EXUDATIVE RETINAL DETACHMENT CAUSED BY METASTATIC CHORIOCARCINOMA TO THE CHOROID

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Abstract—Background: Choriocarcinoma is an aggressive malignant tumor. It commonly invades the uterus, causing necrosis and hemorrhage through vascular invasion. Metastasis is also common, with the main areas affected being the lung, brain, liver, pelvis, vagina, spleen, intestine, and kidney. Objectives: To illustrate the presentation of a rare disease, metastatic choriocarcinoma to the eye, causing a retinal detachment, and to elucidate the characteristics and treatment of that rare disease. Case Report: A 23-year-old woman presented to our Emergency Department with a left painful red eye with decreased visual acuity and was subsequently diagnosed with choriocarcinoma with metastasis to the lungs, brain, and choroid, causing a left exudative retinal detachment. Conclusion: Gestational choriocarcinoma with metastasis to the eye is a rare disease and a rare cause of exudative retinal detachment. In general, metastatic choriocarcinoma is highly curable with chemotherapy. However, metastasis to the eye, regardless of the type of tumor, portends a poor prognosis.

Keywords—choriocarcinoma; metastasis; choroid; retinal detachment

INTRODUCTION

Gestational choriocarcinoma is a highly malignant tumor that can arise from any type of trophoblastic tissue such as molar, ectopic, or intrauterine pregnancy. Histologically, choriocarcinoma is composed of syncytiotrophoblasts and cytotrophoblasts, and the appearance of those cells is essentially pathognomonic of choriocarcinoma. Approximately 50% of choriocarcinoma cases are preceded by a hydatidiform mole, 25% follow a normal term pregnancy, and 25% follow an ectopic pregnancy or spontaneous abortion. Choriocarcinoma is a rare disease, with an incidence of 1 in 40,000 pregnancies and 1 in 40 hydatidiform moles in Western countries. In the past, choriocarcinoma with metastasis had a mortality rate approaching 100%, but more recently, cure rates are >90%, even with widespread metastasis (1).

CASE REPORT

A 23-year-old Jamaican woman presented to our Emergency Department with a complaint of painful, red left eye with blurry vision. Physical examination of the affected eye revealed periorbital edema, conjunctival injection, normal cornea, presence of afferent pupillary defect and visual acuity positive for only gross objects, unable to discriminate number of fingers, in all fields. The fundus could not be adequately visualized. Visual acuity and examination of the other eye were normal. Ophthalmology was consulted and she was diagnosed with left exudative retinal detachment. Given the exudative nature of the retinal detachment, their suspicion was for a rheumatologic etiology.

Upon review of systems, the patient reported several other complaints which she deemed irrelevant to her current eye issue. She reported one episode of minor hemoptysis the previous week, with several more minor episodes within the past few months. She also reported...
a history of irregular menses for many years, with her last normal menses 6 months prior. To the best of the patient’s knowledge, she had never been pregnant. The urine beta-human chorionic gonadotropin (hCG) was positive during this visit. Beta-hCG serum quantitative level was 120,074 IU/L. A bedside transvaginal ultrasound was performed with no visualization of intrauterine pregnancy and no visualized ectopic pregnancy. An official transvaginal ultrasound performed by the Department of Radiology revealed no evidence of ectopic or intrauterine pregnancy, and heterogeneous endometrium with some questionable cystic areas. It was reported that this could represent a small amount of blood clot related to a missed abortion vs. trophoblastic disease.

These findings raised concern for gestational trophoblastic neoplasia (GTN) with metastasis to the lungs causing the hemoptysis, and to the eye causing the exudative retinal detachment. The patient received a non-contrast head, chest, abdomen and pelvis computed tomography (CT) scan that demonstrated multiple bilateral hyperdense masses in the high frontal lobes and right cerebellar hemisphere consistent with metastatic disease to the brain. A retinal detachment of the left eye was noted and deemed secondary to metastatic disease (Figure 1). There were also multiple pulmonary nodules consistent with metastatic disease to the lungs.

Many medical specialties became involved, including Gynecologic Oncology, Medical Oncology, Neurosurgery, and Radiation Oncology. The gynecology team further noticed two friable 0.5-cm exophytic lesions on the right lateral fornix of the vagina, concerning for metastatic disease to the vagina. A decision was made to defer dilation and curettage due to concern for high risk of bleeding, and to proceed directly to chemotherapy. A chemotherapy regimen was agreed upon by Medical Oncology and Gynecologic Oncology that included etoposide, methotrexate, folinic acid, actinomycin D, cyclophosphamide, and vincristine; this regimen is referred to as the EMA-CO regimen. The patient was also started on high-dose dexamethasone to reduce cerebral edema. Before chemotherapy was initiated, the patient received a magnetic resonance imaging (MRI) scan of the brain, which showed multiple hemorrhagic metastases seen in the bilateral frontal lobes and right cerebellar hemisphere. MRI of the cervical, thoracic, and lumbar spine showed no metastasis to the spine.

After 7 days of inpatient evaluation and treatment, the patient was discharged with scheduled follow-up in the oncology clinic the next day and re-admission 1 week later for the second cycle of chemotherapy. On the day of discharge, the patient’s beta-hCG had significantly decreased from the day of admission to 52,514 IU/L. The patient was also referred to Radiation Oncology at an affiliate institution for further evaluation. Radiation Oncology at our institution had deemed the patient not suitable for stereotactic radiation therapy, and whole-brain radiation therapy was deferred during this hospitalization.

DISCUSSION

Choriocarcinoma is one of three diseases categorized under the term gestational trophoblastic neoplasia. Before the advent of chemotherapy, choriocarcinoma had a mortality rate near 100% when metastatic disease was present and near 60% after hysterectomy for nonmetastatic disease (1). Chemotherapy has completely changed the mortality of choriocarcinoma, as now even metastatic choriocarcinoma has a >90% cure rate.

Choriocarcinoma most commonly metastasizes to the lung, brain, liver, pelvis, vagina, spleen, intestine, and kidney. Metastatic choriocarcinoma to the eye is very rare. Our literature search shows only six reported cases of gestational choriocarcinoma with metastasis to the eye, and this entity has not been previously reported in the Emergency Medicine literature (2–7).

As opposed to the most common forms of retinal detachment that occur as a result of a retinal tear or traction on the retina, exudative retinal detachment is related to accumulation of fluid beneath the retina. This entity is most commonly associated with inflammatory conditions, but is sometimes related to choroidal neoplasm or metastasis (8). The management of exudative retinal detachment caused by neoplasm is treatment of the neoplasm.
Most malignant neoplasms of the eye are of a metastatic origin, with breasts and lungs being the most common primary sources. The most common signs and symptoms of choroidal metastasis are conjunctival injection, worsening of vision, visual field defects, retinal detachment, and pain. Regardless of the type of tumor, metastasis to the eye generally portends a poor prognosis. According to two studies, median survival for patients with choroidal metastasis from all sources is 6 to 7 months from time of diagnosis (9,10).

In 2002, the International Federation of Gynecology and Obstetrics developed a staging system for GTN that incorporated the World Health Organization risk factor scoring system for GTN. According to the modified risk-stratification system, our patient was Stage IV score >7, the highest risk category in the system, based on the number, location, and size of metastases and other prognostic factors, including the level of the serum beta-hCG.

The treatment for metastatic choriocarcinoma is first and foremost chemotherapy. Patients with non-metastatic (Stage I) and low-risk metastatic disease (Stage II and III score <7) are treated with single-agent therapy, usually methotrexate. Patients classified as having high-risk metastatic disease (Stage IV and Stage II-III score >7) are generally treated with multiple chemotherapy agents with or without radiation or surgery (11).

Most recent literature suggests that patients undergoing a multiagent regimen of EMA-CO have complete response rates of 71–78% and long-term survival rates of 85–94% (12–18).

In respect to gestational choriocarcinoma with choroidal metastasis, the six published case studies reported only two deaths. Admittedly, most of the articles did not mention how far removed from the date of diagnosis the patients were at the time of publication. Our patient is currently 1 month removed from her diagnosis; however, there are too few cases of patients with gestational choriocarcinoma with metastasis to the eye to accurately predict the prognosis of her survival.

CONCLUSION

Gestational choriocarcinoma with metastasis to the eye is a rare cause of exudative retinal detachment. Ophthalmologic findings consistent with exudative retinal detachment combined with symptoms of hemoptysis and amenorrhea led to further diagnostic testing in this case. Elevated beta-hCG, transvaginal ultrasound suggestive of trophoblastic disease, and final confirmation on CT scan ultimately confirmed the diagnosis of metastatic gestational choriocarcinoma.

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REFERENCES