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Case Report

Burkitt's Lymphoma Presenting as Pancoast Syndrome: A Rare Clinical Entity

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Abstract

This article describes a rare clinical presentation of Burkitt's lymphoma, an aggressive malignancy that presented as an anterior mediastinal mass causing Pancoast syndrome in a 39-year-old pregnant woman. The patient presented with respiratory distress, right upper limb monoparesis, significant weight loss, and multiple bilateral axillary and cervical lymphadenopathies. Blood tests revealed non-regenerative anemia and thrombocytopenia, and a cervico-thoraco-abdomino-pelvic scan showed a lobulated, tissue-dense mass with supraclavicular extension, located at the right paramediastinal apex. The bone marrow biopsy showed disseminated Burkitt lymphoma. The article highlights the diverse clinical manifestations of Burkitt lymphoma and discusses its clinical variants, diagnostic techniques, and symptoms. Early diagnosis and treatment are crucial for a favorable outcome.

Keywords: Burkitt's lymphoma; Pancoast syndrome; Anterior mediastinal mass; Pregnancy

Abbreviations: MALT: Mucosa-Associated Lymphoid Tissue; HIV: Human Immunodeficiency Virus; WHO: World Hhealth Organization; CNS: Central Nervous System; CT: Computed Tomography

Introduction

Pancoast syndrome is a rare clinical entity characterized by a set of symptoms caused by a tumor located in the superior sulcus of the lung. It is commonly associated with lung cancer, but it can also be a manifestation of other malignant neoplasms [1]. Burkitt's lymphoma, a rapidly growing B-cell lymphoma, is not a common cause malignancy that can present with Pancoast syndrome. The diagnosis of Burkitt's lymphoma in the setting of Pancoast syndrome can be challenging, as it may mimic other more common etiologies of this clinical entity. Therefore, a high index of suspicion and prompt diagnostic evaluation are necessary for timely diagnosis and appropriate management. In this article, we describe a case of Pancoast syndrome caused by Burkitt's lymphoma and review the clinical features, diagnostic workup, and management of this rare presentation.

Case Presentation

A 39-year-old woman with a history of chronic thyroid adenoma under surveillance for 9 years, 16 weeks pregnant, presented with respiratory distress and spontaneous pain in the right upper limb with electric shock-like sensations extending to the forearm for the past 15 days, along with significant weight loss of 10kg.

Upon admission, the patient was conscious, hemodynamically and respiratory stable, with right upper limb monoparesis (muscle strength at 3/5) with preserved osteotendinous reflexes and right ptosis and miosis.

The rest of the examination revealed a hard and well-defined 3cm right supraclavicular mass with multiple bilateral axillary and cervical lymphadenopathies, cardiac and pulmonary auscultation was normal, abdominal clinical examination was normal.

Blood tests showed bicytopenia (non-regenerative anemia: Hb=6g/dl, reticulocytes=10000/mm^3 with thrombocytopenia at 101000/mm^3), along with a clear inflammatory syndrome: CRP: 182mg/l, D-dimers: 4384ng/ml, PCT<0,05µg/l. BHCG was positive (104 600UI/ml).

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The abdominal-pelvic ultrasound revealed a stopped pregnancy estimated at 12 weeks. A cervico-thoraco-abdomino-pelvic scan showed a lobulated, tissue-dense mass (45UH) measuring 45x45x50mm with supraclavicular extension, located at the right paramediastinal apex, and abutting the pleura, pushing the right carotid and internal jugular veins inward and the right subclavian artery outward, with intimate contact with the homolateral cervical nerve roots.

A bone marrow biopsy and a transthoracic needle aspiration were performed to establish the diagnosis. The bone marrow biopsy showed 90% infiltration by large-sized cells with a high nuclear to cytoplasmic ratio, indicative of disseminated Burkitt lymphoma. A transthoracic needle aspiration of the mass confirmed the diagnosis.

Discussion

Our case is an extremely rare case of Burkitt lymphoma presenting as an anterior mediastinal mass causing Pancoast syndrome. Diffuse large B cell lymphoma is the most common subtype of non-Hodgkin lymphoma, accounting for about a third of reported cases [2]. The most common primary pulmonary lymphoma is low-grade mucosa-associated lymphoid tissue (MALT) lymphoma, which originates in the bronchi, and the second most common is diffuse large B-cell lymphoma [3].

Pulmonary primary lymphomas generally have a non-progressive pattern and a good prognosis. High-grade lymphomas have a poor prognosis, most of which are caused by transmission from low-grade lymphomas or occur in immunocompromised conditions [4].

Burkitt's lymphoma, first described in Ugandan children in 1958 by Denis Parsons Burkitt, has a cell doubling time of 2 days and constitutes the human fastest growing known tumor [5]. It represents the first tumor associated with a viral infection, the first tumor associated with HIV infection and the first tumor associated with a chromosomal translocation responsible for the activation of an oncogene [6]. The 5-year survival rate for Burkitt's lymphoma ranges from 60% to 85% [7].

Three clinical variants of Burkitt's lymphoma are described by the World Health Organization (WHO) classification describes: endemic, sporadic (the predominant type in malaria-free areas) and immunodeficient. These clinical variants share similarities in terms of their morphology, genetic characteristics, and immunophenotype [8].

Pathological confirmation of Burkitt's lymphoma is essential, but imaging also plays a crucial role in the diagnosis, treatment, and monitoring of patients. In emergency situations with acute symptoms, ultrasound or Computed Tomography (CT) is usually employed, with ultrasound being particularly useful for detecting palpable masses or intussusception in pediatric patients [9]. CT provides rapid, whole-body assessment with better soft tissue resolution than ultrasound. Magnetic Resonance Imaging (MRI), although not routinely used in emergency settings, has an increasing role in Burkitt's lymphoma evaluation, especially for superior soft tissue characterization in assessing tumor extension and Central Nervous System (CNS) involvement. MRI protocols should be customized according to the anatomy being evaluated [10].

Burkitt's lymphoma is known for its aggressive behavior and often involves areas outside of the lymph nodes. It may even progress to acute leukemia. The tumor is characterized by medium-sized lymphoid cells with scattered macrophages, giving it a "starry sky" appearance. The cells overlap in a "puzzle" pattern, with little deformation or overlap. They have a high nuclear to cytoplasm ratio and exhibit finely divided chromatin, discrete nucleoli, and scant basophilic cytoplasm. Mitotic figures, apoptotic bodies, and areas of necrosis are readily observable. Immunohistochemistry tests indicate that Burkitt lymphoma cells are positive for CD45, CD19, CD20, CD10, and bcl-6, with light chain restriction, and negative for CD5, CD34, TdT, and bcl-2. Ki67 proliferation is typically greater than 95% [7].

Circumstances of revelation of Burkitt's lymphoma are varied, ranging from the asymptomatic form diagnosed during a standard check-up or complementary check-up prescribed for another reason, to symptomatic forms with constitutional symptoms or symptoms related to the anatomical location of the tumor, or patients with a paraneoplastic syndrome (disorders related to the immune system reaction to cancer). Burkitt's lymphoma has most commonly an abdominal symptomatology presentation (60-80%). The usual symptoms include abdominal pain, distension, nausea, vomiting, and gastrointestinal bleeding [11]. Head and neck region is the next most frequent site of presentation, involving lymphadenopathy, and sinus, nasal oropharyngeal, or tonsillar involvement. Infrequently, the jaw may be affected. Bone marrow infiltration occurs in approximately 20% of patients, and some cases are classified as Burkitt's leukemia, characterized by extensive marrow infiltration with more than 25% blasts, which may present with bone pain. Other rare presenting sites include the CNS, thyroid gland, mediastinum, breasts, and skin [7].

In our case, it was pancoast syndrome, a constellation of signs and symptoms associated with extrinsic compression of anatomical structures at the thoracic inlet at the apex of the lungs. Pancoast syndrome, also known as Pancoast-Tobias syndrome, is a medical condition characterized by tumors in the upper part of the lungs. It was first described in the early 1900s in three cases of superior pulmonary sulcus tumor, and later revisited in 1932 by Tobias, who identified the bronchopulmonary origin of the tumor [1]. The classic symptoms of Pancoast syndrome include neuralgia of the upper limb on the same side as the tumor, limited active movements, costal lysis of the first four ribs or vertebrae, and a Claude Bernard Horner syndrome [12]. The Horner syndrome, described for the first time in history in 1869 by Johann Friedrich Horner, is identified by three symptoms, which are ipsilateral ptosis, miosis, and anhidrosis, and is caused by the invasion or compression of the paravertebral sympathetic and inferior (stellate) cervical ganglion by the tumor. Between 14% and 50% of patients with lung tumors have Horner's syndrome, and 40% of patients with Pancoast tumors experience this symptom [13]. It is important to differentiate this syndrome from brachial neuritis, which has also been described as paraneoplastic lymphoma syndrome, particularly the Hodgkin variant.

Pancoast syndrome can be challenging to diagnose because it occurs in the outermost part of the lungs. Transthoracic aspiration is typically used for diagnosis [14]. The type of cell involved is usually a slow-growing, well-defined squamous cell carcinoma that can spread to nearby subpleural lymph nodes and structures. In some cases, the technical challenges of histological diagnosis, along with severe pain and high probability of bronchial carcinoma, may lead to initial radiotherapy based on clinical and radiological diagnosis [14]. However, other conditions like benign tumors such as hamartomas and pulmonary

adenomas, as well as non-tumor pathologies such as pulmonary tuberculosis, destructive staph infections, aspergillosis, or histoplasmosis, may cause similar symptoms, making an accurate diagnosis crucial. In cases where the tumor has endobronchial extensions, a bronchial biopsy may be helpful [4]. In our patient's case, identifying the underlying cause led to a change in treatment.

The source of the tumor in our instance was uncertain. One possibility is that it originated in the lung tissue or the subpleural lymphoid tissue and then disseminated to the mediastinal lymph nodes and neck, which is similar to primary extranodal Burkitt lymphoma of the lung. Another hypothesis is that the tumor originated in the neck or neck lymph nodes and extended in a backward direction to the subpleural lymph nodes and bronchial submucosal lymphoid tissue.

The FAB LMB study or Berlin-Frankfurt-Münster protocols are followed for the management of Burkitt's lymphoma in most medical center. These protocols involve initial cytoreduction using a combination of cyclophosphamide, prednisolone, and vincristine, followed by more intense chemotherapy with varying drug combinations [7]. High risk of pronounced tumor lysis is possible in first days of therapy pose, but the use of urate oxidase has considerably reduced this danger. However, due to the toxic effects of these protocols, advanced supportive care is necessary, which may not be available in low-income countries.

Conclusions

In conclusion, this case highlights the rare presentation of Burkitt lymphoma as an anterior mediastinal mass causing Pancoast syndrome. Burkitt lymphoma is an aggressive malignancy that can present with varied symptoms, including abdominal pain, lymphadenopathy, and constitutional symptoms. Prompt diagnosis and treatment are crucial for a favorable outcome. Clinicians should be aware of the diverse clinical manifestations of Burkitt lymphoma and consider it as a differential diagnosis when evaluating patients with lymphadenopathy and other associated symptoms.

Author Statements

Declaration of Conflicting Interests

The authors declared no conflicts of interest with respect to the research, authorship, and/or publication of this article.

Consent for Publication

Written informed consent was obtained from the patient for publication of the case report and any accompanying images.

Availability of Supporting Data

All clinical finding and radiological results included in this case report can be found in the archived medical file of the patient.

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Authors' Contributions

All authors have contributed to this work since conception, reading and endorsing the final version of the manuscript.

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