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CASE REPORT

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A rare case of choriocarcinomatous pulmonary embolism revealed by ischemic strokes

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ABSTRACT

Choriocarcinoma is a malignant tumor with high metastatic potential belonging to the spectrum of gestational trophoblastic tumors, which can be of germ-cell origin (nongestational) or gestational. Choriocarcinomatous pulmonary embolism represents a rare and aggressive extra-gonadal form of gestational trophoblastic disease, most often occurring after a normal or pathological pregnancy. The presentation is similar to that of cruoric embolism, but it is characterized by local invasion and significant hematogenous dissemination leading to distant metastases. Increased human chorionic gonadotropin hormone (hCG) levels are suggestive of the diagnosis. We report a case of choriocarcinomatous pulmonary embolism involving the inferior lobar artery of the left lung in a 44-year-old female patient admitted for multiple ischemic strokes. The diagnosis of choriocarcinoma was done on a computed tomography scan in the presence of high levels of hCG contrasting with negative pelvic imaging and a history of previous spontaneous abortion. The outcome was favorable after chemotherapy which was an additional argument in favor of the diagnosis of choriocarcinoma.

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Pulmonary embolism; choriocarcinoma; ischemic strokes; human chorionic gonadotropin hormone (hCG).

Introduction

Choriocarcinoma is an aggressive form of gestational trophoblastic neoplasia in most cases arising in the uterus, this highly malignant epithelial tumor is characterized by a proliferation of cytotrophoblasts and syncytiotrophoblasts. It has been reported to occur in 1 to 50,000 pregnancies in patients during childbearing age with previous complete hydatidiform moles, or after nonmolar pregnancies (normal gestation, ectopic pregnancy, or abortion). The primary or secondary choriocarcinomatous involvement of the pulmonary arteries is associated with important morbidity and mortality. This unusual presentation of choriocarcinoma is underdiagnosed on imaging because of its non-specific semiology, which makes it difficult to distinguish it from thromboembolic disease and other malignant vascular tumors (sarcomas). This requires the use of a set of clinical and biological arguments. Primary choriocarcinoma of the pulmonary artery without any evidence of placental involvement has been rarely reported in literature we report the case of a choriocarcinomatous pulmonary embolism revealed by multiple ischemic strokes in a 44-year-old patient.

Case Report

A 44-year-old female patient, with no particular history of cardiovascular disease and a spontaneous abortion 3 years prior to her admission to the Department of Neurology for multiple and recurrent ischemic strokes.

A computed tomography (CT) scan performed 1 month before her admission showed multiple systematized supratentorial ischemic lesions of varying stages involving the territories of the middle cerebral arteries with areas of hemorrhagic transformation (Fig. 1).

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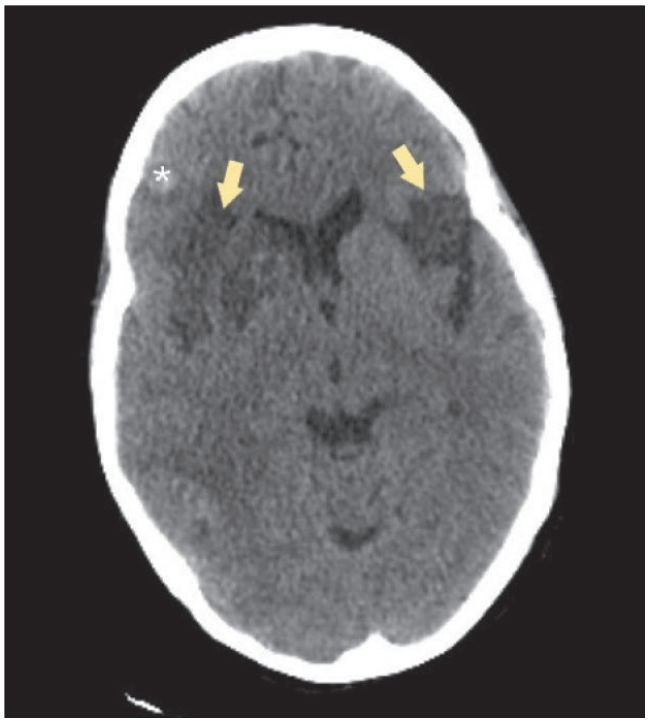


Figure 1. A non-contrast axial CT scan of the brain reveals bilateral frontal cortico-subcortical hypodensity areas (arrows) with a pattern suggestive of ischemia, affecting the territories of the middle cerebral arteries. Furthermore, there is evidence of a hemorrhagic transformation in the right frontal region (*).

The etiological investigation included echocardiography, an ambulatory electrocardiogram (ECG) monitoring (Holter ECG), a CT angiography of the supra-aortic arteries and the circle of Willis, and a thrombophilia testing (Protein S and C, antithrombin III, and factor V Leiden), which all came back normal.

The laboratory tests showed no signs of inflammation, and the antibodies tested for, including anti-phospholipid antibodies, antinuclear antibodies, and anti-neutrophil cytoplasmic antibodies, were all negative. However, a significant elevation in human chorionic gonadotropin (β-hCG) levels at 55, 000 IU/l (Normal <5 IU/l) with an increasing trend led us to consider a possible germ cell tumor.

To explore the possibility of a gestational trophoblastic tumor as the underlying cause, the patient underwent pelvic exploration using ultrasound and magnetic resonance imaging (MRI) following a standard protocol, including T2 sequences in all three planes, T1 sequences with and without fat saturation, diffusion-weighted imaging with apparent diffusion coefficient mapping, and dynamic gadolinium injection in axial and sagittal planes. This examination did not reveal any pelvic mass (Fig. 2a and b).

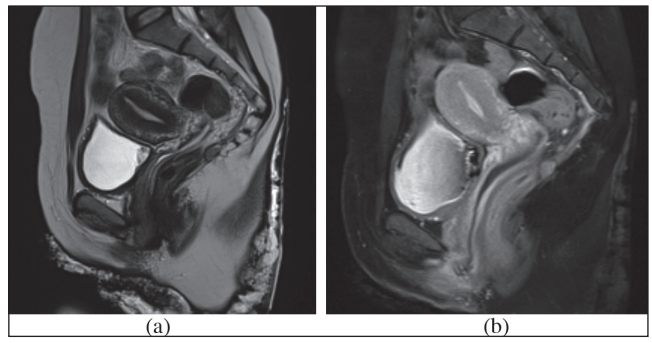


Figure 2. MRI with T2-weighted sequence in the sagittal plane (a) and T1-weighted sequence with Gadolinium contrast injection after fat saturation in the sagittal plane (b) does not reveal any endocavitary mass.

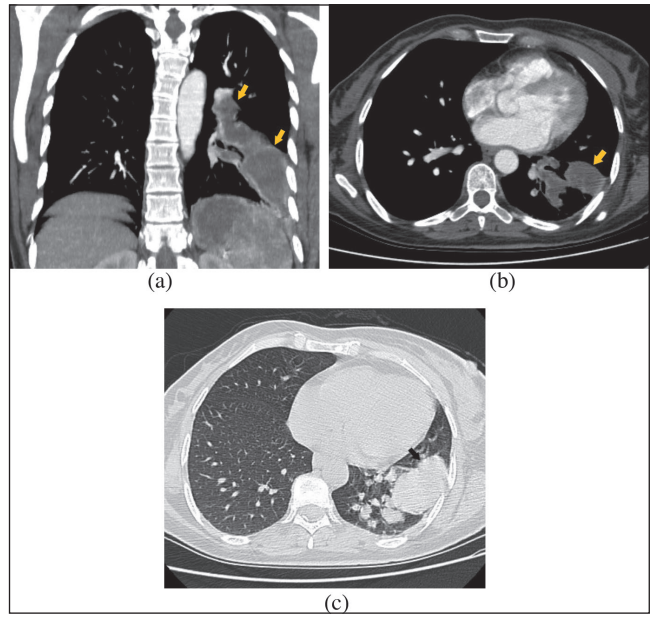


Figure 3. Coronal (a) and axial (b–d) sections of the thoracic CT angiography showing an obstructive and expansive endoluminal material within the inferior left lobar artery, extending to its segmental branches (yellow arrows). The dilation is primarily observed in the laterobasal branch with trans-lobar extension (black arrow). Endoluminal material is seen in the left inferior pulmonary vein (red arrow).

A thoracoabdominal CT was performed to investigate potential extra-pelvic involvement. It revealed a fusiform aneurysmal dilation of the inferior lobar artery of the left lung. Within this dilation, there was a hypodense and heterogeneous endoluminal content that completely obstructed the artery. The dilation extended to the segmental and sub-segmental branches and was outlined by contrast medium. The compression of bronchial structures was most prominent at the lateral basal branch with translobar extension at that level (Fig. 3a–c).

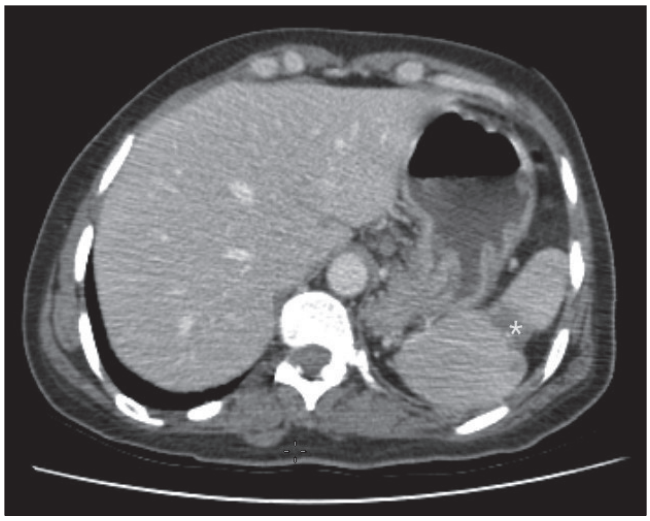


Figure 4. Axial contrast-enhanced abdominal CT scan showing a hypodense, wedge-shaped non-enhancing mid-splenic lesion (*), accompanied by capsular retraction suggestive of an ischemic origin.

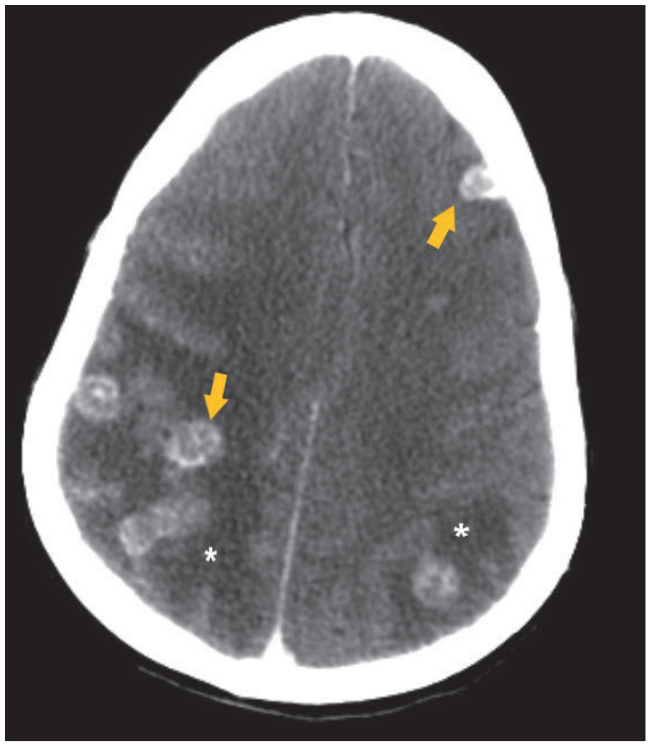


Figure 5. Axial contrast-enhanced brain CT scan revealing multiple bilateral supratentorial lesions with a ring-like enhancement pattern (arrows), accompanied by areas of vasogenic edema (*).

Furthermore, partially obstructive material was observed within the adjacent left inferior pulmonary vein next to the aforementioned endoluminal mass. This finding suggests an embolic origin for the ischemic strokes (Fig. 3d). Additionally, there was no involvement or damage to the lung parenchyma,

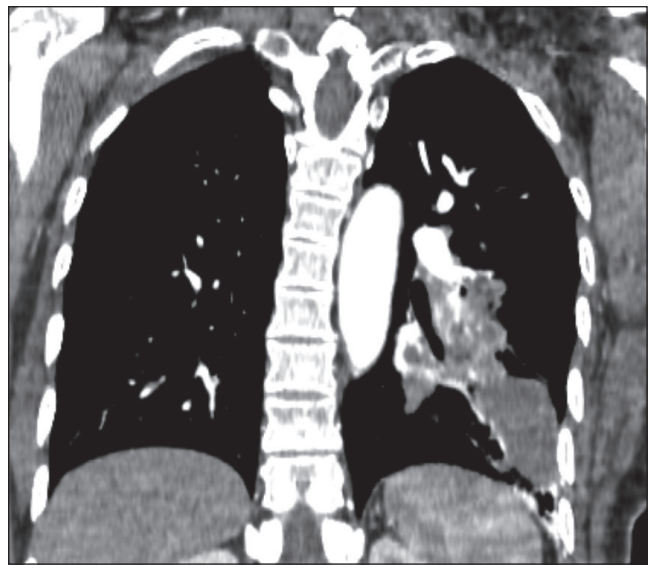


Figure 6. Follow-up thoracic CT scan after 3 months in the coronal section, demonstrating partial regression in size of the endoluminal mass.

and no significant mediastinal lymphadenopathy was observed.

The CT of the abdomen revealed a hypodense, wedge-shaped non-enhancing mid-splenic lesion. It caused capsular retraction, suggestive of an infarction, without any detectable abnormalities in the splenic pedicle (Fig. 4).

Due to the worsening headaches and behavioral changes, a brain CT scan was performed. It showed the presence of multiple enhancing intra-axial supratentorial lesions with a ring-like enhancement pattern, surrounded by significant perilesional edema in a glove-like distribution. These findings suggested a secondary origin (Fig. 5).

The combination of pulmonary embolism, elevated β-hCG levels without pelvic involvement, history of gestational events, and secondary brain lesions strongly suggested metastatic choriocarcinomatous pulmonary embolism. Certain somatic tumors, like lung adenocarcinoma, can exhibit choriocarcinomatous differentiation and also produce β-hCG but they typically present as a primary lung tumor, the patient may not have a history of gestational events. Hence, the importance of pathological examination is to distinguish between the two entities.

This case was presented and discussed in a multidisciplinary meeting, where a transbronchial biopsy was suggested for confirmation. However, considering the invasive nature of the procedure and the patient's debilitated condition, it was deemed potentially lethal and life-threatening, and therefore, it was not performed.

As a result, the diagnosis was established based on the increased levels of β-hCG, the metastatic nature of the condition, and the history of spontaneous abortion, following the criteria formulated by the International Federation of Gynecology and Obstetrics and the guideline of the European Society for Medical Oncology.

A polychemotherapy regimen was initiated following the protocol of etoposide, methotrexate, actinomycin D, cyclophosphamide, and vincristine. After two cycles of treatment, there was a notable reduction in the size of the endoluminal tumor on the follow-up imaging after 3 months (Fig. 6).

This positive response was accompanied by a decline in β-hCG levels, further supporting the initial diagnostic hypothesis. Despite this therapeutic approach, the patient passed away due to intracranial hypertension 4 months after the initial diagnosis.

Discussion

Choriocarcinoma is a rare malignant tumor belonging to the group of gestational trophoblastic tumors, characterized by excessive secretion of choriogonadotropin hormone (β-HCG) [1,2]. Although various extra-gonadal sites have been reported, it appears to have a predilection for the retroperitoneum, mediastinum, and skull.

Embolism remains one of the most exceptional presentations of extra-gonadal choriocarcinoma, and its pathophysiology is not fully understood. It can occur in isolation or as part of metastatic disease [1,2].

First described in 1959 by Bagshawe and Brooks, choriocarcinomatous pulmonary embolism represents a diagnostic and therapeutic emergency that can be potentially serious or even fatal due to pulmonary hypertension and its high metastatic potential [2].

Several hypotheses have been proposed to explain the etiopathogenesis of this localization. One of them suggests a malignant transformation of villous trophoblasts that entered the pulmonary artery during abortion or childbirth. However, others consider that choriocarcinoma of the pulmonary artery results from metastatic dissemination of a spontaneously regressive uterine tumor [1].

The clinical presentation is nonspecific, and the most common signs resemble those of thromboembolism, including progressive respiratory distress, chest pain, hemoptysis, and occasionally right-sided heart failure. In rare cases, multifocal metastatic involvement may be a presenting feature [3].

The diagnosis is confirmed by significantly elevated levels of β-HCG hormone secreted by the trophoblastic tissue. Based on Seckl's research, it is not necessary to obtain a histological diagnosis before initiating chemotherapy, especially considering the substantial morbidity associated with surgery. This implies that chemotherapy can be started based on clinical and biochemical indications of choriocarcinoma, such as elevated levels of the β-HCG hormone, without the requirement for histological confirmation. This approach helps mitigate the potential complications and risks associated with surgical interventions.

On CT, choriocarcinomatous embolism presents as a hypodense endoluminal mass that is centrally or marginally located, proximal, unilateral, and completely obstructive. The mass has lobulated or irregular contours and shows little to no enhancement after contrast medium injection. It extends to the lobular and segmental branches and often demonstrates transmural extension with hypoperfusion of the underlying pulmonary parenchyma.

Unlike thromboembolism, this mass tends to dilate the vessels and attaches to the arterial wall at an acute angle with resistance to anticoagulant treatment. The formation of pseudoaneurysms has also been reported in advanced cases [4].

While the diagnostic utility of CT pulmonary angiogram is limited, the described lesion characteristics raise suspicion of malignant vascular tumors of the pulmonary artery, mainly sarcomas and metastases. Sarcomas of the pulmonary artery exhibit a similar imaging pattern; however, contrast enhancement and transmural invasion are frequently observed. Unlike thromboembolism, sarcomas demonstrate increased uptake of 18-FDG in positron emission tomography.

Another favored diagnosis is secondary endovascular involvement, although it tends to occur in the segmental and sub-segmental branches, resulting in bead-like dilations.

Like all gestational trophoblastic neoplasms, choriocarcinoma cells have a rapid doubling time, explaining their sensitivity to chemotherapy. Therefore, early management, as recommended by guidelines from the European Society for Medical Oncology, does not always require histological confirmation to prevent local and distant dissemination [5].

A polychemotherapy is proposed, achieving a remission rate of over 89% for non-metastatic forms. However, surgery and circulatory assistance (extracorporeal membrane oxygenation) are reserved for cases with hemodynamic instability.

This treatment approach requires regular radiological and hormonal monitoring through weekly measurement of plasma β-HCG levels [1].

Conclusion

Choriocarcinoma-associated pulmonary embolism is an exceptional and potentially curable presentation. It should be suspected in imaging studies of reproductive-age patients presenting with an expansive, totally obstructive, contrast-enhancing thrombus resistant to anticoagulant treatment. The radiological appearance is nonspecific, necessitating the use of β-HCG measurement and pathology, which represent the cornerstone of diagnosis. This condition is associated with significant morbidity and mortality.

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