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Case Report

A Rare Presentation of Extrapulmonary Small Cell Cancer with Bone Involvement: Case Report and Insight into Clinical Management

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Abstract

Extrapulmonary Small Cell Carcinoma (EPSCC) is a rare and aggressive malignancy that shares histopathological features with small cell lung cancer but originates outside the lungs. Bone metastasis in EPSCC, especially involving the spine, pelvis, and ribs, is common and correlates with poor prognosis. This case report describes the clinical course of a 51-year-old male with a history of pulmonary small cell carcinoma who presented with diffuse pain and anemia due to extensive osteoblastic metastatic disease. Despite a multimodal treatment approach including chemotherapy, immunotherapy, and bone-targeting agents, the patient's condition progressively worsened. Over several follow-up visits, his symptoms, including weakness and pain, were managed with palliative chemotherapy and supportive care. The patient eventually transitioned to hospice care after a discussion regarding his poor prognosis. This case highlights the challenges in managing EPSCC with bone metastasis and underscores the need for further research into effective therapies, as current treatment regimens offer limited survival benefits. Enhanced awareness and early intervention may improve outcomes and quality of life for affected individuals.

Introduction

Extrapulmonary small cell cancer (EPSCC) is a rare and aggressive neoplasm characterized by small cell carcinoma that arises outside the lungs. The incidence of EPSCC is low, accounting for approximately 2% of all small cell carcinoma cases, with a higher prevalence in older adults [1]. Unlike its pulmonary counterpart, which is more commonly recognized, EPSCC typically originates in other anatomical sites, including the gastrointestinal tract, genitourinary system, and skeletal-muscular system [2]. Bone metastasis in EPSCC often involves the axial skeleton, particularly the spine, pelvis, and ribs, being most frequent. The presence of bone metastasis often correlates with poor prognosis, contributing to pain, fractures, and neurological deficits, which can further complicate treatment and quality of life [3]. The prognosis for patients with EPSCC is generally poor, with a median survival rate that varies widely depending on the tumor's site, stage at diagnosis, and response to treatment. The aggressive nature of this cancer often leads to advanced disease at diagnosis, resulting in a 5-year survival rate estimated at less than 10% [4].

Treatment typically involves a multimodal approach, including chemotherapy, radiation therapy, and, in some cases, surgical resection. This case report highlights the clinical presentation and management course associated with EPSCC metastasis to the bone, offering valuable insights into its prognosis and treatment strategies. The case further underscores the need for increased awareness and research into this rare malignancy, which may improve management and goal of care planning for affected individuals.

Case Presentation

A 51-year-old male with a medical history of pulmonary small cell carcinoma, metastatic disease, hypertension, and hyperlipidemia presented to the oncology clinic for ongoing management of chemotherapy and radiation for his extrapulmonary small cell carcinoma. During his initial visit, the patient reported experiencing severe, diffuse pain throughout his body. Laboratory results revealed a white blood cell count (WBC) of 5.0 109/L, hemoglobin 7.9 g/dL, platelets 213 109/L, mean corpuscular volume (MCV) 82.1 fL. The complete metabolic panel (CMP) was notable for an alkaline phosphatase level of 1476 IU/L, while other results were within normal limits. A CT scan of the chest, abdomen, and pelvis at that time showed diffuse osteoblastic metastatic disease, without evidence of soft tissue metastasis in the chest, abdomen, or pelvis. Based on these findings, the oncologist has developed a treatment plan, which includes palliative chemotherapy with cisplatin and etoposide, Atezolizumab, the bone remodeling agent Aredia, and Norco for pain management. Additionally, the patient reported generalized weakness and was administered an iron infusion. Complete Metabolic Panel (CMP) is significant for alkaline phosphatase 1476 IU/L, the rest was normal.

At his second follow-up visit in July 2022, six weeks after his

initial consultation, the patient remained anemic. Laboratory results showed a white blood cell count (WBC) of 4.3 × 10⁹/L, hemoglobin of 6.5 g/dL, mean corpuscular volume (MCV) of 87.1 fL, iron level of 29 mcg/dL, total iron binding capacity (TIBC) of 314 mcg/dL, iron saturation of 92%, and ferritin of 2111 ng/mL, which indicated iron overload likely due to previous blood transfusions. The anemia at this visit was primarily attributed to his extensive cancer and probable bone marrow metastasis. The patient was started on Procrit 10,000 units weekly for eight weeks.

By his third visit in August 2022, the patient reported significant improvement in symptoms, including better strength, increased appetite, and weight gain. A complete blood count (CBC) showed an increase in hemoglobin to 8.1 g/dL. The treatment plan was adjusted to transition the patient to immunotherapy alone.

At his fourth follow-up visit in September 2022, six months after transitioning to monotherapy with Atezolizumab, the patient was stable with no new symptoms. However, during his fifth visit in September 2022, he reported a recurrence of weakness and diffuse pain. Despite a stable hemoglobin level of 11.6 g/dL, a repeat CT scan revealed progression of diffuse osteoblastic metastatic disease, and chemotherapy with cisplatin and etoposide was reintroduced.

By his sixth visit in December 2022, the patient remained relatively stable. A bone scan showed diffuse increased bony activity, with focal areas indicating worsening bone metastasis. CT imaging confirmed extensive bone metastasis, prompting the addition of Tecentriq to his chemotherapy regimen. Additionally, he was prescribed Procrit 20,000 units twice a week to manage his anemia-related symptoms.

During his seventh visit in April 2023, the patient's symptoms worsened significantly. He complained of increased weakness and was admitted to the emergency room a week prior for severe lethargy, with his hemoglobin dropping to 8.6 g/dL. At this point, the oncologist had a discussion with the patient's family regarding the patient's poor prognosis. The family was informed that the cancer was incurable and would continue to progress. The patient was subsequently admitted to hospice care and passed away four months later.

Discussion

EPSCC is a rare, aggressive malignancy that originates outside of the lungs, yet shares histopathological features with small cell lung carcinoma (SCLC). This tumor typically demonstrates a high propensity for early metastasis, often to the liver, bones, and brain. The exact molecular mechanisms driving EPSCC are not fully understood, but like its pulmonary counterpart, it is associated with mutations in tumor suppressor genes such as p53 or RB1, leading to uncontrolled cellular proliferation and resistance to apoptosis. This cancer type commonly presents at advanced stages with a high potential for metastasis, particularly to distant organs such as the liver, bone, and brain. Among the various sites of metastasis, bone involvement is a significant factor that adversely impacts prognosis, contributing to both morbidity and mortality. The presence of bone metastases in EPSCC patients is associated with a poor overall survival rate, as these lesions often cause severe complications such as pain, fractures, and spinal cord compression. Recent studies indicate that survival rates are drastically reduced for patients with extrapulmonary small cell cancer that has spread to bone, with a 1-year survival rate often falling below 50% due to the rapid disease progression [5].

The treatment of EPSCC with bone metastasis is challenging, as the disease's aggressive nature makes it less responsive to conventional therapies. A combination of systemic chemotherapy, localized radiotherapy, and supportive care forms the cornerstone of the treatment plan. Chemotherapy regimens that include agents like etoposide and cisplatin or carboplatin are standard and typically offer modest benefits in terms of disease control. However, these therapies are often palliative rather than curative, and long-term survival remains rare. The use of radiotherapy, particularly for bone lesions causing pain or neurological complications, can provide some symptomatic relief and improve the quality of life, but it does not significantly alter the course of the disease [6]. In some cases, the addition of targeted therapies, such as immune checkpoint inhibitors, is being investigated, although results have been mixed, and further studies are necessary to establish their role.

The prognosis for patients with extrapulmonary small cell cancer and bone metastasis remains poor, with median survival times generally ranging from 6 to 12 months. Early diagnosis and aggressive treatment can improve survival slightly, but the prognosis is heavily influenced by the extent of metastasis, the patient's overall health, and the response to initial therapy. Some studies have suggested that patients who respond well to chemotherapy can live longer, but these individuals still face high recurrence rates and rapid progression upon relapse. For example, a study by Tokito found that while chemotherapy could extend survival in a subset of patients, the median survival for those with metastatic EPSCC involving bone was still under a year, with many patients experiencing deterioration within the first few months following treatment [7].

In light of the poor prognosis, treatment strategies for EPSCC with bone metastasis increasingly focus on improving quality of life and providing palliative care. For instance, bisphosphonates or denosumab, which inhibit bone resorption, are sometimes used to manage bone complications and reduce the risk of skeletal-related events (SREs) like fractures. These interventions may alleviate some of the symptoms associated with bone metastases, such as pain and hypercalcemia, thereby improving the patient's quality of life. Despite the lack of curative treatments, ongoing research into molecular targets and novel therapies, including immunotherapy and combination approaches, offers hope for better outcomes in the future [8]. However, until more effective therapies are identified, the treatment course for patients with EPSCC and bone metastasis will largely be supportive and focused on symptom management, with survival remaining limited.

Conclusion

Extrapulmonary small cell carcinoma with bone metastasis presents significant clinical challenges due to its aggressive nature, poor prognosis, and limited response to conventional therapies. This case underscores the importance of early diagnosis, multimodal treatment, and symptom management to improve quality of life for affected patients. Despite aggressive interventions, survival remains poor, with most patients experiencing rapid disease progression. Ongoing research into targeted therapies, including immunotherapy and combination approaches, is crucial for developing more effective treat-

ments. When treating such patients, clinicians must be aware of the challenge in management along with poor prognosis and consider palliative care strategies to manage symptoms and improve patient outcomes.

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