**A Curious Case of Autoimmunity, Pancytopenia, and Disseminated Intravascular Coagulation**

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**Abstract**

**History and examination:** A 21-year-old female patient presented to us with severe low back pain for 4 months. On examination, patient was afebrile, with severe pallor, and tenderness in both sacroiliac (SI) joints. Patient was being admitted and evaluated, and during the course of evaluation, developed severe headache, which was severe in intensity and associated with nausea and projectile vomiting.

**Initial investigations:** An X-ray of the bilateral SI joints revealed inflammation, and the antinuclear antibody (ANA) turned out to be 4+ with pancytopenia and raised lactate dehydrogenase (LDH), but the liver function tests were normal. Rest of the hematological profile was unremarkable. During the course of the evaluation, she developed a severe headache, which, on imaging, showed presence of cerebral edema with chronic subdural hematoma, and a concomitant coagulopathy workup revealed evidence of disseminated intravascular coagulation (DIC).

**Discussion:** Taking the whole picture into consideration, a malignant process in the body was suspected, and serum tumor markers carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA19-9), and cancer antigen 125 (CA-125) were sent, all of which were raised. Validating the clinical clue was the bone marrow biopsy done for pancytopenia, which revealed malignant epithelial infiltration. A contrast-enhanced computed tomography (CECT) thorax and whole abdomen were done to find out the primary, which showed a neoplastic mass at the gastroesophageal junction along with bony metastases in the vertebrae and left adrenal. Tissue from the primary lesion was taken for histopathological examination (HPE) through upper gastrointestinal endoscopy. Although HPE revealed grade III poorly differentiated stomach adenocarcinoma, the patient had succumbed to the disease process by the time the diagnosis came to light.

**Conclusion:** In short, this case perfectly illustrates how solid organ malignancies might be a mimic of multisystem disorders, thereby delaying diagnosis and worsening the prognosis even further.

**CASE DESCRIPTION**

A woman aged 21 years presented with severe low back pain for 4 months. She was pale, afebrile, with tenderness in both sacroiliac joints that was incapacitating. An X-ray of both sacroiliac (SI) joints showed inflammation features, warranting consideration of connective tissue disease.

**Case Description**

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**Introduction**

Delayed diagnosis due to absence or paucity of classical symptoms is one of the key prognostic factors in cancers and is a feature of adenocarcinoma in the young.1 Gastric cancer (GC) usually presents in middle and old age, between 50 and 70 years, with 50% of the freshly diagnosed stomach cancer patients aged >74 years.2 Local symptoms predominate, and GC is generally associated with a dismal prognosis unless localized at the time of diagnosis. However, 2–8% of GCs are reported in young persons (<41 years), called young adult GCs; they pursue an aggressive time of diagnosis. However, 2–8% of GCs are reported in young persons (<41 years), called young adult GCs; they pursue an aggressive course, frequently with less prominent local features and masquerade as a different disease altogether, baffling clinicians.3 We describe a 21-year-old female who presented with low back pain, who had strongly reactive antinuclear antibody (ANA), pancytopenia with features mimicking hemolytic anemia, disseminated intravascular coagulation (DIC), developed cerebral edema before an incidental diagnosis of gastric carcinoma could be achieved, with an extremely poor outcome.

**Table 1.**

<table>
<thead>
<tr>
<th>Item</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin (Hb)</td>
<td>6.6 gm/dL</td>
</tr>
<tr>
<td>Direct bilirubin</td>
<td>0.6 mg/dL</td>
</tr>
<tr>
<td>Indirect hyperbilirubinemia</td>
<td>2.6 mg/dL</td>
</tr>
<tr>
<td>Lactate dehydrogenase</td>
<td>1294 U/L</td>
</tr>
<tr>
<td>Alanine aminotransferase</td>
<td>87 U/L</td>
</tr>
<tr>
<td>Aspartate aminotransferase</td>
<td>93 U/L</td>
</tr>
<tr>
<td>Alkaline phosphatase level</td>
<td>1256 U/L</td>
</tr>
<tr>
<td>Normal γ-glutamyl transferase</td>
<td>23 U/L</td>
</tr>
<tr>
<td>Normal parathyroid hormone</td>
<td>40.2 mg/dL</td>
</tr>
<tr>
<td>Normal complement C3 level</td>
<td>125 mg/dL</td>
</tr>
<tr>
<td>Normal complement C4 level</td>
<td>37.22 pg/mL</td>
</tr>
<tr>
<td>Serum creatinine</td>
<td>0.8 mg/dL</td>
</tr>
<tr>
<td>Normal platelet count</td>
<td>125,000</td>
</tr>
<tr>
<td>Abnormal LDH</td>
<td>1294 U/L</td>
</tr>
<tr>
<td>Normal INR</td>
<td>0.9</td>
</tr>
<tr>
<td>Normal PT</td>
<td>12 seconds</td>
</tr>
</tbody>
</table>

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A positron emission tomography scan could not be done due to affordability issues. Finally, an upper gastrointestinal endoscopy was done, which confirmed a proximal gastric neoplastic mass, from which biopsy samples were taken. Eventually, the patient succumbed on the 12th day of her admission. An autopsy was not done in view of negative consent for the same by family members; however, the biopsy report that arrived later indicated metastatic carcinoma with overall reduced hematopoietic cellularity of all three cell lines. The epithelial origin of the cells was confirmed by immunohistochemistry staining with cytokeratin.

In search of the primary tumor, we did an ultrasound of the whole abdomen, which revealed a hypoechoic mass in the upper portion of the epigastrium and mild hepatomegaly with normal echotexture of the liver; this was followed up by a contrast-enhanced CT scan of the thorax and whole abdomen, which revealed an irregularly heterogeneously contrast-enhancing wall thickening at gastroesophageal junction having diffuse luminal irregularity with multifocal perigastric and retroperitoneal lymphadenopathy indicating a possibility of gastroesophageal junction neoplasm along with metastatic deposits in the T3, T6, T8-S1 vertebral body and left adrenal gland (Figs 3 and 4). A positron emission tomography scan could not be done due to affordability issues. Finally, an upper gastrointestinal endoscopy was done, which confirmed a proximal gastric neoplastic mass, from which biopsy samples were taken. Eventually, the patient succumbed on the 12th day of her admission. An autopsy was not done in view of negative consent for the same by family members; however, the biopsy report that arrived later indicated metastatic carcinoma with overall reduced hematopoietic cellularity of all three cell lines. The epithelial origin of the cells was confirmed by immunohistochemistry staining with cytokeratin.
proved it to be a grade III poorly differentiated adenocarcinoma of the stomach (Fig. 5).

Case Discussion

Uncommon and rare presentations of common diseases are the greatest challenges in medicine, not only as an academic curiosity but also to raise the alarm early for diagnosis and improved outcomes. Solid organ malignancies with a usual late onset are too frequently diffusely metastatic and multisystem in presentation when they occur at a disproportionate early age, worsening the outcome. The patient we report here had a GC diagnosis that was preterminal and hence inconsequential in targeted care because of the absence of local symptoms, baffling bone disease due to metastasis, positive markers of autoimmunity with hemolysis and DIC.

Gastric cancer (GC) is the fourth most common cancer worldwide, and effective care with surgery and neoadjuvant chemotherapy is largely dependent on early diagnosis. This patient had no gastrointestinal symptoms at all. She complained of severe lower back pain for the last four months, which was later discovered to be due to metastasis in the bone. In the last 2 decades, 40% of patients with GC present with metastasis and 12% of all metastasis occurs in the bone.4

There are several aspects of the case that make it worthy of mention. First, the low back pain, along with her young age and strongly reactive ANA, misled us in the first place, making us consider connective tissue disorder. It is important to note that significant and clinically consequential ANA reactivity in GC has not been reported so far. The diagnostic booby trap was widened in the context of a finding of elevated levels of LDH along with anemia and thrombocytopenia. This raised the possibility of immune hemolysis, but other features were absent. LDH and tumor markers CEA, CA19-9, and CA-125 were also elevated due to the underlying gastric carcinoma. Poorly differentiated adenocarcinoma and high levels of LDH, CEA, and CA19-9 are shown to be poor prognostic factors of gastric carcinoma.5

Second, she suddenly developed a severe headache, which, on a noncontrast CT scan of the brain, revealed diffuse cerebral edema with bilateral chronic subdural hematoma without midline shift. There have been very few reported cases of GC presenting with a bilateral subdural hematoma, and it is indeed a very unusual presentation of GC.6,7 The coagulation profile revealed raised PT, aPTT, INR, and FDP. All of these proved she developed an acute DIC. GC patients with DIC are rare but carry the risk of severe complications resulting in a dismal prognosis.8

Third, widespread bone metastasis was all too evident and worsened the prognosis further. She had markedly elevated alkaline phosphatase (ALP) (1256 U/L). Bone marrow aspiration and trephine biopsy showed infiltrative malignant cells of the epithelial type in the bone marrow along with a reduction of all three hematological cell lines, confirming a case of diffuse metastatic malignancy with unknown primary. The low back pain and tenderness in SI joints were probably due to metastasis to the bones. DIC associated with metastatic malignant bone marrow involvement, like our patient, is a rare complication of gastric carcinoma.9

The net 1-year survival rate for stage IV GC is 21.4% (CI: 20–22.9) for females in the UK.3

Conclusion

In conclusion, one must remember that neoplastic cells can wreak havoc with our immune system. A clinician must always consider the diagnosis of occult malignancy, particularly intraabdominal visceral malignancies like GC, in patients presenting with a combination of bizarre features like bone pain, cytopenia, false positive ANA, and bleeding manifestations.

References