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CASE REPORT

PRIMARY PULMONARY CHORIOCARCINOMA: A CASE REPORT

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ABSTRACT

Primary pulmonary choriocarcinomais a rare malignant tumor of the lung. It is classified as one of the primary extragonadal choriocarcinomas, which often involve midline structures such as the intracranium, mediastinum, stomach, intestines, retroperitoneum, bladder, and prostate. It has two unique characteristics: it is not associated with abnormal gestation and is not part of a germ cell neoplasm involving the reproductive organs. Its diagnosis is often challenging and treatment is usually ineffective. We present the first case of primary pulmonary choriocarcinoma reported from East Africa and one of only 56 cases reported in the literature.

INTRODUCTION

Primary pulmonary choriocarcinoma (PPC) is a rare and unique trophoblastic malignant tumor of the lung and is classified as one of theprimary extragonadal choriocarcinomas (EGCC). An accurate diagnosis of PPC is usually difficult and challenging, and its treatment is often unsuccessful. We present the first case of PPC reported from East Africa and one of only 56 cases (39 females and 17 males) reported in the literature (1).

CASE REPORT

A 32-year-oldwoman presented with cough, low-grade intermittent fever, and fatigue of 3-months duration.She had weight loss and amenorrhea for the same duration. She was para 2 (3 and 8 year old children). During the month prior to admission, she developed shortness of breath and mild hemoptysis. Chest radiograph, obtained two weeks prior to admission, demonstrated right lower lung opacity (Figure 1).

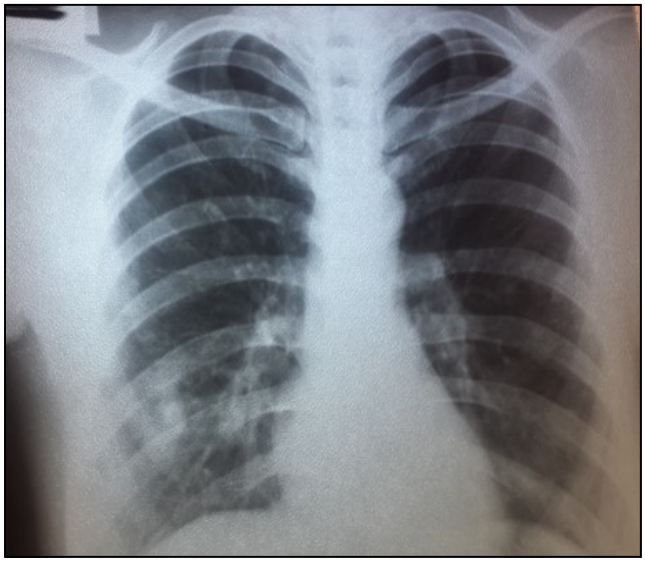

Figure 1. Initial chest radiograph showing right lower lung zone opacity.

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She was admitted to a public hospital where she received ceftriaxone & azithromycin for a presumed severe community acquired pneumonia. She failed to improve and was referred after 15 days to Tikur Anbessa Specialized Hospital (TASH).

On transfer, she was an ill appearing woman in respiratory distress. Vital signs included: pulse rate = 128 beats per minute, blood pressure = 100/70mmHg, respiratory rate = 40breaths per minute, temperature (T^o) = 36.5^{o}C.Chest examination revealed bilateral diffuse rales.

Complete blood count (CBC), liver function tests, coagulation profiles, lactic dehydrogenase (LDH) and blood culture were all unremarkable.

Urine hCG (x2), serum HIV, ANA and ANCA were negative. Chest radiograph and CT (Figure 2) on transfer showed multiple bilateral nodular opacities, right mid-lung mass, and airspace disease at the lung bases.

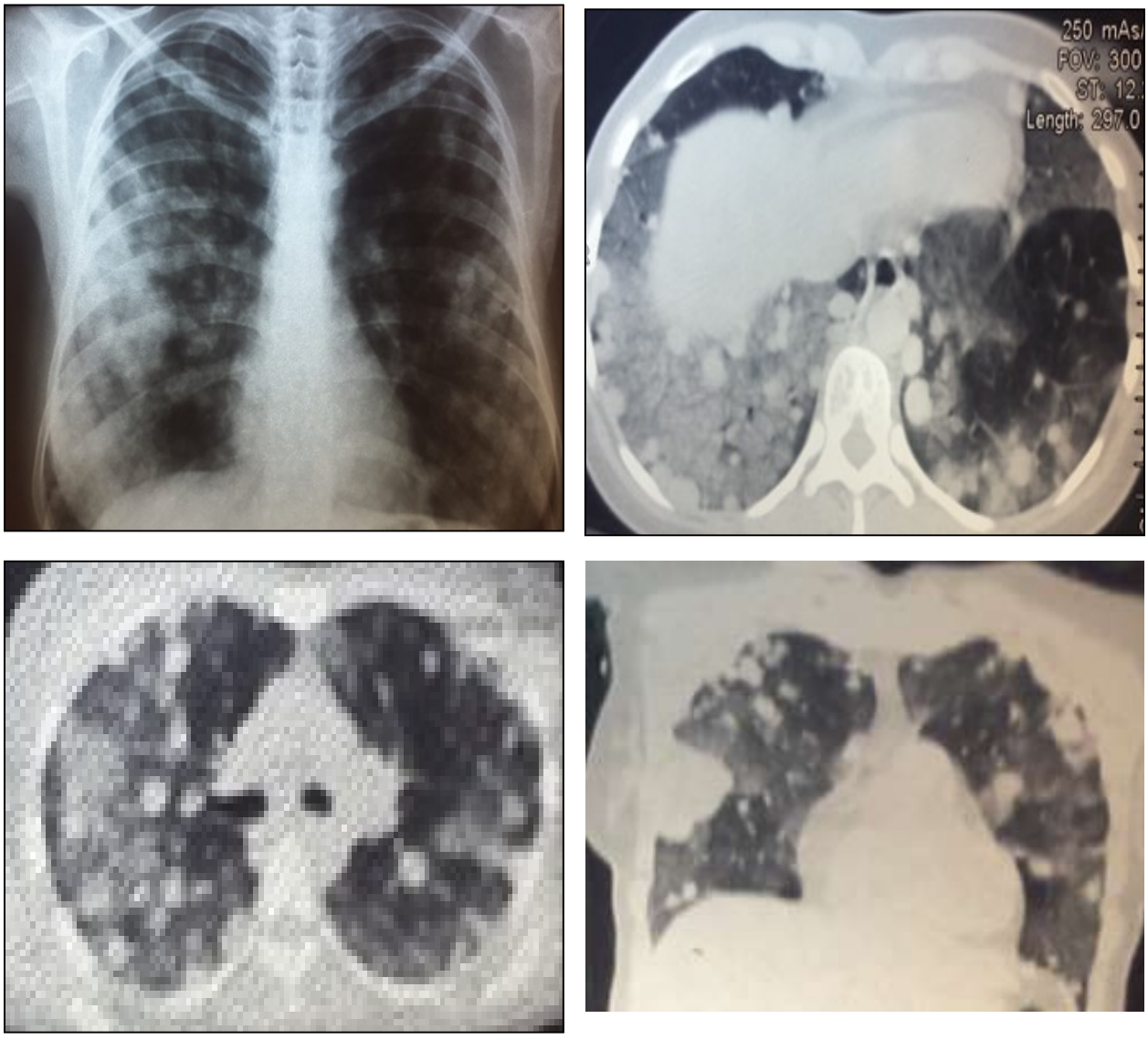

Figure 2. Left upper panel (Chest radiograph): bilateral
nodular and right midlung zone homogeneous opacities; right upper and lower panels:multiple bilateral nodular opacities, right mid-lung mass, and airspace disease at the bases (Chest CT).

Additionally, she had a normal abdomino-pelvic ultrasound, abdomino-pelvic CT scan, and brain and spinal MRI scans. Alpha fetoprotein, carcinoembryonic antigen, and CA-125 were within normal limit. Serum β-hCG level was markedly elevated at 19,950 mIU/ml and increased after 2 days to235,000 mIU/ml.

Ultrasound guided fine needle aspirate revealed clusters of cells with large irregular nuclei consistent with carcinoma. With the elevated serum β-hCG level, a diagnosis of PPC was made. Subsequently, she developed massive hemoptysis and severe hypoxemia despite high flow of supplemental oxygen. She was intubated and placed on mechanical ventilation. Broad-spectrum antibiotics for possible superimposed hospital acquired pneumonia were initiated. Chemotherapy using a regimen of bleomycin, etoposide and cisplatinwas started. She had continuous massive endotracheal bleeding which was difficult to arrest. She died after 10 days of ICU admission and 2 days of starting chemotherapy initiation.

DISCUSSION

Primary pulmonary choriocarcinomas a subtype of EGCC that is entirely localized to the lung (2).A higher prevalence of PPC occurs in the second and third decades of lifewith a median age of 34 years (1,2).

The mechanism of development of PPC is not certain; abnormal migration of embryonic germ cells, metastatic tissue from a regressed primary gonadal tumor or a molar pregnancy, or cellular transformation of lung to trophoblastic tissue have all been reported (2-4).

Patients with PPC often present with non-specific clinical features. Chest pain, non-productive cough, hemoptysis, and dyspnea are frequent manifestations (5).Woman may also have hormonal problems such as amenorrhea or vaginal bleeding. In males, signs of feminization including gynecomastia, testicular atrophy, and loss of libido may be seen(6).

An accurate diagnosis of PPC is usually difficult and challenging. Metastatic choriocarcinoma to lung is frequent and an elevated serum β-hCG can be seen as a paraneo plastic finding with lung cancer. However, extreme serum β-hCG elevation as in this case is not expected(7).

Chest radiographs and chest CT scans may show a well-defined, centrally located, solitary or lobulated opacity. Rarely, multiple nodular lesions are found, resembling metastases. Other extra-thoracic imaging studies are important to rule out a primary gonadal focus of choriocarcinoma (8).

Grossly PPCs are large, well circumscribed, hemorrhagic and partly necrotic tumors. They may involve the mainstem bronchi. Diagnosis can be confirmed by positive immunohis to chemical tissue staining for β-hCG and negative for TTF-1(9).

PPCs have a poor prognosis and show limited response to treatment. There are no established treatment guidelines due to the tumor's rare occurrence and often advanced stage at diagnosis. However, surgical resection with neo-adjuvant cisplatin based chemotherapy is preferred and reasonable. Radiation therapy has a limited role as the tumor is not radiosensitive (1,10).

Conclusion: We report the first case of PPC from East Africa. This case illustrates the need for clinicians to consider PPC in the differential when they encounter a young patient with lung nodules or masses and no obvious primary tumor site.

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