Sacral chordoma incidentally discovered in a patient with acute gastrointestinal perforation during his first visit to the hospital

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ABSTRACT

Introduction: Chordoma is a rare recurrent locally malignant bone tumor. Case Report: A 41-year-old male patient presented to Emergency Medicine Department (EMD) with features of an acute abdomen. His chest X-ray showed air under the diaphragm, and the computed tomography (CT) abdomen revealed a soft tissue mass suggestive of a sacral chordoma. A few months before his EMD visit, he has received multiple courses of nonsteroidal anti-inflammatory drugs (NSAIDs) for back pain. The patient was operated upon as an emergency for the gastrointestinal perforation. He was operated upon electively for the mass later on. Histopathological exam after total resection at other center confirmed the diagnosis of chordoma. Conclusion: After stabilization of patient and pain control, further history taking will be valuable to know the primary cause for the acute abdomen presentation.

Keywords: Acute surgical abdomen, Gastrointestinal perforation, NSAIDs, Sacral chordoma

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INTRODUCTION

Chordomas are rare bone tumors with local invasion and frequent recurrences. These are relatively radioresistant. The tumors originate from the primitive notochordal remnants that run the entire length of the spine [1]. Chordomas may occur at the skull base (35%), sacral regions (50%), and cervical vertebrae (10%) [2]. The annual incidence of chordoma is one in million in the United States, and one in 800,000 in the United Kingdom [3, 4]. The most frequent local effect of sacral chordoma is pain that causes nocturnal awakening. Patients often describe this pain as a gnawing and unremitting [5]. In one study, the 10-year tumor survival rate of sacral chordoma was 46% [6]. In most cases, complete surgical resection followed by radiation therapy offers the best long-term control [7]. The latest guidelines for management of chordoma were published in Lancet in 2015 [8].

CASE REPORT

A 41-year-old male patient with no past history of medical illness presented to EMD with a severe sudden
upper abdominal pain which started two hours before his arrival. The pain was accompanied by vomiting, but without fever, chills, hematemesis or melena, and no urinary complaint. The patient graded pain as 10 out of 10 and he looked sick. His blood pressure was 128/75 mmHg, pulse rate was 75/min, temperature was 37.3 °C, and oxygen saturation was 97% at room air.

His abdominal examination revealed generalized severe tenderness, distension, and guarding mainly at the upper abdomen. His chest and heart examination were unremarkable. His plain chest X-ray revealed free air under the right diaphragm (black arrows) (Figure 1).

The provisional diagnosis was an acute abdomen most probably due to gastrointestinal perforation. The patient received intravenous (IV) morphine sulfate 5 mg, ondansetron IV 8 mg, 0.9% sodium chloride IV 1 L, IV pantoprazole 40 mg, and piperacillin/tazobactam 4.5 g IV. After pain control partially achieved, a more detailed history could be taken.

During the previous six months, the patient was suffering from recurrent episodes of low back pain which worsen at night. He was treated with different courses of NSAIDs. The patient suffered from previous occasional heartburn, but no abdominal pain, hematemesis, or melena before his EMD visit. His investigations at our EMD revealed white blood cells (WBCs) were 6220 cell/mm³, hemoglobin was 14.7 g/dL, platelets were 190,000/mm³, and differential cell count was within normal.

The patient’s blood urea nitrogen, serum creatinine, electrolytes, liver function tests, serum amylase, serum lipase, C reactive protein, and lactic acid were within a normal range.

Computed tomography abdomen and pelvis sagittal view with IV contrast showed free intraperitoneal air anterior to the stomach (upper black arrow), and a large sacral soft tissue mass which displaced and compressed the urinary bladder. This mass showed aggressive features of erosions of the sacral vertebrae (lower white arrow) (Figure 2).

The patient was assessed by a surgical team who had the same provisional diagnosis of acute abdomen due to gastrointestinal perforation which is most probably due to NSAIDs usage.

Diagnosis and radiological findings were explained to the patient. This high possibility of sacral chordoma tumor was clarified including the need for diagnosis confirmation and management plan that may not be advisable during the same emergency operation. Spine surgeon and neurosurgeon were consulted preoperatively and advised not to approach the sacral tumor for reception or even biopsy, to avoid contaminating the retroperitoneal region and in case of malignancy spreading the cells in the peritoneal cavity.

Operative findings: A mini-laparotomy showed a 5 × 5 mm prepyloric anterior gastric perforation with turbid intra-abdominal fluid. On exploration, we identified the retroperitoneal pelvic mass, pushing anteriorly the bladder and rectum. No liver masses or omental deposits were noted. Procedure: Wedge biopsy of the anterior surface of gastric perforation, suture repair with interrupted Vicryl 3/0 sutures and omentoplasty. Irrigation, suction, and drainage were performed.

Figure 1: Plain chest X-ray revealed free air under the right diaphragm (black arrows).

Figure 2: CT abdomen and pelvis sagittal view with IV contrast showed free intraperitoneal air anterior to the stomach (upper black arrow), and a large sacral soft tissue mass which displaced and compressed the urinary bladder.
Postoperatively, the patient was hemodynamically stable, admitted to the surgical ward. At day 2 postoperative, workup-up was completed with a magnetic resonance imaging (MRI) abdomen and pelvis with contrast which showed a large sacral soft tissue heterogeneous mass $16 \times 14.3$ cm (MRI abdomen sagittal view) (Figure 3).

Small areas of moderate to high signal intensity suggestive of proteinaceous or blood products (white arrow) and findings were in favor of sacral chordoma (MRI abdomen and pelvis axial view) (Figure 4). The patient had an uneventful recovery, and was discharged on day 4 after surgery.

Second operation: The patient decided to have the surgery of the pelvic mass, later on, at another hospital. Three months later, he was operated upon electively, at another medical center, where his sacral chordoma was totally resected and followed by radiotherapy. Histopathology studies with hematoxylin and eosin stain revealed a tumor with lobules of neoplastic cells (yellow arrow), and interspersed myxoid areas (blue arrow) (Figure 5A).

High power histopathology slide revealed the neoplastic cells (physaliphorous cells) had abundant clear vacuolated/bubbly cytoplasm (yellow arrow), with large pleomorphic nuclei (blue arrow) (Figure 5B). Histopathological findings confirmed the diagnosis of chordoma.

DISCUSSION

The acute abdomen accounts for up to 40% of all emergency-surgical hospital admissions and considered in the differential in millions of visits to Emergency Department annually of abdominal pain in the United States and the United Kingdom [9, 10].

A high percentage of these cases are due to gastrointestinal perforation [9]. Mortality due to secondary peritonitis is still relatively high 30–50%, despite advances in antibiotics, surgical techniques, radiographic imaging, and resuscitation therapy [11].

Perforation is complicated in 2–10% of peptic ulcer patients [12]. The duodenal, antral, and gastric body ulcers account for 60%, 20%, and 20% of perforations due to peptic ulcer disease (PUD), respectively [13]. In a systemic review of studies published before 1997 and during period 1997–2008, mortality rate in all cases due to upper gastrointestinal bleed or perforation fell from
11.6% in pre-1997 to 7.4% in 1997–2008. Mortality rate of upper gastrointestinal bleed or perforation in patients taking NSAIDs increases from 14.7% in pre-1997 to 20.9% in 1997–2008 [14].

Relative risk (RR) of upper gastrointestinal bleed/perforation varies among individuals of NSAIDs. Celecoxib has the lowest RR of bleed/perforation 1.42, while piroxicam and ketorolac have the highest RR 8.00 and 14.54, respectively [15].

The median age at diagnosis of chordoma is 60 years, with skull base affecting younger patients than the spinal cord injury patients. The median time from initial symptoms till diagnosis is longer than two years, with clinical presentation at onset varies according to site [8].

The MRI is the recommended modality for primary tumor assessment because it allows for delineation of the different soft tissue, components of the tumor, and adjacent structures. After injection of gadolinium-based contrast most chordomas show minimum to moderate heterogeneous enhancement [16].

Recommended management of sacral chordoma includes trocar CT-guided biopsy which should be posterior and along midline. Surgery should include the biopsy track and aimed at achieving complete en block resection which is the most determinant for long-term outcome. Unfortunately, adequate margins are only achieved in roughly 50% of cases [17].

In a study among 101 patients with sacral chordoma either primary tumor or first time local recurrent tumor, combination of surgical resection and adjuvant radiotherapy has a good overall survival, local relapse free survival, and distant relapse free survival [18].

Our patient at EMD was hemodynamically stable, and after achieving partial pain control we could have a more detailed history regarding the low back pain that he suffered from and the courses of NSAIDs that he received.

Patient’s vital signs were monitored while he was in Radiology Department. We could have a CT scan of the abdomen and pelvis with IV contrast while monitoring his vital signs closely.

The sacral mass which was revealed in the CT scan could explain this six months history of low back pain with nocturnal worsening. These NSAID courses were incriminated for the prepyloric perforation and acute surgical abdomen presentation of our patient.

CONCLUSION
After stabilization of patient with acute surgical abdomen and pain control, further history taking will allow treating team to know the primary cause for acute abdomen.

REFERENCES

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