Squamous cell carcinoma as a late complication of intracerebroventricular epidermoid cyst

Case report

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Primary intracerebral squamous cell carcinoma is a rare sequela of benign cerebral epidermoid cysts. A case of presumed malignant transformation in a lateral ventricular epidermoid cyst is described, in which squamous cell carcinoma arose 33 years after the surgical resection of a benign epidermoid cyst. Three years after diagnosis of malignancy, the patient remains functional with a slowly invasive tumor. This case provides a rare description of the natural history of forebrain squamous cell carcinoma, which would appear to be a more indolent tumor than previously realized.

KEY WORDS • brain neoplasm • epidermoid cyst • squamous cell carcinoma

INTRACEREBRAL squamous cell carcinoma is a rare complication of intracerebral epidermoid cysts. Most reported cases have involved cerebellopontine or parapontine epidermoid cysts and have been discovered only at autopsy.7 The case is presented of a 59-year-old woman who had had an intraventricular epidermoid cyst removed at the age of 23 years; 33 years later she developed squamous cell carcinoma arising in the original cyst bed. In the 3 years since, the tumor has followed an indolent, slowly invasive course. This is the fourth published case of forebrain epidermoid cyst associated with squamous cell carcinoma, and represents the longest survival yet recorded in such a patient.

Case Report

This 59-year-old Caucasian woman presented in 1949, at the age of 23 years, with headache and diplopia. Neurological examination at that time revealed papilledema and bilateral sixth nerve palsies. A pneumoencephalogram showed a right lateral ventricular mass, which was resected. Pathological examination disclosed a benign cerebral epidermoid cyst. Postoperatively, the patient was asymptomatic until 1960, when she developed generalized tonic-clonic seizures. She was initially managed with administration of phenytoin alone, but by 1970 the seizure frequency had increased, necessitating treatment with both phenobarbital and phenytoin. The seizures remitted with treatment, only to recur in 1982 with left partial sensory and generalized tonic-clonic seizures. A computerized tomography (CT) scan in 1982 revealed a contrast-enhancing right parietooccipital mass abutting the posterior wall of the third ventricle (Fig. 1 left).

A repeat CT scan 8 months later showed increasing tumor size, obstructive hydrocephalus, and a right lateral ventricular tumor. Angiography revealed a vascular mass of the right posterior thalamus and splenium. A right craniotomy with subtotal resection of the tumor was performed in November, 1982. Pathological examination of the excised specimen revealed an epidermoid cyst with foci of moderately to poorly differentiated squamous cell carcinoma (Fig. 2). Postoperative neurological deficits included a dense left hemiparesis, homonymous hemianopsia, hemihypalgesia, and left-sided hemineglect. The patient subsequently received 5000 rads of bitemporal radiation therapy. Postoperative CT scans revealed periatrial enhancement of the tumor remnant at the roof of the third ventricle.

The patient did well for the next 2½ years; her left-sided deficits improved and she could walk independently. In May, 1985, she became ataxic and noted increasing left-sided weakness, urinary incontinence,
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and impaired memory; these symptoms were followed by lethargy which progressed to stupor. A repeat CT scan showed lateral and third ventricular dilatation with a small distorted fourth ventricle. New contrast-enhancing tumor was demonstrated invading the right internal capsule and thalami bilaterally; the tumor traversed the splenium of the corpus callosum causing aqueductal compression posterior to the third ventricle (Fig. 1 right).

A ventriculoperitoneal shunt was placed for relief of the obstructive hydrocephalus. Rapid ventricular decompression was noted on CT, coincident with marked clinical improvement. The patient recovered her pre-morbid alert, oriented, and fluent mental status within 3 weeks after shunting. Her hemiparesis showed marked improvement, although her hemianopsia and hemisensory deficits persisted. The patient was discharged home, where she remains functional and ambulatory 3 years after diagnosis of the intracerebral squamous cell carcinoma.

Discussion

Primary intracerebral squamous cell carcinoma shares a rare but well-described association with benign intracranial epidermoid cysts. Cerebral epidermoid cysts are relatively common tumors, comprising roughly 0.5% of all intracranial tumors. However, their complication by squamous cell carcinoma is an unusual occurrence. Garcia, et al., defined true intradural squamous cell carcinoma; according to their classification, 19 cases have hitherto been reported. Of these, 16 were associated with epidermoid cysts, suggesting malignant transformation. In all but three of these cases, the tumors originated in the cerebellopontine or parasellar regions. In the remaining three cases, epidermoid cysts containing squamous cell carcinoma were found in the frontal lobes. Apparently these represent the only cases of primary forebrain squamous cell carcinoma yet reported.

Previous to our case, the longest reported duration of symptoms in a cerebral epidermoid cyst with squamous cell degeneration was 13 years. However, only twice has a cerebral epidermoid cyst been diagnosed before the development of known squamous cell carcinoma. More typically, the two have been diagnosed simultaneously at surgery or autopsy.

In summary, the case of a 59-year-old woman with squamous cell carcinoma invading the posterior diencephalon and splenium of the corpus callosum is described. This tumor was discovered 33 years after subtotal resection of a benign intraventricular epidermoid...
cyst. Three years after partial resection of the malignant tumor, with subsequent whole-brain irradiation and ventriculoperitoneal shunting, the patient remains functional with a slowly invasive forebrain carcinoma.

References

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