Neoplastic cerebral aneurysm from metastatic gestational choriocarcinoma

Case report


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This case of metastatic gestational choriocarcinoma presented as intracerebral hemorrhage from an atypical distal middle cerebral artery aneurysm. Operative evacuation of the intracerebral hematoma was undertaken and histopathological examination revealed choriocarcinoma invading the vessel wall. Neoplastic cerebral aneurysms are unusual, being reported in metastatic choriocarcinoma, cardiac myxoma, bronchogenic carcinoma, and undifferentiated carcinoma. Metastatic choriocarcinoma should be considered in the differential diagnosis of intracerebral hemorrhage in women of child-bearing age. Recent advances in treatment have resulted in a 75% cure rate for metastatic choriocarcinoma.

KEY WORDS · aneurysm · choriocarcinoma · hydatidiform mole · pregnancy

Peripheral located aneurysms in the brain are either idiopathic or secondary to infection, tumor embolus, moyamoya disease, or trauma. Neoplastic aneurysms have most frequently been reported in cases of cardiac myxoma. Eight reported cases were secondary to choriocarcinoma. More recently, there have been reports of cases involving other metastatic carcinomas. Neoplastic aneurysms are atypical, being fusiform and often associated with adjacent vascular abnormalities. They are commonly found in the peripheral branches of the middle cerebral artery. In this paper, we describe a case of neoplastic aneurysm from a metastatic gestational choriocarcinoma.

Case Report

This pregnant 16-year-old girl presented with vaginal bleeding; pelvic ultrasound showed a multicystic lesion consistent with hydatidiform mole. Serum β subunit human chorionic gonadotropin (HCG) levels were markedly elevated (> 1,024,000 IU/liter). The placenta was evacuated by suction curettage, and histopathological examination showed a typical hydatidiform mole.

After 8 days, recurrent bleeding necessitated repeat curettage, which showed persistent trophoblastic disease. Seventeen days later, the patient suddenly developed a severe left-sided headache and collapsed after 24 hours. On admission to the Royal Adelaide Hospital she was mute and restless, but obeyed simple commands.

Examination. She had tonic gaze to the left, and a right hemiparesis which predominantly affected her upper limb. There was mild neck stiffness. An emergency computerized tomographic (CT) head scan showed a large left frontal intracerebral hematoma which had ruptured into the ventricular system. A left carotid angiogram showed an aneurysm arising from an ascending frontal branch of the left middle cerebral artery (Fig. 1).

Operation. At an emergency left frontotemporal craniotomy, a vascular lesion was encountered in the anterior wall of the hematoma cavity. This consisted of an aneurysmal dilatation and adjacent tortuous blood vessels. The aneurysm began to bleed profusely, requiring induced hypotension to aid hemostasis. The lesion was excised and histopathological examination showed...
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The patient was treated with combined chemotherapy, consisting of three 8-day cycles of vincristine (1 mg/sq m), methotrexate (300 mg/sq m), cyclophosphamide (500 mg/sq m), and folic acid rescue. Initially, she was also given hydroxyurea (1 gm/day) for 2 days, and actinomycin D (0.5 mg/day) for 5 days. The final course included Adriamycin (30 mg/sq m). Concurrently, she underwent whole-brain irradiation with a total of 6200 rads fractionated over 46 days. Serum HCG concentration fell to undetectable levels, and her hemiparesis resolved. However, there was a persistent residual subtle dysphasia and memory disorder. At her last review, 12 months postoperatively, she was essentially asymptomatic with no evidence of choriocarcinoma by clinical, radiological, or serum HCG studies.

Discussion

Gestational choriocarcinoma is a rare, highly malignant, neoplasm of trophoblastic tissue occurring in about one of 40,000 pregnancies in the United States of America. Significantly higher incidences are reported in the Far East and parts of the Third World. Molar pregnancy is a common antecedent, but less than 10% of hydatidiform moles progress to choriocarcinoma. The neoplasm is disseminated via the maternal circulation and has an incidence of cerebral metastases of between 15% and 28%. The majority of patients with metastatic cerebral disease also have concurrent pulmonary involvement. Despite recent advances in the treatment of choriocarcinoma, cerebral metastases are responsible for significant mortality and residual morbidity. Two-thirds of the patients who die of advanced metastatic choriocarcinoma show autopsy evidence of cerebral involvement.

Patients with central nervous system involvement from choriocarcinoma may present with an acute stroke or encephalitic syndrome. There may be features of a space-occupying lesion, including spinal cord compression, but about 8% of cases are asymptomatic despite subsequently demonstrated cerebral lesions.
cranial hemorrhage is the most frequent mode of presentation of cerebral involvement and accounts for two-thirds of the cerebral clinical manifestations. The intracranial hemorrhage may be subdural, subarachnoid, or intracerebral.

Pathological examination of surgically resected or autopsy material in cases with intracranial hemorrhage has demonstrated tumor emboli occluding cerebral vessels, invasion of vessel walls, and (less commonly) aneurysm formation. It is proposed that invasion of the vessel wall occurs following tumor embolization, with consequent weakening of the vessel wall leading to its destruction and later hemorrhage. Focal damage to the vessel wall may also cause aneurysmal dilatation which, in time, may lead to rupture or thrombosis. This behavior is analogous to that of normal trophoblastic tissue with its propensity to invade the vessels of the uterine wall in order to establish pregnancy.

A literature review revealed eight cases of neoplastic cerebral aneurysm from gestational choriocarcinoma. Only two reports presented both radiological evidence of aneurysm formation and histopathological documentation of trophoblastic invasion and destruction of the vessel wall. Our case demonstrates focal destruction of the media and internal elastic lamina, with aneurysm formation. Neoplastic aneurysms are also reported in cardiac myxoma (18 cases), bronchogenic carcinoma (two cases), and undifferentiated carcinoma (three cases). The mechanism of aneurysm formation in cardiac myxoma is uncertain. It may involve invasion of the vessel wall by tumor, but could also represent noninvasive vascular damage secondary to embolic impaction.

It is likely that the incidence of neoplastic aneurysms is higher than suggested in the literature. Unless there is clinical suspicion, angiography is rarely performed in a patient with a metastatic cerebral tumor even if associated with intracranial hemorrhage. It is also probable that some neoplastic aneurysms are destroyed by the resulting hemorrhage, making radiological and histopathological diagnosis impossible. Other angiographic appearances in metastatic choriocarcinoma include arterial occlusion, angioma formation, and carotid-cavernous fistula.

We hypothesize that variations in the location, extent, and rapidity of vessel embolism may lead to a spectrum of vascular damage: namely, 1) aneurysm formation, with the lesion size depending on extent of vessel wall damage; 2) rapid complete destruction of the vessel wall leading to hemorrhage without recognizable aneurysm formation; 3) involvement of multiple adjacent vessels resulting in an angiomma-like lesion; and 4) cerebral infarction secondary to occlusion of the lumen of the vessel wall. A combination of these processes may operate in a given case. Although the major feature in our patient was aneurysm formation, the adjacent vessels were tortuous and dilated, suggesting an operative diagnosis of aneurysm associated with a vascular malformation.

The early diagnosis of metastatic choriocarcinoma and aggressive management of cerebral involvement is important because combined chemotherapy and cranial irradiation result in a 75% to 100% cure rate.

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