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Metastatic clear cell sarcoma of the pancreas: A rare case report

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Abstract

BACKGROUND

Clear cell sarcoma (CCS) is a rare soft-tissue sarcoma. The most common metastatic sites for CCS are the lungs, bones, and brain. CCS is highly invasive and mainly metastasizes to the lung, followed by the bone and brain; however, pancreatic metastasis is relatively rare.

CASE SUMMARY

We report a rare case of CCS with pancreatic metastasis in a 47-year-old man. The patient had a relevant medical history 3 years ago, with abdominal pain as the main clinical manifestation. No abnormalities were observed on physical examination, and the tumor was found on abdominal computed tomography. Based on the medical history and postoperative pathology, the patient was diagnosed with CCS with pancreatic metastasis. The patient was successfully treated with surgical interventions, including distal pancreatectomy and splenectomy.

CONCLUSION

This report summarizes the available treatment modalities for CCS and the importance of regular postoperative follow-up for patients with CCS.

INTRODUCTION

Clear cell sarcoma (CCS) is a rare soft-tissue sarcoma (STS) that accounts for less than 1% of all cases and was originally reported in 1965[1,2]. The incidence of CCS is estimated to be approximately 0.014/100000 depending on the surveillance, epidemiology, and end results databases[3]. CCS originates from the tendon and aponeurosis, and is characterized by local invasive growth of the tendon and other soft tissues[4]. CCS is a highly invasive type that mainly metastasizes to the lung, followed by the bone and brain; however, pancreatic metastasis is relatively rare[3-8]. To our knowledge, only two cases of primary pancreatic CCS have been reported[9,10]. Here, we provide details of the clinical manifestations and imaging features of a patient with metastatic CCS of the pancreas.

CASE PRESENTATION

Chief complaints

A 47-year-old male presented with abdominal pain and diarrhea.

History of present illness

Symptoms started more than a half month before presentation with abdominal pain and diarrhea.

History of past illness

None

Personal and family history

None

Physical examination

None

Laboratory examinations

None

Imaging examinations

Computed tomography (CT) and magnetic resonance imaging (MRI) revealed an isolated lesion in the pancreatic tail without metastasis (Figure 1 and 2).

FINAL DIAGNOSIS

The preoperative diagnosis did not rule out the possibility of CCS recurrence combined with the patient's disease history.

TREATMENT

The patient underwent surgical treatment. Under general anesthesia, the abdomen was prepped and draped in the usual fashion, with the patient in the supine position. A midline incision of the abdomen was made, and the peritoneal cavity was entered. Generalized abdominal exploration revealed no metastases to the liver, spleen, or peritoneal omentum of the abdominopelvic cavity. The gastrocolic ligament was opened in the avascular zone, and the stomach was turned upward to reveal the pancreas. A solid tumor measuring 3 × 3 × 4 cm with a hard texture and smooth surface located in the pancreatic body was found during surgery. A distal pancreatectomy and splenectomy were successfully performed. The posterior, transverse, caudal, and dorsal pancreatic arteries and pancreatic freeing were ligated. The pancreatic tissues were gradually dissected by suturing 2 cm away from the tumor, close to the neck of the pancreas. The pancreatic body and tail and the spleen were excised. No tumor residue was found in the pancreatic margins or the spleen.

OUTCOME AND FOLLOW-UP

The patient recovered well after surgery without complications and was free of any discomfort until the last follow-up (6 months postoperatively), with refusal of any other therapy or examination.

DISCUSSION

CCS is a malignant tumor arising from the neural spinal cells caused by the *EWSR1-ATF1* gene fusion resulting from t(12;22) (q13;q12) translocation[11]. CCS often occurs in the second to fourth decades of life and affects the lower extremities. CCS metastasis to the gastrointestinal system (CCSLGT) is rare and was first reported by Zambrano *et al* in 2003[12]. CCSLGT is prevalent in the walls of the small intestine, stomach, colon, and peritoneum, but is rare in the pancreas[13,14]. The manifestations of CCSLGT include abdominal pain, intestinal obstruction, anemia, nausea, and vomiting. The clinical presentation in this case was abdominal pain with nausea, which is consistent with the literature.

Currently, there is no unified management method for advanced CCS, and surgery remains the standard treatment, similar to most STSs[4,7,15,16]. Ensuring that the margins of the tumor are clean and retaining a ring of normal tissue around the tumor is essential. Preoperative imaging assessment in conjunction with patient history is appropriate. CT provides initial postoperative follow-up and evaluation. MRI is an important modality that can reveal the location and extent of tumor invasion[17,18]. In this case, the patient was initially diagnosed with metastatic CCS on preoperative MRI, thus providing a good plan for surgery and ensuring clean surgical margins.

CCS has a poor prognosis, with limited effectiveness of other postoperative treatments. According to the literature, chemotherapy and radiotherapy for CCS are unsatisfactory[19-22]. Some studies have demonstrated that targeted therapy or immunotherapy may be effective; however, no confirmatory clinical trials have been conducted[23-27].

Despite optimal management of local disease, CCS has a high probability of recurrence or metastasis, and when it progresses to stages III and IV, the 5-year survival rate significantly reduces to 15-35%[3]. Long-term follow-up is necessary for patients with CCS, because new metastatic lesions may appear after a longer period[28]. Postoperative management is important in CCS to effectively detect early metastases. In this case, the patient was not followed up after the first operation until symptoms appeared, leading to delayed treatment. At this time, the patient resisted follow-ups and examinations that would have facilitated disease monitoring.

CONCLUSION

A rare case of CCS in a patient who developed pancreatic metastases 3 years after surgery has been reported. Regularized follow-up and re-examination were not performed after the first operation, resulting in patient being seen only after the onset of symptoms, which may have delayed the treatment. Therefore, regular postoperative follow-up is important for patients with CCS because recurrence has a long-term course.

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