Olfactory Neuroblastoma as Acute Postpartum Depression

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Olfactory neuroblastoma is a rare neuroectodermal malignant tumor. It was first described by Berger et al in 1924,[1] and about 250 cases have since been reported, none of them in the perinatal period. The tumor emerges from the olfactory membrane located in the sinonasal area and anterior cranial fossa. It has a broad histologic spectrum and can be confused with peripheral neuroectodermal tumors.[2] Early symptoms are intranasal obstruction, epistaxis, and anosmia.[3] As the tumor grows, it can invade the oral cavity,[4] the orbits, and the brain.[3,5] In very rare cases the initial symptom is depression and frontal lobe dysfunction.[6] We describe a patient with olfactory neuroblastoma in whom the presenting symptom resembled postpartum depression.

Case Report

The patient, 40 years of age, gravida 3, para 4, first came to our clinic for prepregnancy consultation. All previous pregnancies and deliveries, the most recent 15 years ago, had been uneventful. She had a history of mild hypertension (140/90 mmHg), which from the 12th week of her last pregnancy was treated with methyldopa 250 mg twice a day.

Results of her primary workup for hypertension performed by her family physician, which included kidney, adrenal, and liver function tests, were normal. In addition, she suffered from chronic sinusitis for which she received homeopathic therapy and antibiotics and nasal spray for acute exacerbations. She was observed by an otolaryngologist before and during pregnancy. Findings on a computerized tomographic (CT) scan of the brain for sinusitis performed within the previous year were normal. She was also mildly obese.

Her intrauterine device was removed at her request after having been explained the risk of hypertension in pregnancy. She conceived 3 months later. We advised her to attend the high-risk pregnancy clinic. Aside from mild hypertension treated with a daily dose of 100 mg aspirin and 50 mg of atenolol, the pregnancy course was uneventful. Because of suspected macrosomia (estimated fetal weight, 4000 g), labor was induced at 38 weeks' gestation by intracervical prostaglandin E_2, and a 3920-g baby boy was delivered spontaneously with epidural anesthesia. There were no immediate complications.

Twelve hours later the patient complained of severe headache. Blood pressure and obstetric findings were normal, and the headache was attributed to the epidural anesthesia. Treatment consisted of bed rest, fluid loading, and dipyrone. Twenty-four hours later the patient complained of abdominal pain, became apathetic, lost interest in the newborn, and refused to get out of bed. Catheterization yielded 1800 mL of urine and led to some improvement in her condition. The next day (48 hours postpartum) she again complained of headache and became apathetic, refusing to get out of bed, or eat; she urinated in bed and did not seem to care.

We suspected postpartum depression. The psychiatrist, however, found no signs of acute psychiatric disease and referred her to a neurosurgeon because of the complaint of headache and her apathy. Asymmetry on preliminary neurologic examination raised suspicion of organic disease. An emergency brain CT scan showed a large space-occupying, fluid-filled left frontal lesion that was exerting extreme pressure on the left lateral ventricle. There was also a lesion showing partial enhancement in the anterior aspect of the brainstem bilaterally, with calcifications and thickening of the basal bone. Another space-occupying lesion was found in the nose invading both orbits medially and invading and filling both ethmoids to the brain stem.

An emergency left temporal craniotomy was performed using general anesthesia. The cystic lesion was drained and 70 cc of yellowish brown

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fluid was aspirated. The tumor at the base of the anterior fossa was removed; it was flaky and vascular and well differentiated from the anterior lobes. A left rhinotomy was performed to remove the tumor from the nose and left orbit. The postoperative period was uneventful with no motoric sequelae or cerebrospinal fluid leakage. A follow-up CT scan showed residual tumor on the right side and in the ethmoidal cells. The histopathologic examination revealed esthesioneuroblastoma.

Magnetic resonance imaging 3 weeks later showed partial recurrence of the frontal cyst and the residual tumor in the ethmoid cells and orbits. A team of neurosurgeons, otolaryngologists, and ophthalmologists performed a second operation. The tumor was completely removed. The postoperative recovery was uneventful, and 7 days after surgery the patient was discharged and referred for radiotherapy.

Three months after radiotherapy the patient's condition was being treated with prophylactic anticonvulsants. At that time she was feeling well and had no signs or symptoms of active disease.

Discussion

Although it is well known that intracranial tumors occurring in women of reproductive age can produce their first signs and symptoms during pregnancy, to the best of our knowledge, this is the first report of olfactory neuroblastoma occurring in the perinatal period. Pregnancy often masks the existence of the neoplasm, and the diagnosis can easily be missed.[7] Such symptoms as vomiting, headache, and visual disturbances are attributed by both patient and physician to the pregnancy itself or to preeclampsia. The influence of pregnancy on brain tumors is not completely known. Some authors believe that tumors grow during pregnancy because of the positive water balance as a result of the altered hormonal environment.[8] Others implicate the engorgement of the blood vessels that feed the tumor.[9] In our case, too, the brain neoplasm caused symptoms compatible with mild postanesthesia complications.

Olfactory neuroblastoma is a slow-growing tumor, accounting for only 2 to 3 percent of all malignant intracranial tumors.[10] It occurs in all age groups, but rarely during childhood, with a peak in the second and fifth decades of life (mean age at occurrence, 41 years).[11,12] There is no true sexual predominance, but some authors report a male-female ratio of 2:1.[13] Diagnosis is difficult and requires a high degree of suspicion because the signs and symptoms are nonspecific and the histopathologic findings are often unclear. Ultrastructural examination and identification of specific neural proteins[14-16] provide confirmation. Clinical symptoms resemble those of other intranasal neoplasms: nasal obstruction, anosmia, and epistaxis.[10,17] Later, as the tumor invades adjacent structures, the sinuses, orbits, palate, and brain,[3-5,18] there may be diplopia, ophthalmoplegia, exophthalmus, blurred vision, and blindness.[19] Our patient had unilateral exophthalmus and blurred vision caused by tumoral pressure on the orbits.

There can also be distant metastases involving the cervical lymph nodes, brain, lungs, bone, and liver,[17,20,21] and local recurrence appears in about 50 percent of cases.[17,22] The biologic behavior is unpredictable, ranging from a benign neoplasm to a highly malignant tumor. The tumor is not usually associated with hormone secretion, but in some cases it has been found to be an arginine vasopressin producer causing inappropriate antidiuretic hormone secretion syndrome.[4,23] In one case olfactory neuroblastoma was found to produce adrenocorticotropic hormone, causing Cushing syndrome.[24] Classification is based on the extent of tumor invasion,[20] and the staging system proposed by Kadish et al[21] provides a reference point for therapy and prognosis.

Olfactory neuroblastoma has been traditionally treated by radical surgery. Several authors have reported that the tumor is radiosensitive[21] and can also be treated successfully by chemotherapeutic agents.[25,26] The best treatment today seems to be oral surgery and neurosurgery combined with radiation and chemotherapy. Our patient underwent surgery twice for complete tumor removal and subsequent radiation therapy with good results at 3 months' follow-up. We cannot yet determine the precise prognosis.

The 5-year survival rate ranges from 20 percent to 71 percent[17,20,27] for olfactory neuroblastoma. Analysis of the various studies indicates no relation between the type of treatment and survival rate.[20,21] Apparently the biologic behavior of the tumor and the prognosis are determined not only by the extent of spread at the time of diagnosis and beginning of treatment but also by the histologic type and substance of secretions. Some of the latter still require further investigation.

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