Rare Tumor in a Rare Location in Patient With Constipation

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— Could occupational exposure to sawdust explain this case?

by Kate Kneisel, Contributing Writer, MedPage Today March 14, 2022

A 56-year-old man presented to a hospital with a 3-week history of severe constipation, accompanied by back pain that worsened when he ate. He also reported headache and excessively watering eyes, although he was less concerned about these symptoms.

The patient had a smoking history of 32 pack-years, although he noted that he stopped smoking many years ago, and did not regularly drink alcohol. He explained that he had not taken any treatment for his constipation, had not lost weight, and had not felt weak or lacking in energy. He noted that he works as a carpenter, with frequent exposure to sawdust.

Clinicians found he had no relevant past medical history. His WHO performance status was 0. Following his initial assessment, he developed a mild nose bleed.

The clinical examination was somewhat unrevealing, showing mainly abdominal distention, as well as spinal syndrome without any associated neurologic symptoms. Specifically, the patient had no signs of intracranial hypertension or sinusitis, and his eyes showed no obvious exophthalmia.

Blood tests showed a normal complete blood count; however, blood analysis indicated that the patient had moderate thrombocytopenia (65,000 platelets/L) and hypoalbuminemia (28 g/L).

The laboratory report showed severe cholestasis and cytolysis with no significant icterus, and normal blood electrolytes and renal function.

The patient's C-reactive protein level was 18 mg/L, his procalcitonin level was 4.71 µg/L, and his neuron-specific enolase was elevated at 169 µg/L.

Clinicians performed an abdominal CT scan, which revealed heterogeneous liver nodules with compression of the inferior vena cava, and multiple vertebral lytic lesions suspicious as possible metastases (Figure). Colonoscopy and gastroscopy revealed no unusual findings, although there was evidence of slight thickening of the colon.

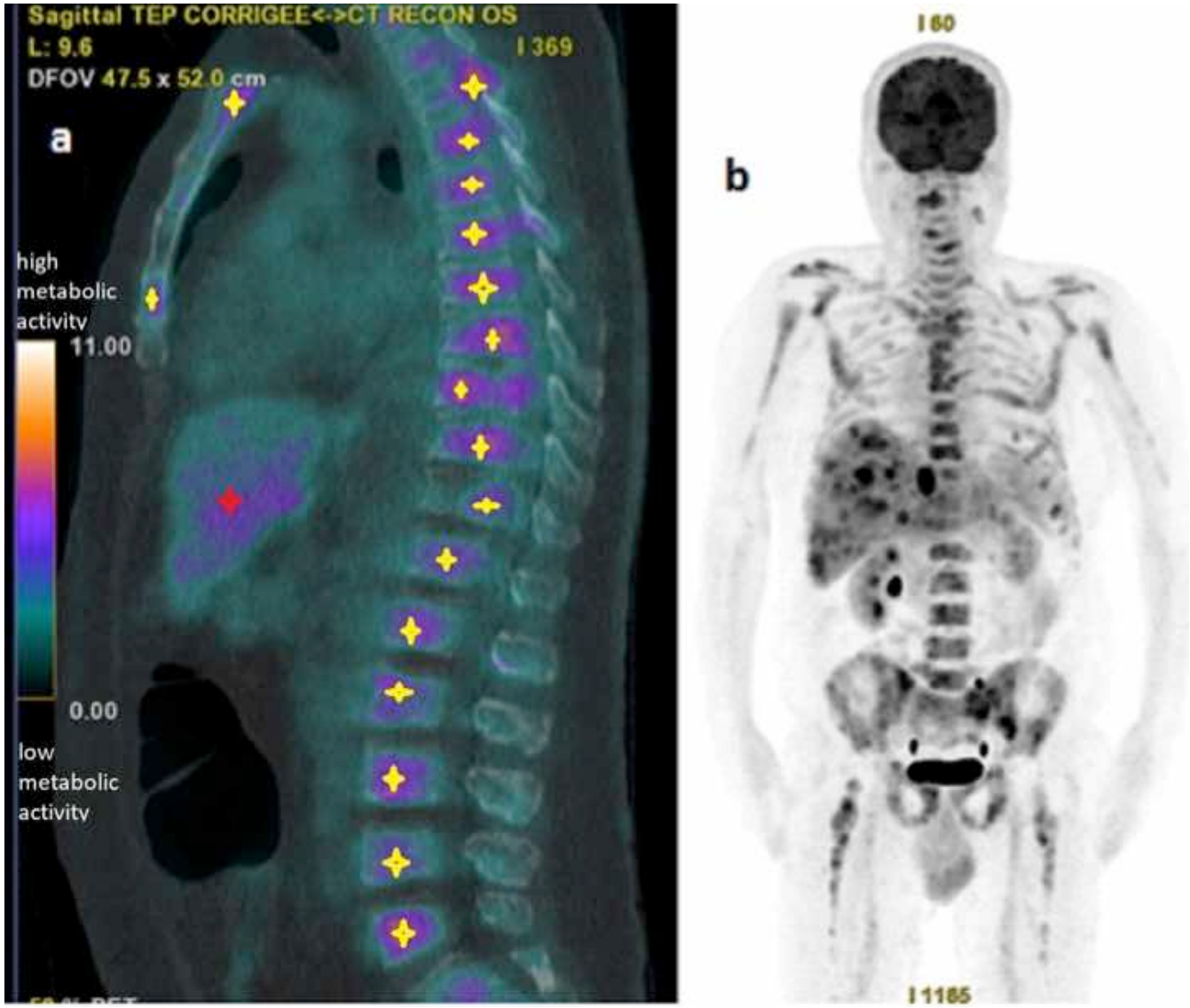

Figure. Sagittal (A) and coronal (B) PET scan overall views showing initial multiple metastatic locations including liver nodules (red cross) and vertebral lytic lesions (yellow crosses).

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They also performed a liver biopsy and PET scan, which showed a 42-mm hypermetabolic lesion in the left ethmoidal bone; a sample of the liver tumor was suggestive of small cell neuroendocrine carcinoma. They noted additional metastases in the left adrenal gland and multiple adenopathy and bone locations, which they felt accounted for the patient's epidural pain.

The team considered performing an ethmoidal biopsy to confirm that the patient did not have an additional previously undetected small cell carcinoma (SCC) located elsewhere. However, they did not pursue further diagnostic measures, given that the chest and abdominal CT scans had not identified pulmonary or digestive primary tumor candidates, and such findings would not lead to different treatment approaches in any case.

A previous study noted that SCC is rarely found outside pulmonary or digestive system sites, with primary head and neck locations accounting for only about 10% of all cases.

The case authors noted that recent studies on the treatment of neuroendocrine tumors of the head and neck have not shown any significant changes over the past 10 years. As in other types of

neuroendocrine carcinomas, the combined use of platinum and etoposide is recommended, with cisplatin-etoposide doublet chemotherapy used most widely for pulmonary SCCs. "Thus, our patient received doublet chemotherapy with dose adaptation due to thrombocytopenia ... [and ongoing] classical corticosteroid treatment and radiotherapy to address the epidural location," they wrote.

Discussion

Clinicians reporting this case of a patient presenting with digestive symptoms and occupational exposure to sawdust, who was ultimately found to have a small cell carcinoma with an atypical ethmoidal location noted that because of these aspects, "[this case] is an interesting illustration of the heterogeneity of small cell neuroendocrine carcinomas."

The patient's initial clinical presentation with primarily digestive symptoms served to obscure the diagnosis, they suggested. Thus, when "diagnostic wavering ensued," they reviewed not only the patient's clinical history but also the current literature, which includes about 100 reports of small cell neuroendocrine carcinoma affecting the paranasal sinuses.

The authors noted their belief that this is only "the second reported case with an ethmoidal location," adding that most head and neck carcinomas involve the larynx and hypopharynx.

Paranasal carcinomas are typically associated with ear, nose, and throat (ENT) symptoms, such as protrusion of the eyeball(s), headache, impaired vision and sense of smell, facial pain, and rhinorrhea. In addition to these symptoms, the group noted that the sole other documented case of ethmoidal SCC involved seizures and inappropriate antidiuretic hormone secretion.

The authors explained that this patient's presentation with primarily digestive and spinal symptoms, without any significant symptoms related to the notably large primary tumor, "emphasizes the need for a systematic review of the risk factors when malignancy is suspected."

About 80% of head and neck SCC cases occur in men at a median age of about 74 years, the group wrote. Of SCCs that occur outside the lungs or digestive system, the most important risk factor seems to be a history of smoking. This case expands on previous descriptions of SCC of the paranasal sinuses, they noted.

The group cautioned that this presumably index case of SCC "associated with both smoking history and occupational exposure to sawdust ... provides only anecdotal evidence and does not confirm the causal effect of sawdust exposure. However, the rarity of SCC at the ethmoidal site and the well-known carcinogenic effect of sawdust exposure for the ethmoid epithelium makes it plausible."

In addition to its overall rarity, unusual location, and uncharacteristic presentation, the authors concluded that this case of ethmoidal SCC "underlines the importance of ascertaining risk factors and suggests that sawdust exposure should prompt an investigation of the ethmoidal location, regardless of the histological nature."

Disclosures

The authors reported no conflicts of interest.