranslation), [hematochezia](http://dict.youdao.com/w/hematochezia/#keyfrom=E2Ctranslation), abdominal pain, abdominal distension or significant weight loss. The patient had a 5-year medical history of hypertension and he was a hepatitis-B carrier of 30 years and a smoker of 40 pack-years. There was no family history of cancer.

On palpation, a 20 mm×20 mm relatively well-defined and soft mass with no tenderness was observed along with multiple enlarged cervical nodules. Abdominal examination did not reveal any organomegaly or palpable lumps.

Ultrasonography of the neck two months ago revealed a relatively undefined hypoechoic mass measuring approximately 15 mm×27 mm in its greatest dimension in the right parotid gland and submandibular gland (Figure 1) along with compared with multiple enlarged right supraclavicular and upper cervical lymph nodes. A needle biopsy of the mass was performed and the pathologic report found malignant tumor cells. The patient was recommended for surgery for the mass in the parotid gland. The preoperative blood routine examination showed that the HGB was 106 g/L. Therefore, the patient underwent positron emission tomography/computed tomography (PET/CT). A 36 mm×33 mm intestinal mass with increased glucose uptake, and multiple peripheral lymph nodes in the right mid-abdomen were found (Figure 2), and the maximum standard uptake values (SUV) was 6.6. An intestinal lesion with increased glucose uptake in the right [hypogastrium](http://dict.youdao.com/w/hypogastrium/#keyfrom=E2Ctranslation) was also seen and the SUV was 7.0. The mass in the right parotid and peripheral lymph nodes were also compared with increased glucose uptake, and the SUV was 10.3. Preoperative tumor maker, such as [CA125](javascript:__doPostBack('dgPersonList$_ctl3$_ctl0','')), [CA15-3](javascript:__doPostBack('dgPersonList$_ctl4$_ctl0','')), [CA19-9](javascript:__doPostBack('dgPersonList$_ctl5$_ctl0','')),[CA72-4](javascript:__doPostBack('dgPersonList$_ctl6$_ctl0','')), [AFP](javascript:__doPostBack('dgPersonList$_ctl7$_ctl0','')), [cyfra21-1](javascript:__doPostBack('dgPersonList$_ctl8$_ctl0','')), [NSE](javascript:__doPostBack('dgPersonList$_ctl9$_ctl0','')),[SCC](javascript:__doPostBack('dgPersonList$_ctl10$_ctl0','')), [CEA](javascript:__doPostBack('dgPersonList$_ctl11$_ctl0','')), and [ProGRP](javascript:__doPostBack('dgPersonList$_ctl12$_ctl0','')), did not show [abnormal](http://dict.youdao.com/w/abnormity/#keyfrom=E2Ctranslation) expression.

***Treatment***

The patient received an exploratory laparotomy and the excision of multiple intestinal neoplasms. Operative exploration showed no ascites, pelvic, periaortic, peritoneal, omental deposits, or liver metastasis. No tumors were palpated in the cavity of the stomach, duodenum, colon, rectum, or the mesentery root. Three masses were found at the jejunum and ileum. Intra-operatively, the first tumor was present in the jejunum, located at 80 cm distal to the duodenojejunal junction. [Intussusception](http://dict.youdao.com/w/eng/acute_intussusception/#keyfrom=dict.phrase.wordgroup) was observed from here, and the involved bowels were swelled and expanded (Figure 3). The second tumor was at end of the [intussusception](http://dict.youdao.com/w/eng/acute_intussusception/#keyfrom=dict.phrase.wordgroup) (approximately at the fourth group of intestines). The third tumor was present in the ileum, located at 80 cm proximal to the ileocecal junction. The three tumors of varying sizes invaded the serosa, and the surface of serosa shrunk and was depressed. Multiple enlarged lymph nodes were observed in the intestinal mesentery. Following serial ligation of the mesenteric vessels, resection of the involved bowels with the masses and mesentery was performed with a proximal margin of 10 cm and a distal margin of 10 cm. The first tumor and the second tumor were removed together in one segment of the intestine (Figure 4a and 4b). Then, a primary anastomosis formed. The patient recovered gradually and then received a right parotidectomy with retaining facial nerve and right cervical lymph node dissection 17 d after abdominal surgery because the pathology of the parotid gland neoplasms was not clear, and there were only solitary parotid gland and local cervical lymph node metastasis detected by PET/CT.

***Postoperative pathology***

**Intestinal neoplasms:** Grossly, the specimen consisted of two segments of the small intestine; the larger was approximately 26 cm in length with attached mesentery, and the other segment was 7.8 cm with attached mesentery. Two tumors were on the larger intestine; 11 cm from one margin there was a 2.5 cm× 2.2 cm× 1 cm tumor and 19 cm from the same margin there was a 6.5 × 5.5 × 4 cm tumor. A 2.5 cm× 1.9 cm× 1 cm tumor was on the other segment of small intestine. The cut surface of the three tumors had hard, obscure borders, white to tan in appearance.

[Microscopically](http://dict.youdao.com/w/microscopically/#keyfrom=E2Ctranslation), the jejunum and ileum tissues were infiltrated with malignant cells, which was consistent with CCS-GI (a type of gastrointestinal neural ectoderm tumors, GNET) by [morphology](http://dict.youdao.com/w/cellular%20morphology/#keyfrom=E2Ctranslation) and immunohistochemistry (Figure 5a). The tumors invaded the mucosal and muscular layers. There was no exact focal necrosis, vessel invasion or nerve invasion. The mitotic index exceeded 20/10 HPF, and the tumor was grade G3 according to the FNCLL (French Fédération Nationale des Centres de Lutte Contre le Cancer) system.

Lymph node metastases (1/29), not invading outside lymph node capsule: (1) Peripheral lymph nodes of jejunum 1/26; and (2)Peripheral lymph nodes of ileum 0/3.

Immunohistochemistry: S100 (3+), Vim (3+), GFAP (-), HMB-45 (2+), Melan-A (2+), Melanomapan (1+), CD56 (2+), Syn (-), CgA (-), AE1/AE3 (-), CD138 (-), CD19 (-), CD20 (-), CD3 (-), CD38 (-), CD79a (-), Ki-67 (+40%), LCA (-), MUM1 (-), CD117 (lesion+), CD34 (-), DOG1 (-), CD10 (-), Calponin (-), P63 (-), EBER (-).

[Gene detection](http://dict.youdao.com/w/gene%20detection/#keyfrom=E2Ctranslation): EWSR1 gene fusion transcripts were undetectable by fluorescence in situ hybridization (FISH).

**Parotid gland neoplasms:** Grossly, a 1 cm diameter nodule was found in 5.5 cm× 3 cm× 2 cm tissues; the cut surface of the nodule had a tough, grey to yellow appearance.

[Microscopically](http://dict.youdao.com/w/microscopically/#keyfrom=E2Ctranslation), parotid gland tissues were infiltrated with malignant cells, which was consistent with clear-cell sarcoma [morphology](http://dict.youdao.com/w/cellular%20morphology/#keyfrom=E2Ctranslation) and immunohistochemistry and similar to the previous intestinal tumor [morphologically](http://dict.youdao.com/w/cellular%20morphology/#keyfrom=E2Ctranslation) (Figure 5b). Lymph tissues were found in the tumor and tumor edge, which may be metastatic lesions.

No lymph node metastases (0/30): (1) right cervical lymph nodes level II 0/10; (2) right cervical lymph nodes level III 0/12; (3) right cervical lymph nodes level V 0/5; (4) peripheral lymph nodes of superficial lobe of right parotid gland 0/2; (5) peripheral lymph nodes of caudate lobe of right parotid gland and tumor 0/1.

Immunohistochemistry: S100 (3+), Melan-A (3+), Melanomapan (3+), HMB-45 (3+), AE1/AE3 (-), CK18 (-), Calponin (-), P63 (-), SMA (-).

***Follow-up***

Twenty days after the surgery on the parotid gland, the patient received computed tomography (CT) imaging of the neck, thorax and abdominopelvic area; no recurrence or metastasis was observed. Then, he began 6 cycles of chemotherapy with the regimen of EI (Epirubicin 100 mg + Ifosfamide 2 g D1-4+Mesna 0.4 g 0 h, 4 h, 8 h after the Ifosfamide D1-4). At the time this article was written, the patient was in the first cycle of the chemotherapy.

**Discussion**

CCS-GI is so rare that only 53 cases (including our case) have been reported in the literature to date (Table 1)[3,5-39]. Most of the literature on CCS-GI describes a single tumor in the diagnosis; only two cases[25,38] report two simultaneous tumors during the diagnosis to date. CCS-GI often involves the ileum and jejunum, stomach and colon[4-7,9-12,14-35,38,39]. Because of the aggressive clinical course, regional and distant metastases are common in CCS-GI at presentation[5-7,9,10,15,17,21,25,27,29,31,37,39]. Lymph nodes, liver, and mesentery are the most common destinations of metastases at the time of presentation. The patient in our report had three synchronous masses in in the jejunum and ileum, with a metastasis to the [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation), and he came to the hospital mainly due to the swollen [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation). Lymph nodes inside and outside of [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) make it a common metastatic destination for [head and neck neoplasm](http://dict.youdao.com/w/head%20and%20neck%20neoplasm/#keyfrom=E2Ctranslation)s[40], but it is very rare for gastrointestinal tumors. Among the limited literature on CCS-GI, this is the first case of CCS-GI with metastasis to the [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation).

CCS-GI shows specific histopathological, immunohistochemical, ultrastructural, and genetic features[2,4]. In 2010, Kosemehmetoglu *et al*[41] first divided CCS-GI into two subtypes according to the histomorphology: (1) clear-cell sarcoma-like gastrointestinal tumor (CCSLGT); and (2) clear-cell sarcoma of soft tissue (CCS-ST). However, there was disagreement about whether they were two independent entities[31]. In 2003, Zambranode *et al*[10] reported 6 cases of CCSLGTs. They found CCSLGT was at least focally positive for the S100 protein, but most did not express melanocytic markers such as HMB-45 or Melan-A. While Huang *et al*[36] found parts of CCS-STs were positive for the S100 protein, and most could express melanocytic markers such as HMB-45 or Melan-A. Some reports found that > 90% of cases of CCS were associated with the reciprocal translocation t (12; 22) (q13; q12), resulting in fusion of the EWSR1 gene located at 22q12 and the ATF1 gene located at 12q13[2,41-46]. To date, these translocations have never been observed in malignant melanoma[13,22,43-46], which has a very similar histologic appearance to CCS[20]. Immunohistochemical staining of CCS reveals positivity for the S100 protein as well as melanocyte-specific markers, with this combination of staining allowing for CCS to be distinguished from malignant melanoma histologically. In our case, the tumor was consistent with CCS-GI by [morphology](http://dict.youdao.com/w/cellular%20morphology/#keyfrom=E2Ctranslation), positive for S100 protein, and expressed melanocytic markers such as HMB-45 and Melan-A, but EWSR1 gene fusion transcripts were undetectable by FISH.

Currently the most effective treatment for the CCS-GI is extensive resection of the tumor and peripheral lymph nodes; chemotherapy and radiotherapy appear to have little effect[31]. The clinical behavior of CCCS-GI appears to be highly aggressive, with high local recurrence, lymph node or visceral metastases, or death generally in < 36 mo[41,46]. The patient underwent excision of multiple intestinal neoplasms and right parotidectomy before the first cycle of the chemotherapy and no recurrence or metastasis was observed from the follow-up to date.

In conclusion, CCS-GI is a highly rare soft-tissue sarcoma with distinct morphological, immunohistochemical, and genetic features. This case demonstrates that the [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) is a potential metastatic site for CCS-GI. For the routine method to diagnose and treat of CCS-GI, more cases need to be accumulated for further analysis.

**COMMENTS**

***Case characteristics***

A 51-year-old male presented with a two-year history of a growing painless mass lesion under the right ear, which grew noticeably in the last six months, and a one-year history of night sweats and frequent stool.

***Clinical diagnosis***

A relatively well-defined and soft mass with no tenderness was observed along with multiple enlarged cervical nodules.

***Differential diagnosis***

Small intestinal stromal tumors, lymphoma, [head and neck neoplasm](http://dict.youdao.com/w/head%20and%20neck%20neoplasm/#keyfrom=E2Ctranslation), Sarcomatoid carcinoma

***Laboratory diagnosis***

The patient had no remarkable findings for the laboratory tests.

***Imaging diagnosis***

PET/CT showed an intestinal mass with multiple peripheral lymph nodes and mass in the right parotid.

***Pathological diagnosis***

Intestinal neoplasms and parotid gland neoplasms were consistent with clear-cell sarcoma by [morphology](http://dict.youdao.com/w/cellular%20morphology/#keyfrom=E2Ctranslation) and immunohistochemistry.

***Treatment***

The patient received a curative resection and postoperative chemotherapy.

***Related reports***

Only 53 cases of CCS-GI have been reported in the literature to date; CCS-GI shows distinct morphological, immunohistochemical, and genetic features.

***Term explanation***

Clear-cell sarcoma of the gastrointestinal tract ( CCS-GI) is a highly rare soft tissue sarcoma.

***Experiences and lessons***

The present case report is the third diagnosis with simultaneous multiple CCS-GI to date and the first case of CCS-GI with metastasis to the [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation).

***Peer-review***

The authors have described a case of multiple clear-cell sarcomas of the small intestine with [parotid gland](http://dict.youdao.com/w/parotid%20gland/#keyfrom=E2Ctranslation) metastasis. The article highlights the morphological, immunohistochemical, and genetic features of the tumors.

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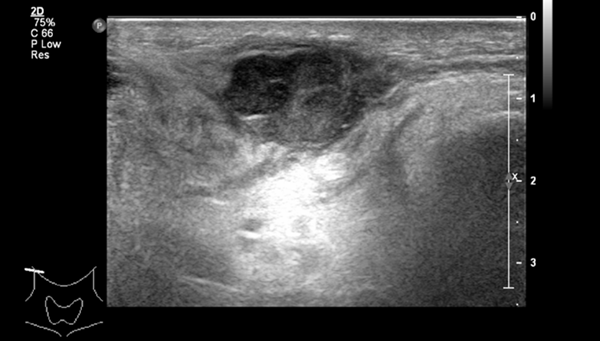
Grade A (Excellent): 0

Grade B (Very good): B

Grade C (Good): 0

Grade D (Fair): D

Grade E (Poor): 0

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**Figure 1 ultrasonogram of the neck showed 15 mm×27 mm mass in the right parotid gland.**

****

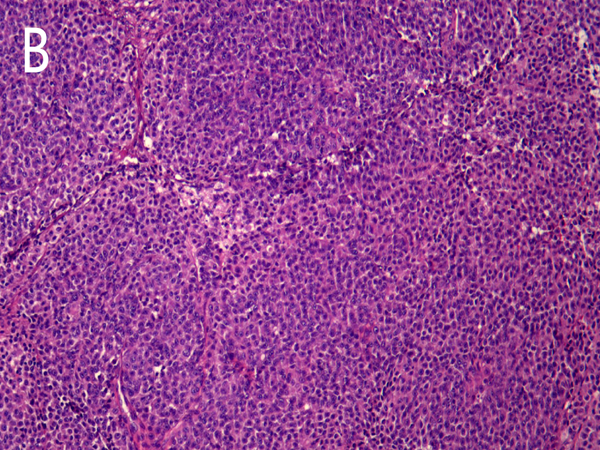
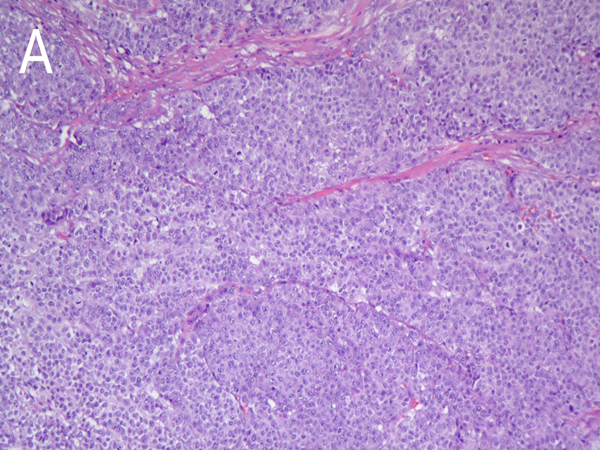
**Figure 2 PET/CT showed a 36 mm×33 mm intestinal mass with multiple peripheral lymph nodes in the right midabdomen.**



**Figure 3** [**Intussusception**](http://dict.youdao.com/w/eng/acute_intussusception/#keyfrom=dict.phrase.wordgroup) **was observed 80 cm distal to the duodenojejunal junction and involved bowels swelled and expanded.**



**Figure 4 Involved bowels with the masses and mesentery was resected with proximal 10 cm and distal 10 cm margin.**

****

**Figure 5** [**Microscopic observation**](http://dict.youdao.com/w/microscopic%20observation/#keyfrom=E2Ctranslation) **of intestinal neoplasms and parotid gland neoplasms.**A:Microphotography shows polygonal malignant cells of intestinal neoplasms were separated by fibrous tissues, arranging in sheets and nests, with eosinophilic or clear cytoplasm and there was no exact necrosis, vessel invasion and nerve invasion. [Nucleolus](http://dict.youdao.com/w/nucleolus/#keyfrom=E2Ctranslation) was obvious and the mitotic index exceeded 20/10 HPF (Hematoxylin-Eosin G×10); B: Malignant cells of parotid gland neoplasms were similar to the intestinal tumor by microphotography (Hematoxylin-Eosin G×10).

**Table 1 clinical, pathological, immunohistochemical, and genetic features of clear-cell sarcoma of the gastrointestinal tract in previously reported cases**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Ref. | Age(yr)/sex | Location | Maximum diameter of tumor(cm) | S-100 | HMB-45 | Melan-A | Genetic  findings | Outcome |
| Alpers *et al*[5] | 26/F | Jejunum | 1.5 | ND | ND | ND | ND | Liver mets |
| Ekfors *et al*[3] | 38/M | [Duodenum](http://dict.youdao.com/w/duodenum/#keyfrom=E2Ctranslation) | 3 | Positive | Positive | ND | ND | Not given |
| Donner *et al*[6] | 37/M | Ileum | 6.5 | Positive | Negative | ND | t(12;22)(q13;q12-13) | Liver mets at 24 and 36 mo |
| Fukuda *et al*[7] | 74/M | Colon | 3 | Positive | Positive | ND | EWSR1-ATF1 by RT-PCR | Liver mets at 9 mo |
| Hu *et al*[8] | 10/M | Rectum | 5 | Positive | Positive | ND | ND | NA |
| Pauwels *et al*[9] | 30/M | Stomach | 4 | Positive | Negative | ND | t(12;22)(q13;  q12) | LN and peritoneal mets at diagnosis |
| Zambrano *et al*[10] | 15/F | Jejunum | 5 | Positive | Negative | Negative | t(12;22)(q13;  q12) | DOD 16 mo |
|  | 21/F | Jejunum | 4 | Positive | Negative | Negative | ND | DOD 12 mo |
|  | 35/F | Ileum | 3.5 | Positive | Negative | Negative | ND | Liver mets at 12 mo |
|  | 37/F | Ileum | 4.5 | Positive | Negative | Negative | ND | NA |
|  | 32/M | Ileum | 5 | Positive | Negative | Negative | ND | NA |
|  | 13/M | Stomach | 6.7 | Positive | Negative | Negative | ND | Local recurrence at 12 mo;2nd Local recurrence at 36 mo |
| Achten *et al*[11] | 57/M | Jejunum | 6.5 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NA |
| Venkataraman *et al*[12] | 21/F | Ileum | 7 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NA |
| Covinsky *et al*[13] | 47/F | Pancreas | NA | Positive | Positive | Positive | EWSR1-ATF1  by RT-PCR and  FISH | NED 24 mo |
|  | 85/F | Mesentery | NA | Positive | Positive | Positive | EWSR1-ATF1  by RT-PCR and  FISH | DOD 1 mo |
| Taminelli *et al*[14] | 35/M | Ileum | 1.8 | Positive | Negative | Positive | EWSR1-ATF1/ by RT-PCR | DOD 15 mo |
| Friedrichs *et al*[15] | 41/M | Jejunum | 8.7 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | Liver mets at 6 mo |
| Huang *et al*[16] | 40/M | Stomach | 3 | Positive | Negative | Positive | ND | NED 9 mo |
| Antonescu *et al*[17] | 81/F | Colon | 7.5 | Positive | Negative | Negative | EWSR1-CREB1 by RT-PCR | Mets to liver and  peritoneum at  60 mo |
|  | 42/F | Ileum | 5.7 | Positive | Negative | Negative | EWSR1-CREB1 by RT-PCR | NA |
|  | 42/F | Ileum | 3.5 | Positive | Negative | Negative | EWSR1-CREB1 by RT-PCR | Peritoneal and  liver mets at  diagnosis |
|  | 51/F | Jejunum | NA | Positive | Negative | Negative | EWSR1  rearrangement  by FISH | Peritoneal and  liver mets; AWD |
|  | 18/F | Jejunum | NA | Positive | Negative | Negative | EWSR1-ATF1 by RT-PCR | Local recurrence |
| Granville *et al*[18] | 16/M | Ileum | 5 | Positive | Negative | ND | EWSR1-ATF1 by RT-PCR; t(12;22)(q13;q12) | DOD 15 mo |
| Comin *et al*[19] | 31/F | Ileum | 2.8 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NA |
| Lyle *et al*[20] | 46/M | Jejunum | 11 | Positive | Positive | Positive | EWSR1 rearrangement by FISH; EWSR1-ATF1 by RT-PCR | NED 7 mo |
|  | 49/M | Cecum | 10.5 | Positive | Positive | Positive | EWSR1 rearrangement by FISH; EWSR1-ATF1 by RT-PCR | DOD 12 mo |
|  | 60/M | Jejunum | 10 | Positive | Positive | Positive | EWSR1-ATF1 by RT-PCR | DOD 28 mo |
|  | 62/M | Ileum | 4 | Positive | Positive | Positive | EWSR1 rearrangement by FISH; EWSR1-ATF1 by RT-PCR | DOD 12 mo |
| Abdulkader *et al*[21] | 37/M | Jejunum | 8.2 | Positive | Negative | ND | EWSR1 rearrangement by FISH | Liver mets at 2 mo |
| Lagmay *et al*[22] | 10/F | Stomach | 7.8 | Positive | Negative | Negative | EWSR1 rearrangement by FISH; EWSR1-ATF1 by RT-PCR | NED 4 mo |
| Joo *et al*[23] | 60/M | Ileum | 2.4 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NA |
|  | 46/M | Jejunum | 6 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NA |
| Terazawa *et al*[24] | Early 20s/F | Ileum | 3 | Positive | ND | ND | EWSR1-ATF1 by RT-PCR | NED at 24 mo |
| Shenjere *et al*[25] | 53/F | Ileum | 5 | Positive | Negative | Negative | EWSR1-ATF1  by RT-PCR | Regional LN mets at diagnosis/  NED at 7 mo |
|  | 26/F | 1Small and large bowel | 13.5/10.1 | Positive | Negative | Negative | EWSR1-CREB1 by RT-PCR | NA |
|  | 66/M | Ileum | 2.5 | Positive | Negative | Negative | EWSR1-CREB1 by RT-PCR | Regional LN mets at diagnosis/NED |
| Balkaransingh *et al*[26] | 15/M | Ileum | NA | ND | ND | ND | EWSR1 rearrangement by FISH | NA |
| Yang *et al*[27] | 15/M | Ileum | 4 | Positive | ND | ND | EWSR1 rearrangement by FISH | Liver mets at 12 mo |
| Suarez-Vilela *et al*[28] | 36/F | Jejunum | 1.5 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NA |
| D’Amico *et al*[29] | 69/F | Ileum | 4 | Positive | Negative | ND | EWSR1 rearrangement by FISH | Liver mets at 2 mo |
| Lasithiotakis *et al*[30] | 49/F | Jejunum | 3 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NED 20 mo |
| Huang Hui-fen *et al*[31] | 45/F | Colon | 4 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | Liver mets at 20 mo |
| Mallick *et al*[32] | 45/M | Jejunum | 4.4 | Positive | Negative | Negative | ND | NA |
| Kong *et al*[33] | 17/M | Stomach | 6 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NED 10 mo |
| Liu *et al*[34] | 76/M | Jejunum | 2.5 | Positive | Negative | Negative | EWSR1-ATF1 by RT-PCR | NA |
| Thway *et al*[35] | 36/M | Ileum | 3 | Positive | Negative | Negative | EWSR1-CREB1 by RT-PCR | DOD 7 mo |
| Huang *et al*[36] | 36/M | Pancreas | 4 | Positive | Positive | Positive | EWSR1 rearrangement by FISH | Liver mets at 10 mo |
| Yegen *et al*[37] | 25/F | Ileum | 3.2 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | Liver mets at diagnosis and at 15 mo. Ovarian mets and peritoneal dissemination at 47 mo |
| Moslim *et al*[38] | 57/M | 2Duodenum and Jejunum | 5.5/7.5 | Positive | Negative | Positive | EWSR1 rearrangement by FISH | NED 30 mo and then DOD 4 mo later |
| Chen *et al*[39] | 29/F | Jejunum | 6 | Positive | Negative | Negative | EWSR1 rearrangement by FISH | NED 17 mo |
| Our case | 51/M | 3Duodenum and Jejunum | 6.5/2.5/2.5 | Positive | Positive | Positive | EWSR1 rearrangement undetectable by FISH | NED up to date |

1Two simultaneous tumors in small and large bowel; 2Two simultaneous tumors in Duodenum and Jejunum; 3Three simultaneous tumors in duodenum and Jejunum. AWD: Alive with disease; DOD: Dead of disease; FISH: Fluorescence in situ hybridisation; LN: Lymph node; Mets: Metastases; NA: Not acquired; ND: Not done; NED: No evidence of disease; RT: Reverse transcription.