A case report of primary central nervous system lymphoma with intestinal obstruction as the initial symptom

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Abstract

Rationale: Primary central nervous system lymphoma (PCNSL) with initial manifestations of constipation and intestinal obstruction (IO) is rare.

Patient concerns: A 50-year-old Chinese male patient was admitted to the gastroenterology department due to constipation and abdominal distention for 8 days. He had experienced intermittent back pain for 3 years prior to admission. Based on abdominal radiography, he was initially diagnosed with IO and treated with meal restriction and enemas. However, his symptoms worsened, and progressive lower limb weakness was observed.

Diagnoses: A colonoscopy was inconclusive due to the IO. Computed tomography and magnetic resonance imaging revealed space-occupying lesions near centra 9–11 of the thoracic vertebrae. The patient underwent spinal decompression surgery, and pathologic examination led to a diagnosis of PCNSL (diffuse large B cell lymphoma).

Outcomes: The symptoms of the IO improved postoperatively, and the patient partially recovered his lower limb muscle strength. He returned to his homeland for chemotherapy.

Lessons: IO can be an initial, unspecific symptom of spinal cord compression in patients with PCNSL.

Abbreviations: CNS = central nervous system, CT = computed tomography, DLBCL = diffuse large B cell lymphoma, IO = intestinal obstruction, NHL = non-Hodgkin lymphoma, PCNSL = primary central nervous system lymphoma.

Keywords: intestinal obstruction, lymphoma, non-Hodgkin, spinal cord neoplasms

1. Introduction

Intestinal obstruction (IO) is a common acute digestive disorder. Common causes include mechanical (tumors, volvulus, or surgical complications), dynamic (paralysis or cramps), and ischemic factors. Primary central nervous system lymphoma (PCNSL) is an extremely rare form of malignant lymphoma that only occurs in <3% of central nervous system (CNS) tumors. The coexistence of these conditions is rare. Herein, we report a case of a male patient who initially demonstrated manifestations of IO with a final diagnosis of PCNSL.

2. Case report

A 50-year-old male Chinese patient was admitted due to a lack of defecation for 8 days and with symptoms of abdominal distention. His medical history included a trauma-induced fracture of the 3rd transverse process of the lumbar vertebrae 3 years earlier, after which he intermittently suffered from backache. He received physical therapy (cupping) 3 days prior to admission, which effectively alleviated the pain. His older brother had died from lung cancer. The patient seemed well-nourished. A chest radiography did not show any remarkable findings. Based on these test results, incomplete IO was considered. After admission, the patient continued suffering from abdominal distension as well as a lack of autonomous defecation and flatus although he received conventional therapy, including meal restriction and enemas. Three days after admission, progressive lower limb weakness was observed; the results of the straight leg raise of the left and right legs were 45° (+) and 60° (+), respectively, and that of the reinforced straight leg raise of both
legs was (+), with reduction of lower limb muscle strength (left leg: grade II; right leg: grade III) and decreased muscle tension. An axial abdominal computed tomography (CT) scan showed suspicious eccentric thickening of the colon walls (Fig. 2), but no enlarged lymph nodes were seen. No remarkable lesion was found in a subsequent colonoscopy; however, the preoperative preparation was unsatisfactory due to the IO (Fig. 3). Therefore, the CT images were reevaluated with an expanded scope; they revealed a space-occupying lesion to the right of centrums 9–11 of the thoracic vertebrae (Fig. 4). Magnetic resonance imaging of the thoracic spine scan indicated an abnormal signal in centrum 10 of the thoracic vertebrae and space-occupying lesions near centrums 9–11 as well as in the spinal canal (Figs. 5–7). Accordingly, it was assumed that the IO was caused by the space-occupying lesions in the centrums compressing the corresponding spinal cord sections. The patient then underwent a resection of the lesions near the centrums. A pathological examination revealed masses in the centrums and right pedicle of the vertebral arch of centrum 10 of the thoracic vertebrae as well as non-Hodgkin lymphoma (NHL) of the diffuse large B cell lymphoma (DLBCL) type (also, germinal-center type). Immunohistochemical analyses showed: AE1/AE3 (−), Bcl-2 (−), Bcl-6 (+), CD10 (−), CD20 (+), CD3 (marginally +), CD30 (Ki-1) (−), CD31 (−), CD34 (−), CD5 (marginally +), HMB45 (−), Ki-67 (index, 40%), Mum-1 (−), and PAX-5 (+) (Fig. 8). The final diagnosis was PCNSL of the DLBCL type, in combination with an IO caused by spinal compression. After the surgery, the IO symptoms were partially relieved, and the patient recovered some of the muscle strength in his lower limbs. He returned to his homeland for chemotherapy and lost to follow-up.
Important clues and steps for diagnosis are summarized on Table 1.

3. Discussion

Spinal tumors are 10 to 15 times less common than primary intracranial tumors and represent 2% to 4% of all primary tumors of the CNS.\[^{2}\] They are classified as primary versus secondary tumors, and categorized as intramedullary, intradural, extramedullary, or extradural. The most common invasion sites of spinal tumors include the midthoracic (69%), lumbar (27%), and cervical spine (4%).\[^{3}\] PCNSL, defined as NHL that can invade not only the brain but also the leptomeninges, eyes, and spinal cord, is extremely rare.\[^{4}\] Spinal tumors can lead to spinal cord compression and result in a series of unspecific symptoms, such as IO in this case.

PCNSL has specific imaging characteristics. On CT scans, tumor lesions mainly appear as shadows of isodensity or slightly elevated density; on magnetic resonance imaging, they show low or isosignal intensities on T1-WI images and high signal intensities on rho-WI and T2-WI images; gadolinium-diethylenetriamine penta-acetic acid was reported to improve the quality of T1-WI images by markedly enhancing the tumor.\[^{5}\] Molecular imaging by positron emission tomography with 18F-fluoro-2-deoxy-glucose is helpful for diagnosis, identification of the metabolically active tumor compartment, and prediction of the treatment response.\[^{6}\]

Extranodal NHL accounts for 30% to 40% of all NHL cases in different regions.\[^{7}\] Spinal tumors are one of the rarest forms of NHL and are only observed in <5% of NHL cases.\[^{8}\] Leptomeningeal, epidural, and brain metastases are the most common neurologic complications of PCNSL, all of which are
associated with a poor prognosis. DLBCL is the most common type of NHL worldwide and the main type of PCNSL.\(^9\)

Immunohistochemically, the large lymphoid cells were CD20 (+) and CD3 (−). Spinal DLBCL can be categorized into the germinal center B cell [CD10 (+) or CD10 (−)/BCL-6 (+)/MUM1 (−)] and non-germinal center B cell [CD10 (−)/BCL-6 (−) or CD10 (−)/BCL-6 (+)/MUM1 (−)] types. It is known to initially involve the paraspinal soft tissues, such as the paravertebral ganglion or epidural lymphoid tissue, followed by invasion around the spinal cord via the vertebral foramen without destroying bony structures. This is consistent with the findings in our case.

It has been reported that back pain[4,10] and signs of lower body motor neuron involvement[11] were the most common symptoms in patients with spinal cord compression. In our case, the patient showed similar symptoms. Although the physical examination on admission was normal, he had complained of intermittent back pain for years, which had been assumed to be a sequelae of a past transverse process fracture; however, the abnormal sensation in his lower limbs on admission was unexplained. Unfortunately, we missed the clues until the spinal cord compression syndrome was obvious. In our case, the first symptom that was indicative of a spinal cord compression was the reduction of muscle strength in the lower limbs, which has been reported as a regular manifestation of spinal cord injuries.[12] Due to the high mortality rate of surgery, it was debated whether surgical treatment should be performed.[13,14] The patient survived the surgical decompression, and future chemotherapy as well as radiotherapy is considered beneficial.[15]

In our case, the cause of the IO was revealed through surgery and pathological examination. In summary, signs of spinal cord compression, including weakness of the lower limbs and unexplained back pain prior to admission, were observed but inadequate attention was given to these symptoms at the time. Spinal cord injury accompanied by sensory disturbance and dyskinesia of the lower limbs were useful to distinguish this case from other cases of IO. Innervation of the stomach, small intestine, colon, and rectum is mainly formed by the sympathetic nerves of the thoracic (T) 6 to lumbar (L) 3 spinal cord as well as the parasympathetic nerves of the dorsal nucleus of the vagus nerve and sacral parasympathetic nucleus of the sacral (S) 2 to S4 spinal cord. In this case, the spinal cord injury of the thoracic segments impaired both, sympathetic and parasympathetic nerves, and reduced gastrointestinal motility, leading to IO.

Table 1

| Time points   | Event                                                                 |
|--------------|----------------------------------------------------------------------|
| 8 d prior to admission  | Lack of defecation                                                   |
| 3 d prior to admission  | Backpain (alleviated by physical therapy)                            |
| On admission          | Symptoms: abdominal distention, cold limbs                           |
|                        | Primary examinations: abdominal radiography, laboratory tests, physical exams |
|                        | Diagnosis: incomplete intestinal obstruction (?)                     |
| Day 3                 | No symptom improvement after enemas                                  |
|                        | Abdominal CT revealed suspicious thickening of the colon walls       |
| Day 4                 | No lesion found by colonoscopy                                       |
| Day 5                 | CT revealed lesion near thoracic vertebrae (T) 9–11                  |
| Day 6                 | MRI revealed lesion near T 9–11, spinal canal invaded                |
| Day 12                | Surgical decompression                                              |
| Day 14                | Clinical diagnosis after surgery: spinal tumor                        |
|                        | Immunohistochemical diagnosis: DLBCL                                  |
|                        | Final diagnosis: PCNSL of the DLBCL type                             |

CT=computed tomography, DLBL=diffuse large B cell lymphoma, MRI=magnetic resonance imaging, PCNSL=primary central nervous system lymphoma.
Moreover, due to the loss of anorectal coordination resulting from forced defecation due to the spinal cord injury, the patient exhibited a lack of autonomous defecation, another reason for his constipation.

The findings of the current case can inspire gastroenterologists when seeking uncommon causes of IO, which is a common disorder encountered in daily clinical practice. A diagnosis of the functional disorder should be based on an understanding of the underlying pathophysiology. PCNSL should be considered as an atypical cause of IO.

4. Informed consent
Written informed consent for the publication of this case report and the associated images was obtained from the patient before submission.

5. Author contributions
X.K. Li was the attending physician of this case and primarily drafted the manuscript; S.Qi and J.Gao were consulting physicians; Y.T. Jiao was the intern physician, he was involved in the diagnosis and treatment; S.Qi also helped with conducting the literature review; and H.B. Du was the chief physician in charge of this case.

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