Off-midline retroperitoneal choriocarcinoma presenting as neurologic symptoms

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Summary

A 28-year-old woman suffered from frequent headaches. She had a history of a dilatation and curettage for hydatidiform moles. This admission showed markedly elevated levels of human chorionic gonadotropin (hCG) and lactate dehydrogenase. Brain MRI showed a hemorrhagic mass in the left temporal area, with rapid growth. Histology of tumors obtained from multiple areas including retroperitoneum was consistent with choriocarcinoma.

Key words: Brain; Choriocarcinoma; Computed tomography (CT); Magnetic resonance imaging (MRI), Retroperitoneum.

Introduction

Choriocarcinoma has been reported to occur in 1 in 40,000 pregnancies [1]. It can present with variable clinical symptoms owing to the tendency to metastasis. We report the case of a 28-year-old woman with an initial presentation of neurological symptoms of an off-midline retroperitoneal mass.

Case Report

A 28-year-old woman suffered from frequent headache almost every day, but the symptoms could be controlled by painkillers. One month before admission, the symptoms had become worse, with intractable headache, nausea and vomiting that could not be relieved by medication. She had recent chest tightness.

The menstrual cycle of the patient had become irregular two years after this clinical manifestation. She had been pregnant three times, with two live births and one termination. Five years earlier, she received dilatation and curettage for hydatidiform moles. The level of human chorionic gonadotropin (hCG) had returned to the normal range after one year, without medication. The last menstrual period was two weeks before the presentation. As her pregnancy test was positive, she had transvaginal ultrasound, which showed no detectable mass in the uterus or adnexa. Her vital signs were normal. Laboratory test results revealed a markedly elevated level of hCG (185839m IU/ml) and lactate dehydrogenase (700 IU/ml).

During this admission, chest radiograph revealed a soft-tissue mass, measuring 11.5 × 9 cm, in the left upper lung zone, with pleural attachment. Echo-guided biopsy of the chest showed anaplastic carcinoma. Magnetic resonance imaging (MRI) of the brain showed a mass, 1.6 × 2.5 × 1.7 cm, in the left temporal area (Figure 1A). The tumor showed gradient-echo darkness and perifocal edema (Figure 1B), indicating focal hemorrhage. The mass had a high signal on T1-weighted images, a low signal on T2-weighted images, and heterogeneous enhancement on postcontrast images (Figure 1C).

On the 17th day after admission, she became drowsy. Computed tomography (CT) scan of the brain showed rapid growth of the mass (Figure 1D), 2.7 × 3 × 2.5 cm, with heterogeneous density in the left temporal lobe with prominent perifocal edema and a mass effect caused compression of the left lateral ventricle, and impending uncal herniation. CT of the abdomen showed a loculated hypodense mass, 9 × 9.1 × 7.5 cm, with central necrosis and peripheral intense enhancement, located in the infrahepatic region, with transverse abdominal muscle involvement (Figure 1E). There was no demonstrable tumor in the uterus. Diagnostic dilatation and curettage revealed only hyperplastic changes.

The patient received craniotomy with total removal of the brain tumor. A dark brownish blood clot was mixed with grayish tumor, and a well-encapsulated tumor at the temporal tip of the left petrosal bone. Microscopy showed choriocarcinoma with blood lakes surrounded by nests of neoplastic cells characterized by pleomorphic nuclei, prominent nucleoli, and amphophilic cytoplasm, and scattered multinucleated cells. The tumor represented syncytial growth and marked central hemorrhagic necrosis (Figure 1F). The tumor cells were positive for hCG.

The patient received a chemotherapy regimen (bleomycin, etoposide, and cisplatin) three days after the brain surgery. After three courses of chemotherapy, the level of hCG was 7881 mIU/ml and the left lung tumor had become smaller.

The patient then underwent left upper lobectomy of the lung, wedge dissection of the left lower lobe and lymph node dissection. Histology revealed choriocarcinoma. There was no invasion at the hilar and mediastinal lymph nodes.

After that this patient received the fourth course of chemotherapy. The level of hCG decreased to 56183 m IU/ml but then the level persisted and increased slightly. A simple hysterectomy was performed first. Then the patient had a radical right hemicolectomy, which included the right retroperitoneal tumor, partial hepatectomy (segments 3, 5, and 6), partial resection of the inferior wall of the third portion of the duodenum, and ligature of Batson's plexus. Histology of the uterine specimen showed no choriocarcinoma. The specimens of hepatic tumors and the retroperitoneal mass had total necrosis, and the specimens of the abdominal and duodenal walls showed infiltration by choriocarcinoma cells (Figure 1G).

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Fig. 1a

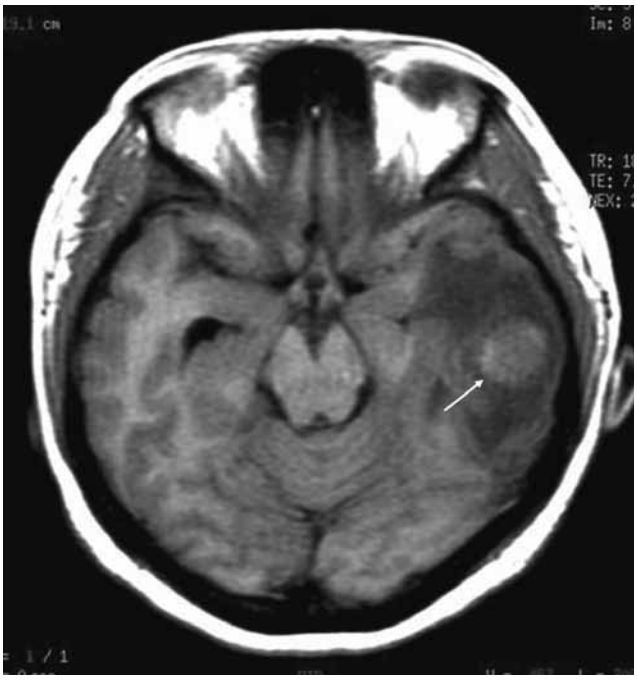


Fig. 1b

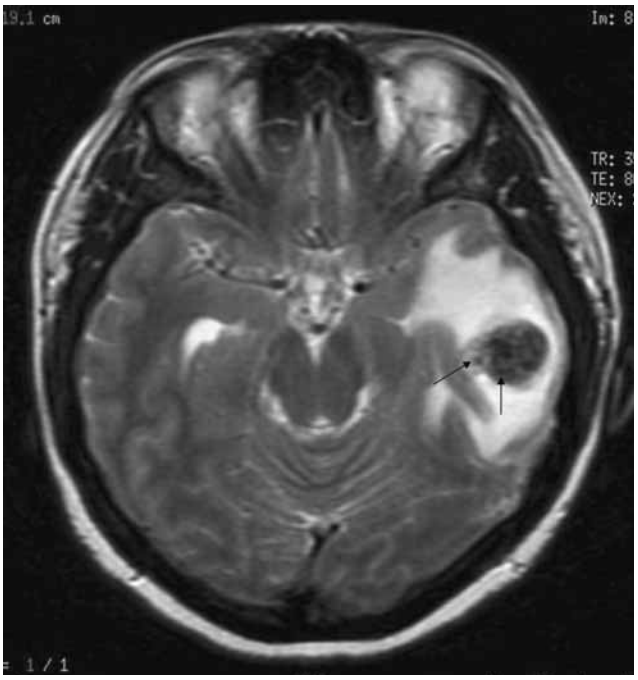


Fig. 1c

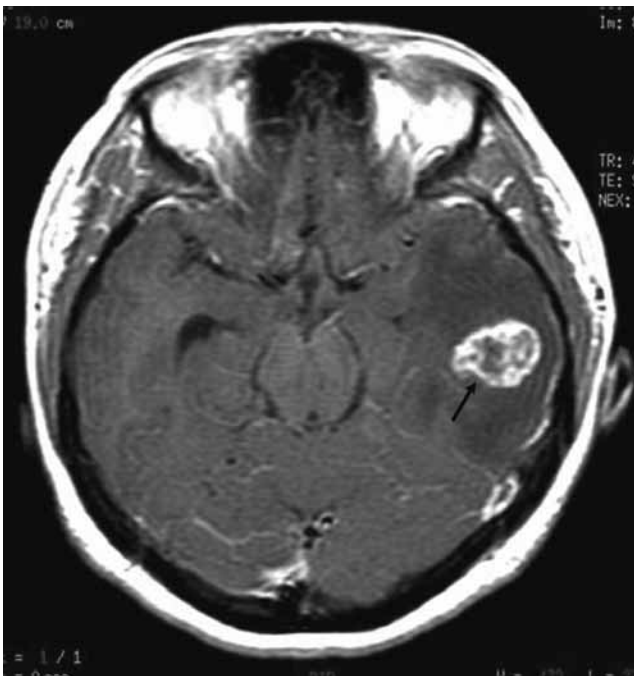


Fig. 1d

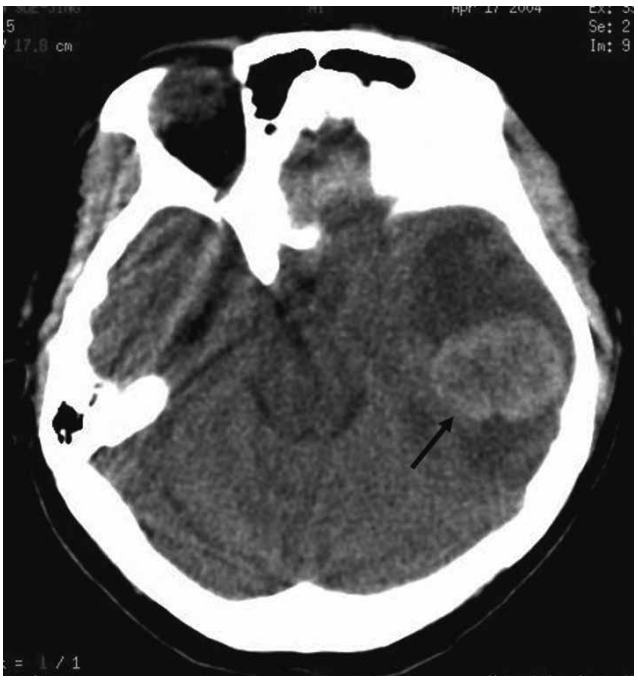


Figure 1. — A 28-year-old woman suffered from frequent headaches.

(A) Axial T1-weighted (TR 1823/TE 7.7) MRI of the brain shows a mass in the temporal lobe with central isointensity and high signal intensity peripherally (arrow).

(B) Axial T2-weighted image (TR 3500/TE 86.5) shows low signal intensity in the central part of the tumor with extensive perifocal edema and spotty dots of dark signal intensity peripherally (arrows). The dark dots become darker on gradient-echo images, suggestive of hemorrhage. Hemorrhage was proven by histology.

(C) Gadolinium-enhanced axial image shows irregular enhancement (arrow), mainly at the periphery of the tumor.

(D) Follow-up CT scan of the brain 17 days after admission shows rapid growth of the tumor (arrow).

Fig. 1e

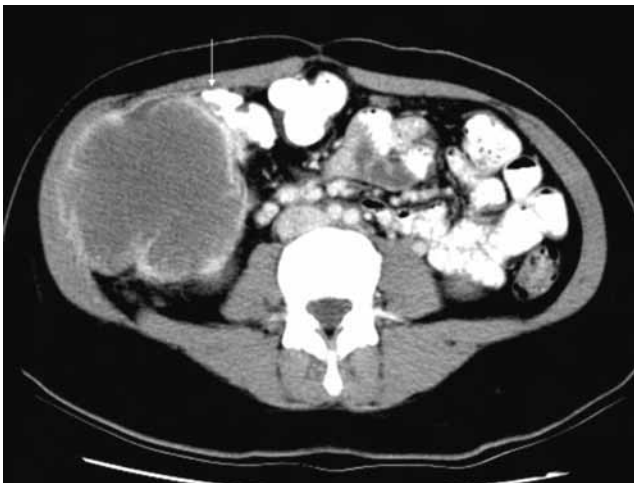


Fig. 1f

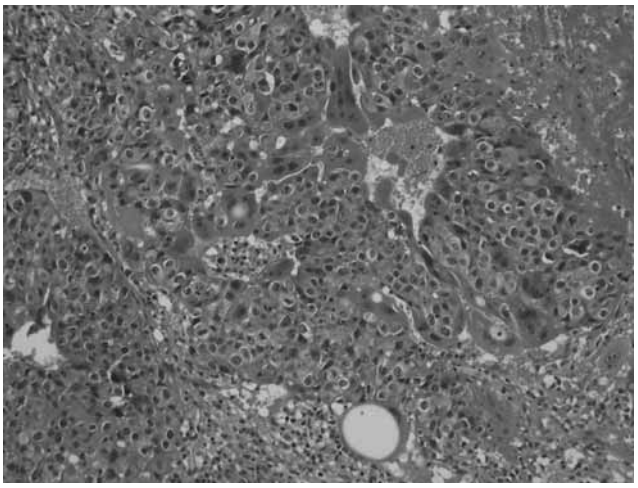
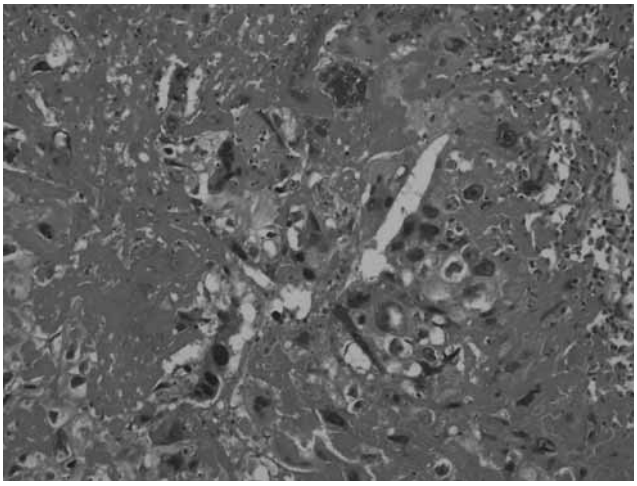


Fig. 1g



(E) Postcontrast abdominal CT scan shows a central necrotic tumor with irregular peripheral enhancement. The tumor invades the abdominal wall and anteriorly displaces the ascending colon (arrow), indicating its location in the retroperitoneum.

(F) Photomicrograph of the specimens obtained from the brain and duodenum. Metastatic choriocarcinoma of the brain with nests of tumor cells showing pleomorhpic nuclei, prominent nucleoli, and amphophilic cytoplasm, intermingled with scattered multinucleated tumor cells. (H & E × 200).

(G) Choriocarcinoma cell infiltration admixed with blood clots in the serosa of the duodenum. (H & E × 200).

Discussion

In choriocarcinoma, metastasis often develops early and is generally blood-borne because of the affinity of trophoblasts for blood vessels. Most have metastases in the lung (60-95%) and vagina (30-50%) [2]. Disease of the central nervous system is seen in 10% of patients, mostly in those with advanced disease. These patients almost always have concurrent pulmonary or vaginal involvement, or both. These lesions may undergo spontaneous hemorrhage leading to acute focal neurologic deficits [3]. Diagnosis of pulmonary embolization or metastasis is suspected in women with an abnormal chest radiograph after recent abortion or pregnancy in the presence of elevated serum hCG levels. These emboli or metastases manifest as multiple parenchymal nodules on radiography. Occasionally, a large intravascular tumor can also develop [4].

Pulmonary emboli or metastases are markedly vascular, as evidenced by extensive hemorrhage in tumor nodules and adjacent lung parenchyma. Symptoms are usually absent, although dyspnea may develop with extensive embolization. Hemoptysis may result from intrapulmonary hemorrhage [5]. Primary nongestational, extragonadal choriocarcinoma is very rare and always in or near the midline, because of primordial germ cell rests that failed to migrate properly [6].

As in our case, metastasis of unknown origin is the first impression. All of the reported cases have had similar features on imaging: low-density masses with heterogeneous enhancement and with an irregular margin. The clue to the nature of these tumors has been the brain MRI. Nonproportional perifocal edema of the brain mass, darker on gradient echo images, is suggestive of metastatic tumor with hemorrhage [7]. Rapid growth is another characteristic during the disease course. In a woman of childbearing age with multiple hypervascular masses with the characteristics of bleeding and rapid growth, choriocarcinoma should be considered.

Over 95% of malignant sequelae occur within approximately six months of evacuation of a hydatidiform mole [3]. The interval in our case was about five years, another unusual point. The other possibility is primary nongestational, extragonadal choriocarcinoma. But in our case, none of the masses were located in the midline. Because of the affinity of trophoblasts for blood vessels, blood-borne metastasis is not unusual. Although no pelvic mass was noted, the previous molar pregnancy could be the

original cause. The probable mechanism may be that some malignant cells escaped to the retroperitoneum and, via venous return, to the lungs, and then spread to brain parenchyma, causing the clinical symptoms.

Multiagent chemotherapy in conjunction with whole-brain irradiation results in acceptable survival rate in patients with metastatic GTN in the brain. Craniotomy is often necessary in fulminant cases [8].

We conclude that in a childbearing woman with multiple disseminated hypervascular masses in the brain, frequent bleeding, central necrosis and a tendency to rapid growth, choriocarcinoma should be considered. Elevated serum hCG levels and careful tracing of any history of a clinical mole, especially early on can lead to a correct diagnosis.

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