CLINICAL CASE

LINFANGITIS CARCINOMATOSA, CASO CLINICO

CARCINOMATOUS LYMPHANGITIS, CLINICAL CASE

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RESUMEN

La linfangitis carcinomatosa es una forma poco común de diseminación pulmonar relacionada con metástasis de neoplasias malignas, principalmente adenocarcinomas, aproximadamente el 6-8% de los pacientes con enfermedad maligna desarrollan linfangitis carcinomatosa, esta condición provoca la invasión progresiva de los vasos linfáticos pulmonares, afectando la circulación linfática y generando problemas respiratorios, el presente caso detalla la experiencia de una paciente con carcinoma ovárico y linfangitis carcinomatosa, destacando la complejidad en el diagnóstico debido a síntomas inespecíficos y su lenta progresión. Se discuten las implicaciones de la linfangitis carcinomatosa, su asociación con diferentes tipos de cáncer, y se subraya la importancia de técnicas de imagen avanzadas para el diagnóstico temprano. Se presenta un análisis de casos previos que resalta la rareza de la condición y su pronóstico desfavorable. En general, se destaca la necesidad de un enfoque integral en el manejo de pacientes con síndromes paraneoplásicos, especialmente aquellos asociados con complicaciones respiratorias como la linfangitis carcinomatosa.

PALABRAS CLAVE: Linfangitis carcinomatosa, cáncer de ovario, carcinomatosis linfangítica pulmonar, neoplasia maligna.

ABSTRACT

Carcinomatous lymphangitis is a rare form of pulmonary dissemination associated with metastasis of malignant neoplasms, primarily adenocarcinomas. Approximately 6-8% of patients with the malignant disease develop carcinomatous lymphangitis. This condition causes progressive invasion of the pulmonary lymphatic vessels, affecting

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lymphatic circulation and generating respiratory problems. This case details the experience of a patient with ovarian carcinoma and carcinomatous lymphangitis, highlighting the complexity of diagnosis due to nonspecific symptoms and its slow progression. The implications of carcinomatous lymphangitis and its association with different types of cancer are discussed, and the importance of advanced imaging techniques for early diagnosis is emphasized. An analysis of previous cases is presented, highlighting the rarity of the condition and its poor prognosis. Overall, the need for a comprehensive approach to the management of patients with paraneoplastic syndromes is highlighted, especially those associated with respiratory complications such as carcinomatous lymphangitis.

KEY WORDS: Carcinomatous lymphangitis, ovarian cancer, pulmonary lymphangitic carcinomatosis, malignant neoplasia.

INTRODUCTION

Paraneoplastic syndromes are clinical conditions that occur in association with malignant neoplasms located elsewhere in the body, but are not directly caused by the local presence of cancer cells.

The onset and progression of paraneoplastic syndromes are often closely correlated with the underlying malignancy and can affect multiple organ systems. Clinical manifestations range from dermatological changes to life-threatening conditions involving critical systems such as the neurological, endocrine, and respiratory systems. In some cases, these syndromes may be reversible following surgical treatment of the primary neoplasm; however, they can also be irreversible or result in permanent sequelae.

Carcinomatous lymphangitis is a rare form of pulmonary dissemination characterized by the progressive infiltration of pulmonary lymphatic vessels secondary to metastasis from a malignant neoplasm, typically occurring in advanced stages of the disease. Among patients with malignancies, intrathoracic metastases occur in approximately 30 to 40% of cases, with about 6 to 8% developing carcinomatous lymphangitis. [1]

The affected lymphatic vessels undergo infiltrative and inflammatory changes that impede lymphatic drainage from the lungs, resulting in progressive accumulation of interstitial fluid and subsequent impairment of gas diffusion.[2] Carcinomatous lymphangitis most commonly originates from adenocarcinomas of the breast, stomach, lung, pancreas, prostate, cervix, and colon.[3] Intrathoracic metastases occur in approximately 30 to 40% of patients with malignant disease, of whom 6 to 8% develop carcinomatous lymphangitis.[4]

Currently, only a limited number of cases of carcinomatous lymphangitis have been reported in the literature. Clinical manifestations typically include progressive dyspnea, non-productive cough, persistent fatigue, and a sensation of chest tightness, which gradually worsen and may ultimately lead to death. Physical examination often reveals diminished vesicular breath sounds without adventitious noises. Due to these nonspecific features, diagnosis is challenging and requires comprehensive evaluation, including pulmonary function tests (such as spirometry, diffusion capacity, and exercise testing), high-resolution computed tomography, and exclusion of more common conditions such as cardiac disease, pneumonia, pulmonary thromboembolism, asthma, sarcoidosis, and pulmonary fibrosis. Treatment primarily focuses on managing the underlying malignancy through chemotherapy and radiotherapy. Given its rarity, nonspecific symptomatology, and insidious progression, carcinomatous lymphangitis is frequently diagnosed at an advanced stage.

CLINICAL CASE

A 43-year-old female patient, gravida 2, para 2, was initially admitted to the institution following referral for an abdominopelvic ultrasound report describing a mixed cystic-solid mass, measuring 112 × 111 × 105 mm, likely originating from the left ovary.

Additionally, chest computed tomography revealed apparent pulmonary micronodules and signs of a bronchial inflammatory process associated with septal thickening. Distal tomographic sections demonstrated diffuse thickening of the gastric wall and regional lymphadenopathy, raising suspicion for an infiltrative pathological process (Figure 1).

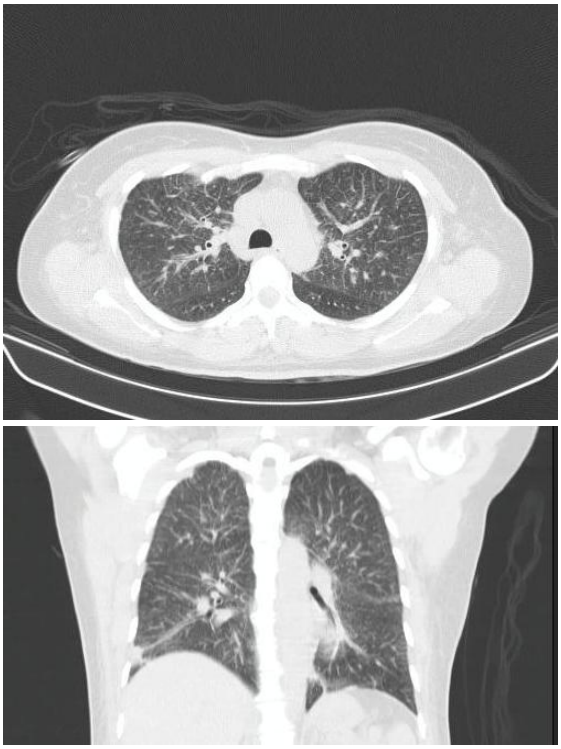

Figure No.1. CT coronal and transverse cuts, own images.

An upper gastrointestinal endoscopy revealed acute erosive esophagitis and atrophic erosive gastritis, likely of autoimmune etiology.

The patient reported a clinical course of approximately two months characterized by dyspnea, cough, orthopnea, chest and back pain, and unintentional weight loss over the past month. She denied any changes in

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bowel habits but reported vaginal bleeding. On physical examination, the patient was in good general condition. Gynecological evaluation included ultrasound, which demonstrated a uterus of normal size with regular contours and homogeneous walls; the endometrium measured 6 mm and appeared homogeneous without evidence of intrauterine pregnancy or retained products of conception. The uterus measured 97 × 73 × 65 mm in longitudinal, anteroposterior, and transverse dimensions, respectively. Two intramural myomas were identified on the anterior uterine wall, measuring 18 × 16 mm and 20 × 15 mm. The right ovary appeared normal, measuring 37 × 25 mm, whereas the left ovary contained a heterogeneous mass measuring 111 × 70 mm. Based on these findings, the patient was admitted to the intramural service, where an exploratory laparotomy was performed. Intraoperatively, a large left adnexal tumor measuring approximately 12 × 15 cm was observed, without evidence of excrescences. The uterus was enlarged and distorted by multiple myomas; the right adnexa appeared normal. There were extensive adhesions between the bladder and anterior abdominal wall, and the retroperitoneum was unremarkable. Given the findings, symptomatic ambulatory management was chosen.

The patient was subsequently re-admitted to the institution due to clinical deterioration. At this time, the thoracic surgery team reviewed the previously obtained chest CT scan and proposed differential diagnoses including carcinomatous lymphangitis, granulomatous diseases, and sarcoidosis. Accordingly, a follow-up chest CT was scheduled three months later to monitor pulmonary parenchymal lesions clinically and radiologically. During the pulmonology evaluation, the presentation was considered consistent with diffuse interstitial lung disease, prompting the need to exclude pulmonary tuberculosis and lymphangitic carcinomatosis. Consequently, a multidisciplinary thoracic approach was undertaken, involving fibro bronchoscopy with bronchoalveolar lavage (BAL) of the lower lobes, thoracoscopy with pulmonary biopsy, and corresponding cultures for mycobacteria and fungi. Additional clinical findings included severe protein-caloric malnutrition, acanthosis nigricans, and secondary anemia. Bronchoscopy examination via an orotracheal approach revealed no endobronchial lesions or bleeding within the tracheobronchial tree; however, the BAL fluid was noted to be turbid. Monoportal video-assisted thoracoscopy identified loose pleural adhesions, congestive multilobar pulmonary parenchymal involvement predominantly in the middle lobe, and nodular lesions on the parietal pleura.

DISCUSSION

Carcinomatous lymphangitis is a rare form of intrathoracic metastasis characterized by infiltrative and inflammatory involvement of the

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pulmonary lymphatic vessels. This condition typically indicates advanced malignancy and is associated with a poor prognosis.

A systematic review and meta-analysis, comprising reports and case series of 139 individual patient events, identified the most frequent primary tumors associated with carcinomatous lymphangitis as breast cancer (17.3%), lung cancer (10.8%), and gastric cancer (10.8%). Secondary involvement originating from ovarian neoplasms was observed in only 2.2% of cases (6). Other reported primary malignancies include colorectal, prostate, pancreatic, cervical, uterine, thyroid, and laryngeal cancers (6).

Lung carcinomatous lymphangitis most frequently coexists with adenocarcinoma, accounting for approximately 80% of cases (7). Pulmonary carcinomatosis secondary to metastasis from gynecological malignancies is a rare occurrence, as is lymph node involvement with carcinomatosis in ovarian cancer, which is reported in less than 1% of cases (8).

In a study investigating the incidence of lung metastases from ovarian cancer involving 357 cases, intrathoracic tumor presence was identified in 169 patients (44.5%) with ovarian cancer, of whom 73% exhibited malignant pleural effusions (8). Furthermore, 12.3% of the patients presented with lung parenchymal metastases, while only 1% demonstrated lymphatic or nodal metastases (8). Over an 11-year period at Samsung Medical Center a tertiary, academic institution and one of the largest cancer centers in South Korea only 27 cases of lymphangitic carcinomatosis associated with gynecological malignancies were reported (9).

An analysis of 255 patients with epithelial ovarian carcinoma demonstrated that 38.0% developed metastases consistent with stage IV disease during the natural history of their illness. Malignant pleural effusions were observed in 24.7% of patients, with a median survival of six months from diagnosis. Pulmonary parenchymal metastases and distant lymph node metastases were each identified in 7.1% of patients (10). In a separate study of 100 ovarian cancer cases with thoracic involvement, pulmonary mass lesions were the most frequent manifestation, occurring in 38% of cases, followed by pleural effusion in 19%, and lymphangitic carcinomatosis in 11% of patients (11).

The diagnosis of metastatic cancer with pulmonary lymphangitic carcinomatosis often requires several months, primarily due to factors related to clinical presentation and imaging challenges. Additionally, individual patient characteristics significantly influence the diagnostic timeline. Most patients present with symptoms such as dry cough and dyspnea resulting from pulmonary tissue involvement, irrespective of the primary tumor origin (12).

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Chest radiography is frequently the initial imaging modality; however, a significant proportion of chest X-rays may appear normal due to the delayed progression of pathological changes (13). Computed tomography (CT), particularly high-resolution CT, is recommended for the evaluation of patients with suspected carcinomatous lymphangitis of the lung, given its higher sensitivity and enhanced ability to facilitate early detection of the disease (14).

CONCLUSION

Dyspnea is a nonspecific symptom with a broad differential diagnosis encompassing a wide range of pathologies. In clinical practice, its significance and the spectrum of potential underlying causes are often underestimated. A comprehensive and systematic approach throughout the various stages of patient evaluation is essential to guide diagnostic reasoning, appropriately interpret ancillary studies, and optimize patient management. Although paraneoplastic syndromes are relatively rare, they must be considered in the assessment of patients with known oncological conditions.

Effective communication and coordination among the various specialties involved in patient care are crucial in managing complex cases, particularly when diagnostic challenges arise and the patient's condition deteriorates. Such interdisciplinary collaboration is essential to consolidate efforts focused on optimizing patient outcomes and to facilitate the consideration of uncommon or atypical etiologies.

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