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CASE REPORT

Clinical presentation of gastric Burkitt lymphoma presenting with paraplegia and acute pancreatitis: A case report

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Abstract

BACKGROUND

The incidence of gastric Burkitt lymphoma (BL), presenting as paraplegia and acute pancreatitis, is extremely low. BL is a great masquerader that presents in varied forms and in atypical locations, and it is prone to misdiagnosis and missed diagnosis. The prognosis of BL remains poor because of the difficulty in early diagnosis and the limited advances in chemotherapy.

CASE SUMMARY

A 53-year-old man was referred to our hospital from the local county hospital due to abdominal pain for two weeks and weakness in the lower extremities for one day. Magnetic resonance imaging of the abdomen and lumbar spine showed a swollen pancreas and gallbladder, with peripancreatic exudation and liquid collection, indicating acute pancreatitis and acute cholecystitis. Additionally, we observed abnormally thickened lesions of the gastric wall, multiple enlarged retroperitoneal lymph nodes and a well-demarcated, posterolateral extradural mass lesion between T9 and T12, with extension through the spinal foramen and definite bony destruction, suggesting metastasis in gastric malignancy. Subsequent whole-body positron emission tomography/computed tomography examination showed multifocal malignant lesions in the stomach, pancreas, gallbladder, bone, bilateral supraclavicular fossa, anterior mediastinum, bilateral axillary and retroperitoneal lymph nodes. Gastroduodenal endoscopy revealed primary BL with massive involvement of the gastric body and duodenum. The patient refused chemotherapeutic treatment and died one week later due to upper gastrointestinal hemorrhage. Afterward, we reviewed the characteristics of 11

patients with BL involving the stomach, pancreas or spinal cord.

CONCLUSION
Clinicians should be aware that BL can be the potential cause of acute pancreatitis or a rapidly progressive spinal tumor with accompanying paraplegia. For gastric BL, gastroscopy biopsies and pathology are necessary for a definite diagnosis.

Key Words: Burkitt lymphoma; Paraplegia; Acute pancreatitis; Case report

Core Tip: The incidence of Burkitt lymphoma (BL) is extremely low, and the clinical symptoms are atypical. The misdiagnosis rate is high, and the patient's prognosis is poor. The patient in this case was eventually diagnosed with BL involving the stomach, pancreas and vertebral column presenting with acute pancreatitis and neurological symptoms secondary to compression of the spinal cord. Chemotherapeutic treatment was refused by the patient, and he eventually died after one week due to upper gastrointestinal hemorrhage. This case reminds us that further transcriptomic and clinical studies are needed to explore desirable biomarkers for early BL. Eleven cases were reviewed with an emphasis on diagnostic criteria and treatment protocols. Clinicians need to raise awareness of BL and reduce misdiagnosis rates.

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INTRODUCTION

Burkitt lymphoma (BL) is a subgroup of high-grade non-Hodgkin's lymphoma (NHL) with an aggressive clinical course that was first described as a clinical entity in children in Central Africa by Denis Burkitt in 1958[1]. Clinically, patients with BL often present with solid tumors or large lymph nodes or symptoms similar to acute leukemia, and bone marrow invasion is present in more than 25% of cases[2]. BL has been classified into three subtypes according to the World Health Organization classification: Sporadic type, endemic type and immunodeficiency-associated type[3]. Endemic BL is most prevalent in children from equatorial Africa and New Guinea. Approximately 50% of endemic BL affects the jaw or kidneys. This endemic subtype could also occur in the distal ileum, cecum, greater omentum, ovaries and breasts. Nearly all cases are associated with Epstein-Barr virus[4]. Sporadic BL most commonly affects children[5] but represents less than 1% of NHL cases among adults[6]. Most sporadic BL occurs in the bowel, respiratory tract-associated lymphoid tissue and gut-associated lymphoid tissue. Immunodeficiency-associated BL is most frequently present in Human Immunodeficiency Virus-positive patients[4]. BL is highly sensitive to chemotherapy. Despite the long-term treatment-related sequelae of patients with BL treated with high-intensity chemotherapy regimens, patients who tolerate highly intensive combination chemotherapy regimens tend to have excellent oncologic outcomes. Currently, most treatment protocols for adult patients are based on pediatric clinical trials, and treatment-related toxicities remain a major barrier for those with advanced age. Hence, the overall prognosis for adult patients remains dismal[7]. Due to the rapid proliferation of BL, early diagnosis is essential for the effective treatment of BL. Until recently, BL was diagnosed mainly on the basis of clinical presentation, histopathological changes, morphology, immunophenotype and genotype. There have been few reports on adult patients with sporadic BL, especially adult patients with severe involvement of the stomach, pancreas and spinal cord. Herein, we report a case of gastric BL in an adult patient presenting with paraplegia and acute pancreatitis, along with a review of the literature.

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CASE PRESENTATION

Chief complaints

A 53-year-old male patient was admitted to the hospital with abdominal pain for two weeks and weakness in the lower extremities for one day.

History of present illness

This patient was admitted to the local hospital because of epigastric pain after alcohol consumption. He described the pain as intermittent, non-radiating and worsening with food consumption. The patient denied nausea, vomiting, constipation, fever or progressive weight loss. Based on abdominal pain, elevated levels of serum amylase, and findings of peripancreatic exudation and effusions by computed tomography (CT), the patient was diagnosed with acute pancreatitis. The patient was treated with antibiotics, proton pump inhibitors, fasting and short-term intravenous feeding and fluid therapy, and the abdominal pain was alleviated slightly. Unfortunately, on the 14th d of hospitalization, this patient developed a sudden onset of aconuresis and paraplegia. He was referred to our hospital for further examination.

History of past illness

The patient reported no remarkable history of past illness.

Personal and family history

There was no family history of malignant tumors.

Physical examination

The patient's vital signs were stable. No superficial lymphadenopathy was palpable. Regarding the pulmonary and cardiac examination, no obvious abnormality was observed. The abdomen was flat and soft. Physical examination revealed epigastric tenderness without rebound tenderness or Murphy's sign. No jaundice or palpable masses were observed. Neurologic examination revealed no abnormality in his cranial nerves. The muscle strength of the upper limbs was normal, while it was grade I in the lower limbs. Deep tendon reflexes in the affected limbs were diminished or absent. Bilateral Babinski signs were positive. Hypoesthesia beneath the T8 sensory dermatome was observed. Meningeal irritation signs were negative. He also showed bladder-urinary dysfunction.

Laboratory examinations

The auxiliary examination at admission showed that the white blood cell count was 14.68 × 10^9/L (normal range, 3.5 × 10^9/L - 9.5 × 10^9/L), RBC count was 3.95 × 10^9/L (normal range, 4.3 × 10^9/L - 5.8 × 10^9/L), HGB was 136.0 g/L (normal range, 130-175 g/L), PLT count was 324 × 10^9/L (normal range, 100 × 10^9/L-350 × 10^9/L), C-reactive protein was 34.64 mg/L (normal range, 0-6 mg/L), procalcitonin was 0.12 ng/mL (normal range, 0-0.05 ng/mL), serum amylase was 266 U/L (normal range, 0-125 U/L), lactic dehydrogenase (LDH) was 526 U/L (normal range, 71-231 U/L), and uric acid was 799 μmol/L (normal range, 71-231 μmol/L). Laboratory tests showed no abnormalities in liver function or electrolytes. His carbohydrate antigen 19-9 was 461.28 U/mL (normal range, 0-35 U/mL), and carbohydrate antigen 12-5 was 126.90 U/mL (normal range, 0-35 U/mL). Other tests revealed normal tumor marker levels, including carcino-embryonic antigen and alpha fetoprotein levels of 0.56 ng/mL (normal range, 0-5 ng/mL) and 2.6 ng/mL (normal range 0-8.1 ng/mL), respectively.

Imaging examinations

A CT scan at admission showed a swollen pancreas and gallbladder, with peripancreatic exudation and liquid collection, indicating a diagnosis of acute pancreatitis and acute cholecystitis. Magnetic resonance imaging (MRI) of the abdomen and lumbar spine at the 14th d after admission showed a swollen pancreas and gallbladder, with less peripancreatic exudation and liquid collection, indicating the remission of acute pancreatitis and acute cholecystitis. Additionally, MRI showed abnormally thickened lesions of the gastric wall, multiple enlarged retroperitoneal lymph nodes and a well-demarcated, posterolateral extradural mass lesion between T9 and T12, with extension through the spinal foramen and definite bony destruction (Figures 1 and 2). Whole-body positron emission tomography-CT (PET-CT) was then performed and showed multifocal malignant lesions in the stomach, pancreas, gallbladder, bone, bilateral supraclavicular fossa, anterior mediastinum, bilateral axillary and retroperitoneal

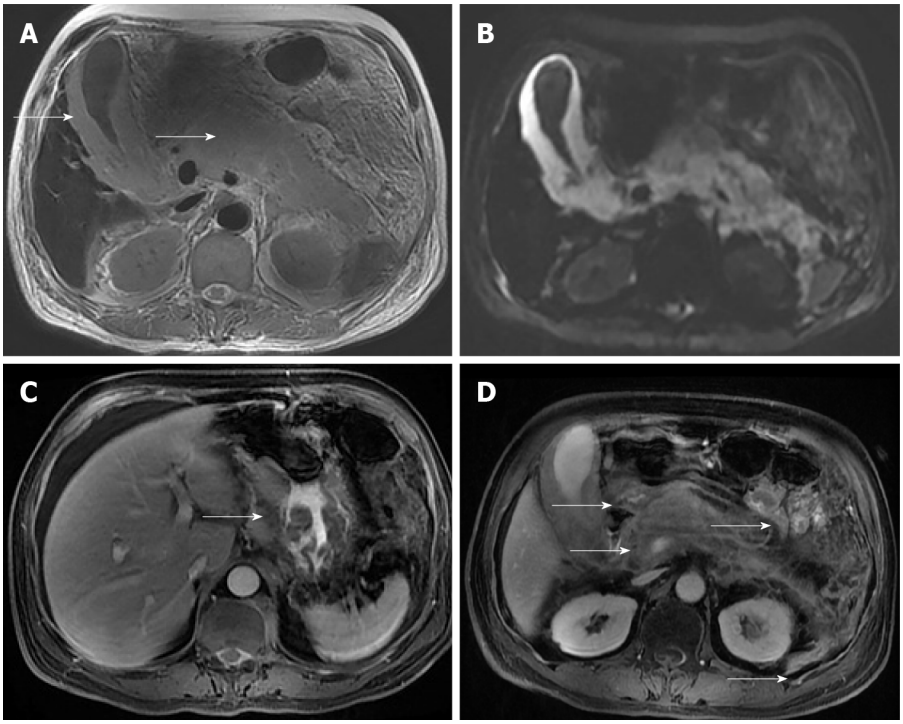

Figure 1 Magnetic resonance imaging of the abdomen at diagnosis. A: Axial T2-weighted magnetic resonance imaging (MRI) demonstrates homogeneous, hyperintense lesion in the whole pancreas and a markedly swollen gallbladder (arrows); B: Diffusion-weighted MRI shows abnormal hyperintensity in gall bladder wall and pancreas; C: Axial contrast-enhanced T1-weighted MRI shows the abnormal thickened lesions of the gastric wall (arrows), which display contrast enhancement in a × homogeneous fashion; D: Axial contrast-enhanced T1-weighted MRI shows the swollen gallbladder and multiple enlarged retroperitoneal lymph nodes (arrows), which display contrast enhancement in a homogeneous fashion.

lymph nodes (Figure 3), indicating multiple metastases of malignant tumors. Gastroduodenal endoscopy revealed massive involvement of the gastric body and duodenum with tumors (Figure 4). Histology and immunohistochemistry of gastric biopsies were suggestive of BL (Figure 5).

FINAL DIAGNOSIS

The histological findings, immunophenotype of the biopsies, and radiological findings were consistent with BL involving the stomach, pancreas and vertebral column. However, the primary lesion of BL is unclear. Because the patient had no symptoms of fever or weight loss, it was classified as group A. Due to the lack of bone marrow aspirate and trephine biopsy, we could not confirm the accuracy of the stage classification of BL in this case. Curiously, this patient presented with acute pancreatitis as the initial manifestation. One possible explanation for the presentation of acute pancreatitis is that the main pancreatic duct was obstructed by the substantial mass. Obstruction of the pancreatic orifice may impair the outflow of pancreatic juice and eventually induce pancreatitis.

TREATMENT

After admission to our department, this patient received short-term fasting, acid suppression, pancreatic enzyme suppression and fluid replacement for acute pancreatitis. Due to suspicion of necrotic pancreatitis, sulbactam sodium/cefoperazone sodium (3 g/d) was administered IV for one week.

Unfortunately, this patient developed sudden onset of aconuresis and paraplegia. According to the neurology consultation, acute myelitis was suspected. To inhibit the inflammatory response and block the antibodies, high doses of glucocorticoids and gamma globulin were applied for three days. Nevertheless, the efficacy of these treatments appeared poor. Concerning the high cost and potential side effects of these treatments, glucocorticoid and gamma globulin treatment was abandoned. Based on

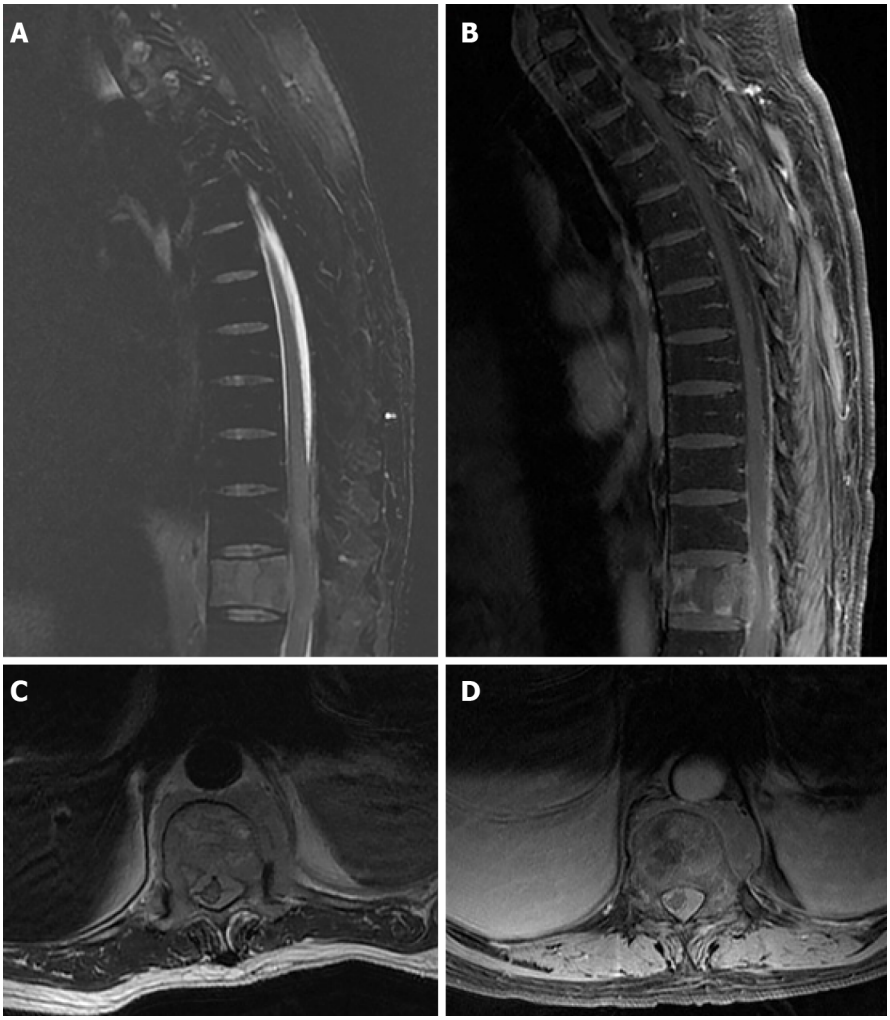

Figure 2 Magnetic resonance imaging of the thoracic and lumbar vertebrae at diagnosis. A: Sagittal T2-weighted magnetic resonance imaging (MRI) shows epidural mass at the centrum and left posterolateral aspect of the spinal cord at the T9 to T12 levels, resulting in severe cord compression; B: Sagittal contrast-enhanced T1-weighted MRI shows the lesions displaying contrast enhancement in a heterogeneous fashion; C: Axial T2-weighted MRI shows that epidural mass involves the centrum and left posterolateral aspect of the spinal cord; D: Axial contrast-enhanced T1-weighted MRI shows the lesions displaying contrast enhancement in a heterogeneous fashion.

the indication for further imaging tests, this patient was diagnosed with gastric BL *via* endoscopic biopsy. Accordingly, a chemotherapy combination of cyclophosphamide, adriamycin, vincristine and prednisolone (CHOP) was recommended for the patient, but he refused the chemotherapeutic treatment.

OUTCOME AND FOLLOW-UP

One week after diagnosis and refusal of chemotherapy, the patient died of upper gastro-intestinal hemorrhage.

DISCUSSION

The incidence of BL is extremely low, and the clinical symptoms are atypical. Thus, we need to raise awareness of BL and reduce the misdiagnosis rates. BL was first described in 1958 by a British surgeon named Denis Burkitt as a sarcoma involving the jaw in African children with characteristic symptoms[1]. There has been some improvement in the understanding of its epidemiological diagnosis and treatment in the ensuing half century. In this article, we report the 11th case of BL involving the stomach, pancreas and spinal cord diagnosed based on the radiological findings and immunophenotype of the biopsies. The clinical features of 10 previous cases of BL

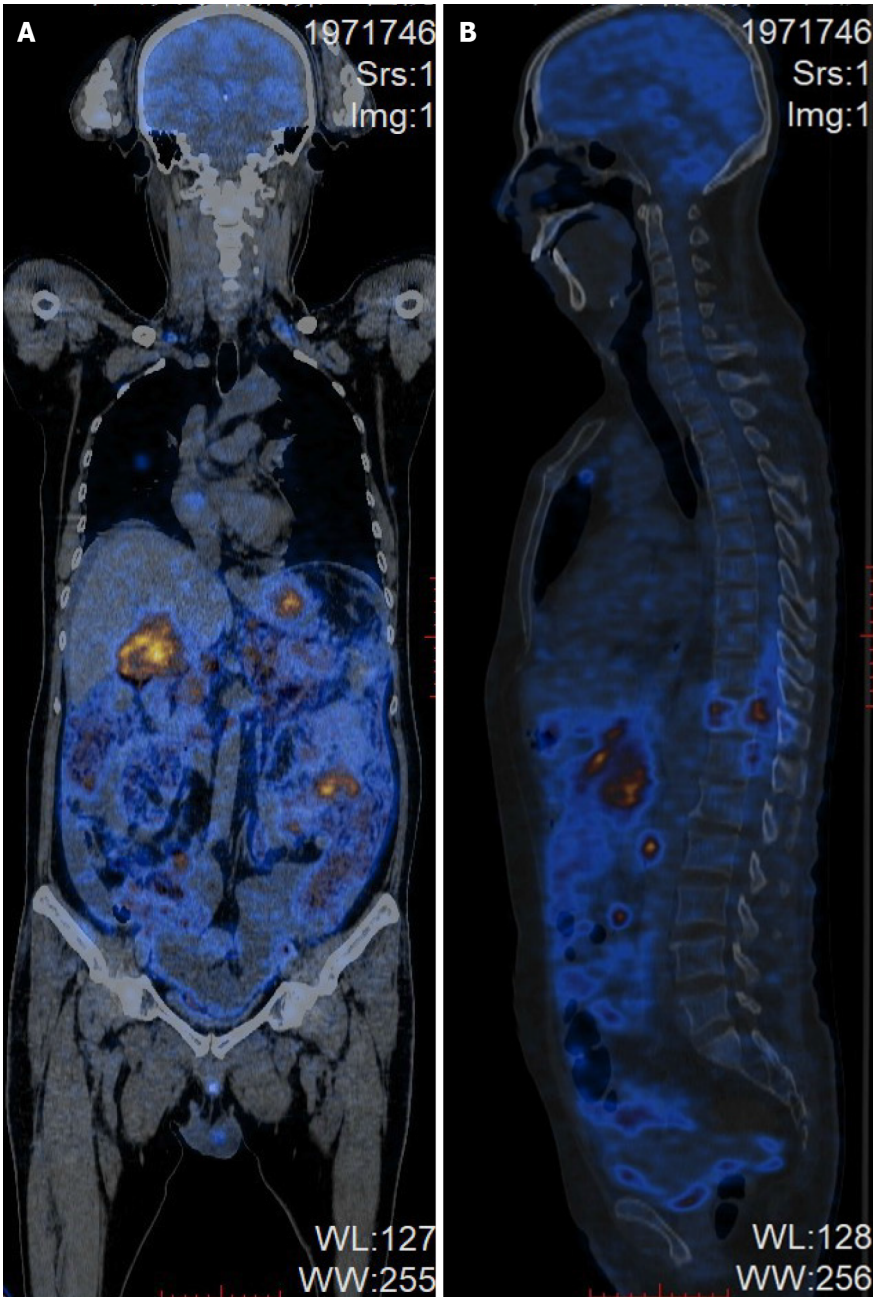

Figure 3 Positron emission tomography-computed tomography of the whole body at diagnosis. A: Coronal images; B: Sagittal images.

involving the stomach, pancreas or spinal cord are summarized in Table 1[8-17].

Among the previous cases, nine cases were reported in foreign countries, while only one patient came from China. From our review of the literature, a clear male predominance (70%) can be established. The ages of the patients range from 9 to 69 years, with a median age of 23 years. The initial symptoms, including abdominal distension, abdominal pain, lumbago, weakness in the lower extremities, fulminant hematemesis and progressive weight loss, are atypical. Regarding the detailed treatment protocols, seven patients received chemotherapy. Only one patient received palliative radiation treatment due to severe spinal cord involvement. Among the four patients who underwent surgical intervention, one patient underwent surgery for intraspinal decompression and mass separation, and the other three patients underwent distal or total gastrectomy. The outcome and follow-up of BL were reported in a total of eight cases. Regrettably, only a 9-year-old patient remained in clinical remission with completed chemotherapy, and no treatment-related sequelae 4 years were observed from initial diagnosis. Severe complications, including gastric perforation, sepsis and bacteremia, are always derived from intensive chemotherapy. Due to lymphoma recurrence or severe complications associated with chemotherapy, the other seven patients died within 6 mo of diagnosis.

Table 1 Clinical features of 10 previous cases of Burkitt lymphoma involving stomach, pancreas or spinal cord

Author	Age	Gender	Initial symptom	Affecting area	Biopsy area	Immunohistochemical studies	EB	HIV	Treatment	Prognosis
Kim *et al*[8]	69	Female	Low back pain radiating down to the right leg	Spinal cord at the L2 to L4 levels, intestine, live, bone and left supraclavicular lymph node	A posterolateral extradural mass lesion between L2 and L3	CD20 (+), CD79a (+), BCL-6 (+), CD10 (+), BCL-2 (-)	+	NA	NA	NA
Seo *et al*[9]	40	Male	Progressive pain and weakness in lower extremities	Spinal cord at the T2 to T4 levels, liver	An intraspinal extramedullary mass from T2 to T4, liver	CD20 (+), CD45RO (-)	NA	+	Chemotherapy and radiation therapy with HAART after surgery for intraspinal decompression and mass separation. Radiation	Died by massive pulmonary thromboembolism at 13 wk postoperatively
Chieng *et al*[10]	9	Male	Progressive pallor, peripheral oedema and respiratory distress	Stomach	Gastric body mass	CD20 (+), CD10 (+) and CD43 (+)	NA	NA	Induction chemotherapy with COP. Further chemotherapy included two courses of COPADAM followed by two courses of CYM and double intrathecal chemotherapy of methotrexate and hydrocortisone	Remains in clinical remission with complete resolution of the protein-losing enteropathy and no treatment related sequelae 4 yr from initial diagnosis
Bolandparvaz *et al*[11]	21	Male	Abdominal pain	Stomach	A huge mass in greater curvature of the stomach	NA	NA	NA	Total gastrectomy and roux-en-y esophagojejunostomy, chemotherapy was given for the patient 1 wk later without any other complication	NA
Gurzu *et al*[12]	60	Female	Fulminant hematemesis, recurring melena, epigastric pain, inappetence, and weight loss	Stomach	A huge mass in the antrum and posterior wall of the gastric body	CD20 (+), CD79a (+), BCL-6 (+), CD10 (+), Ki-67 (100%+), CD3 (-), CD5 (-), CD23 (-), TdT (-), bcl-2 (-), and Cyclin D1 (-)	-	NA	Distal gastrectomy	Died ten days after surgical intervention
Krugmann *et al*[13]	28	Male	Hematemesis and increasing abdominal pain	Stomach	A huge mass in the middle third of the stomach	CD20 (+), CD10 (+), BCL-6 (+), Ki-67 (95%+), CD3 (-), CD5 (-), CD23 (-), Cyclin D1 (-), BCL-2 (-) and TdT (-)	-	NA	Billroth-II surgical resection	Died due to lymphoma recurrence four months after onset
Liao *et al*[14]	26	Male	Fulminant hematemesis, abdominal pain	Stomach	A mass in the body and antrum of the stomach	CD20 (+), CD10 (+), BCL-6 (+), MUM-1 (-), CD30 (-)	NA	NA	Induction chemotherapy with two courses of R-ECHOP. Further chemotherapy included two courses of R-hyper CVAD followed by five courses of intrathecal prophylactic injection of chemotherapy drugs	Lymphoma recurrence six months after onset
Sağlam *et al*[15]	20	Male	Weight loss, back pain, mandible numbness, night sweats, and poor exercise tolerance	The body of the pancreas	A mass in the body of the pancreas	NA	NA	NA	Doxorubicin based combination chemotherapy	Died from sepsis during the second month of chemotherapy
Nistala *et al*[16]	21	Male	Jaundice, increasing swelling in	The head of the pancreas, cystic duct,	The first and second parts of	CD20 (+), CD10 (+), BCL-6 (+), CD5 (-), Mib-1 (99%+)	NA	NA	Two cycles of CHOP followed by hyper CVAD regimen as	NA

			the epigastric region	portal vein and hepatic artery, duodenum	duodenum				definitive therapy	
Konjeti *et al* [17]	68	Female	Belching, abdominal bloating and weight loss	The head of the pancreas, central hepatic duct and portal vein	The pancreatic head mass	CD20 (+), CD10 (+), C-myc (+), BCL-6 (+), CD3 (-), TdT (-), BCL-2 (-), Ki-67 (> 90%+)	NA	NA	Two cycles of chemotherapy regimen consisting of etoposide, prednisone, vincristine (Oncovin), and doxorubicin hydrochloride (Hydroxydaunorubicin hydrochloride)	Die due to the sepsis and bacteremia

EB: Epstein-Barr virus; HIV: Human Immunodeficiency Virus; HAAART: Highly active antiretroviral therapy; COP: Cyclophosphamide, vincristine and prednisolone; COPADAM: Cyclophosphamide, vincristine, prednisone, cytarabine, doxorubicin and methotrexate; CYM: Cytarabine and methotrexate; R-ECHOP: Rituximab, etoposide, cyclophosphamide, doxorubicin, vincristine and prednisone; CVAD: Cyclophosphamide, vincristine, doxorubicin, dexamethasone; CHOP: Cyclophosphamide, doxorubicin, vincristine and prednisolone; NA: Not available.

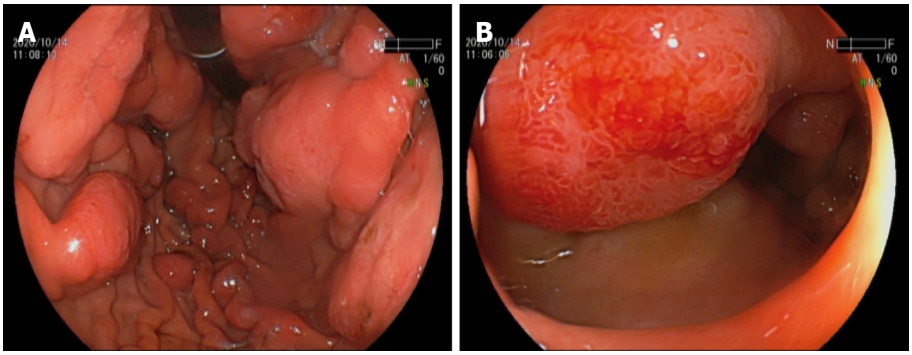

Figure 4 Gastric endoscopy. A: Multiple large (2 to 3 cm in diameter) raised ulcerated tumors involving both the greater and smaller curvatures of the gastric body; B: Numerous smaller tumors involving the anterior wall of the duodenal bulb and the second part of duodenum.

Three variants of BL have been described worldwide: Endemic, sporadic, and immunodeficiency-associated. Among the three subtypes, sporadic BL is regarded as the most common type[18]. The clinical features of BL are variable. In endemic BL, patients tend to present with jaw and other facial diseases. Cases of sporadic BL with an intraperitoneal mass as the initial manifestations are more common. Additionally, the clinical course of sporadic BL is usually aggressive, with frequent extranodal and central nervous system (CNS) involvement and an overall poor prognosis. BL with extranodal involvement usually occurs in the gastrointestinal tract (50%) and head and neck (25%). According to the statistics, CNS involvement is recognized in 13%-17% of all cases of BL[19]. This kind of cancer cell proliferates rather rapidly, with a doubling time of approximately 24 h, and the Ki-67 proliferation index tends to be 90%-100%. Clinically, a blood test usually reveals markedly elevated LDH and uric acid levels in the early stages, indicating a high tumor burden[20]. Herein, we report a case of gastric BL in an adult patient presenting with paraplegia and acute pancreatitis. Similarly, the auxiliary examination in this case also showed markedly elevated LDH and uric acid levels at admission. During hospitalization, this patient developed acute compression of the spinal cord. Abdominal CT at admission revealed no apparent abnormal findings except for the indication of acute pancreatitis. Unexpectedly, MRI of the abdomen and lumbar spine at the 14th d after admission indicated multisite metastasis in gastric malignancy, including in the pancreas, bone, bilateral supraclavicular fossa, anterior mediastinum, bilateral axillary and retroperitoneal lymph nodes. Finally, gastroduodenal endoscopy revealed massive involvement of the gastric body and duodenum with BL. Dawson's criteria are used to label primary gastrointestinal lymphoma, including absence of peripheral lymphadenopathy at the time of presentation, lack of mediastinal lymph node enlargement, normal total and differential white blood cell count, predominance of bowel lesion at the time of laparotomy with only lymph nodes obviously affected in the immediate vicinity and no lymphoma involved in the liver and spleen[21]. In this case, the patient had leukocytosis and multiple enlarged retroperitoneal lymph nodes and therefore did not fulfil the criteria. Hence, it was not a case of primary gastric lymphoma and the primary lesion of BL is unclear. This case reminds us that malignant tumors can originate from hematopoietic

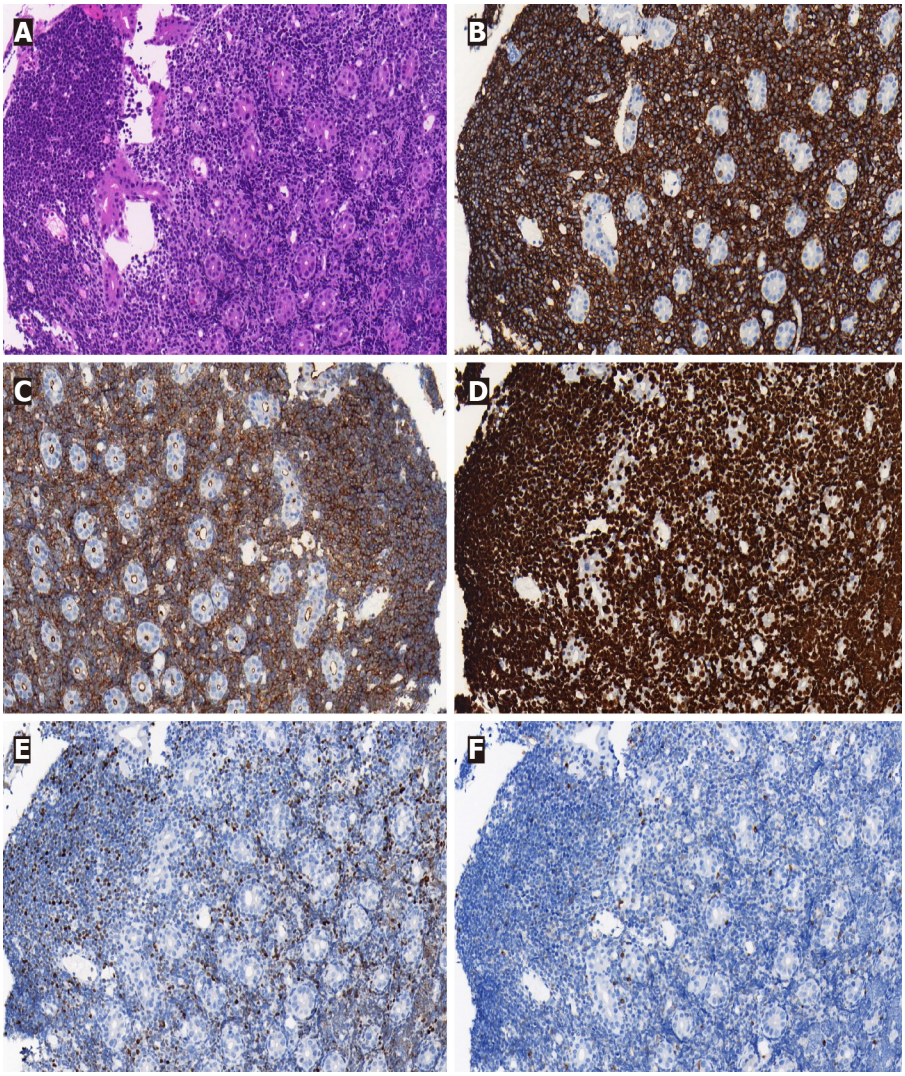

Figure 5 Histology and immunohistochemistry of gastric biopsies (× 200). A: Haematoxylin and eosin staining showed a characteristic "starry sky" appearance; B: Immunohistochemical staining was positive for CD20; C: Immunohistochemical staining was positive for CD10; D: Immunohistochemical staining was positive for Ki-67 (> 90% +); E: Immunohistochemical staining was positive for BCL-6; F: Immunohistochemical staining was negative for BCL-2.

malignancies, especially BL, and that this needs to be taken into consideration when there is abnormally rapid progression of the disease and when there are numerous affected areas or markedly elevated indicators of tumor burden.

Regarding the imaging evaluation of BL, CT scanning and three-dimensional reconstruction are more useful for accurately displaying bone destruction. When spinal cord involvement is suspected for clinical reasons, the preferred choice is MRI since it outperforms CT in depicting associated soft tissues. Additionally, diffusion-weighted MRI is a favorable diagnostic tool in oncologic imaging since it can reflect cellularity and proliferative activity in most malignancies. It is acknowledged that most malignancies are characterized by sustained proliferation, contributing to a high cellular density. More specifically, on diffusion-weighted MRI, BL demonstrates a markedly high signal intensity due to the relative restriction of water associated with high cellular density[22,23]. Since repeated serial imaging is essential for evaluating disease progression, MRI is also superior to CT due to its lack of ionizing radiation. For superior staging and assessment of the treatment response, PET/CT is a better choice since it can evaluate the functional status of abnormally hypermetabolic tissues throughout the whole body[24].

Histologically, the tumor cells of BL are medium-sized with an abundant, basophilic cytoplasm and display the typical "starry sky" pattern. The tumor cells are positive for BCL-6, CD19, CD20, CD22, CD10 and CD79a but negative for CD3, CD5, CD23 and TdT[25]. BL is characterized by the t (8; 14) (q24; q32) translocation of the c-myc and IgH genes, resulting in IgH-myc fusion, which can be detected by molecular analysis *via* fluorescence in situ hybridization. In our case, the tumor cells were negative for

creatine kinase and CD3, indicating that the tumor was not derived from the epithelium or T-cells. Additionally, the tumor cells were positive for CD20, CD79a, CD10, and BCL-6, suggesting germinal center-derived B cells. Combined with the high Ki67 index, the diagnosis of BL can be established.

Systemic chemotherapy is the preferred choice for the treatment of BL. Additionally, conventional radiotherapy, surgery, or a combination of both are recommended as the standard treatment unless severe compression of vital organs by lymphoma is observed[26]. Currently, most treatment protocols for adults are based on pediatric clinical trials. At present, most classical chemotherapy regimens show good efficacy and safety in children and relatively young patients. However, the prognosis of adult patients is poor due to their low response rate and severe treatment-related toxicity. Chemotherapy regimens, including CHOP, hyper-cyclophosphamide, vincristine, epirubicin, dexamethasone, etoposide, prednisone, vincristine, cyclophosphamide, epirubicin and cyclophosphamide, epirubicin, doxorubicin, vincristine, high-dose methotrexate/isophosphamide, cytarabine and etoposide, are still the backbone of therapeutic strategies for BL. Rituximab is an anti-CD20 chimeric antibody that acts by depleting CD20-positive B lymphocytes[27]. It has been reported that common chemotherapy regimens combined with rituximab can significantly improve the 3-year overall survival rate of BL patients (83% *vs* 70%)[28]. Treatment with prophylactic intrathecal methotrexate or cytarabine can lower the incidence of CNS relapse. Hence, it is regarded as a part of the first-line treatment option for BL[29,30]. In this case, the patient refused the chemotherapeutic treatment and died of upper gastrointestinal hemorrhage one week after diagnosis.

CONCLUSION

The incidence of BL is extremely low, and the clinical symptoms are atypical, contributing to the high misdiagnosis rate and poor prognosis. Clinically, malignant tumors originating in hematopoietic malignancies, especially BL, need to be taken into consideration if there is abnormally rapid progression of the disease and if there are numerous affected areas or markedly elevated indicators of tumor burden. CT or MRI could be an option for the detection of BL, while PET/CT is essential for the staging of BL. Histological assessment is indispensable for a definite diagnosis. Regarding the treatment of BL, chemotherapy is the preferred choice. Prophylactic intrathecal methotrexate or cytarabine is also recommended to lower the incidence of CNS relapse.

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