Small Cell Neuroendocrine Carcinoma of the Paranasal Sinuses with Unexpected Location and Clinical Presentation and Occupational Exposure: A Case Report

Pierre Tankere
Sylvie Zanetta
Courèche Guillaume Kaderbhai
Hélène Bellio
Aurélie Lagrange
Martin Garcier
François Ghiringhelli

Corresponding Author: Pierre Tankere, e-mail: ptankere@gmail.com
Financial support: None declared
Conflict of interest: None declared

Patient: Male, 56-year-old
Final Diagnosis: Small cell carcinoma head and neck
Symptoms: Initial clinical digestive presentation • occupational exposure to sawdust
Medication: —
Clinical Procedure: —
Specialty: Oncology • Otolaryngology • Pulmonology

Objective: Unusual clinical course
Background: Small cell carcinoma (SCC) is usually aggressive and associated with a poor prognosis. This type of cancer is rarely found in extra-pulmonary or digestive-tract locations. This report describes an unusual presentation in terms of clinical symptoms and primary location (even among head and neck presentations), and unexpected occupational exposure.

Case Report: This case report is a novel observation of SCC in the paranasal sinuses and is to the best of our knowledge the first case associated with occupational exposure to sawdust, with almost no ear, nose, and throat (ENT) symptoms related to the primary tumor location.
Our patient had no past medical history and none of the usual risk factors, apart from a smoking history of 32 pack-years. He was not taking any regular treatment.
The inaugural clinical presentation was mainly digestive, with neither ENT nor neurological symptoms. Diagnostic waverings ensued and numerous paraclinical tests were performed.
This is one of the very few cases of an ethmoidal location ever reported for SCC. This cancer type is unusual in neck and head locations, but has occasionally been reported in the larynx and hypopharynx. To the best of our knowledge, this is only the second report of an ethmoidal location.
In addition, this patient was a carpenter, implying exposure to sawdust, which is usually associated with adenocarcinoma of the ethmoid. We illustrate here that SCC, which has been described elsewhere without sawdust exposure, is also possible.

Conclusions: Exposure to sawdust should suggest a possible ethmoidal cancer location, even if there are few ENT symptoms. Adenocarcinoma is the most prevalent but clearly not the only possible histological pattern.

Keywords: Carcinoma, Small Cell • Ethmoid Sinus • Occupational Exposure

Abbreviations: SCC = small cell carcinoma; ENT = ear nose and throat

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/934980
Background

Small cell carcinoma (SCC) is seldom found in extra-pulmonary or extra-digestive sites, and primary head and neck locations account for only about 10% of all cases of SCC [1,2]. Here we describe a case characterized by an atypical ethmoidal location, an initial clinical digestive presentation, and occupational exposure to sawdust. Diagnostic wavering ensued. We therefore reviewed the patient’s clinical history and conducted a systematic review of the current literature. The rarely reported location, unusual clinical presentation, and exposure to sawdust in this case is an interesting illustration of the heterogeneity of small cell neuroendocrine carcinomas, reinforcing and extending previous reports.

Case Report

Our patient was a 56-year-old carpenter with no relevant past medical history. Although he quit smoking many years previously, he had a smoking history of 32 pack-years. He reported no regular alcohol consumption and no daily treatment prior to consultation for the episode described here.

The initial clinical presentation included a 3-week history of severe constipation and back pain exacerbated by eating. WHO performance status at diagnosis was 0, with no general symptoms such as weight loss or asthenia at the first medical exam. The patient described slight epiphora and headache, which he felt were unimportant compared with his digestive symptoms and back pain. Mild epistaxis and thrombocytopenia occurred after the initial consultation.

Figure 1. 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).
Results of a clinical examination were rather poor and revealed mainly abdominal distention as well as spinal syndrome without any associated neurological symptoms. In particular, there were no signs of intracranial hypertension or sinusitis, and no obvious exophthalmia.

The complete blood count was normal, but the blood sample revealed moderate thrombocytopenia (65,000 platelets/L) and hypoalbuminemia (28 g/L). The laboratory findings revealed severe cholestasis (ALP 329 IU/L; G.G.T 2231 IU/L) and cytolysis (T.G.O 607 IU/L; T.G.P 1150 IU/L) with no significant icterus. Blood electrolytes and renal function were normal. The C-reactive protein level was 18 mg/L, procalcitonin was 4.71 µg/L, and neuron-specific enolase was elevated at 169 µg/L (UNL<12 IU/L).

The initial investigations also included an abdominal CT scan, which showed heterogeneous liver nodules with compression of the inferior vena cava, and multiple vertebral lytic lesions suggestive of metastases (Figure 1). Colonoscopy and gastroscopy were performed, revealing no abnormalities, but showing slight thickening of the colon.

The diagnosis was confirmed by liver biopsy and PET scan. These revealed a 42-mm hypermetabolic lesion in the left ethmoidal bone (Figure 2) and a hepatic tumor sample compatible with small cell neuroendocrine carcinoma. Further metastases were observed in the left adrenal gland and multiple adenopathy and bone locations, which explained the epidural pain.

Ethmoidal biopsy was discussed as a means to ensure that the patient did not have both ethmoidal adenocarcinoma and small cell carcinoma of unknown location. However, the care team decided against it because the results would not change the therapeutic approach, and without pulmonary nor digestive primary tumor candidate at chest and abdominal CT scan this hypothesis seemed highly unlikely.

A review of the recent literature regarding treatment of neuroendocrine tumors of the head and neck [2] revealed that there...
have been no significant changes over the past 10 years. The combined use of platinum and etoposide is recommended, as in other types of neuroendocrine carcinoma. A retrospective review of 120 patients [3] also confirmed that the most widely used treatment is based on cisplatin-etoposide doublet chemotherapy, similar to pulmonary SCC. Thus, our patient received doublet chemotherapy with dose adaptation due to thrombocytopenia. We used classical corticosteroid treatment and radiotherapy to address the epidural location. The patient is still receiving this first-line treatment.

Discussion

Rare Tumor in a Rare Location

There are about 100 reported cases of small cell neuroendocrine carcinoma of the paranasal sinuses. The most common neck and head locations include the larynx and hypopharynx [2,4]. As far as we know, this is only the second reported case with an ethmoidal location [5].

Clinical Digestive Presentation Obscuring the Diagnosis

The initial symptoms of paranasal carcinoma are usually ENT symptoms, including exophthalmia, headache, visual impairment, facial pain, anosmia, and rhinorrhea. The only other documented ethmoidal SCC also reported seizures and inappropriate antidiuretic hormone secretion [5]. This is the first reported case in which a patient described mainly digestive and spinal symptoms, with no predominant symptoms linked to the primary tumor despite its large size (42 mm). This emphasizes the need for a systematic review of the risk factors when malignancy is suspected.

Risk Factors

Patients with SCC of the head and neck are generally men (80%) with a median age of about 74 years. The risk factors described in SCC with extra-pulmonary or digestive sites are generally similar to other types of SCC, and smoking history seems to be the most important risk factor [2]. Our case report adds new observations for SCC of the paranasal sinuses and is also, to the best of our knowledge, the first case associated with both smoking history and occupational exposure to sawdust [6]. Furthermore, it is the second case of ethmoidal location ever reported. This report 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.

Conclusions

Our case adds new information regarding SCC of the paranasal sinuses and is, to the best of our knowledge, the first case revealed almost exclusively by digestive symptoms and associated with occupational exposure to sawdust. Furthermore, it is one of the very few reported cases of ethmoidal SCC. This 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.

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