Primary intracranial epidermoid carcinoma

Case report

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A case is presented of primary intracranial epidermoid carcinoma in the right cerebellopontine angle which was visualized as a homogeneously enhanced mass on computerized tomography. At autopsy the malignant tissue was found to have invaded the brain stem.

KEY WORDS • epidermoid carcinoma • epidermoid cyst • cholesteatoma • cerebellopontine angle tumor • computerized tomography

Primary intracranial epidermoid carcinoma is an extremely rare neoplasm. Only 13 cases have been reported in the literature. There has been only one recent report on computerized tomography (CT) findings in malignant epidermoid. The following is a case of a malignant intracranial epidermoid detected by CT.

Case Report

This 46-year-old man was admitted to our University Hospital complaining of headache and vomiting. He had a 10-year history of paroxysmal tingling pains in the right forehead lasting a few hours on six or seven occasions, for which he had not undergone evaluation. Six months before admission, he began complaining of numbness over the right side of the face, which persisted. Three months later he developed diplopia on lateral gaze to the right and drooling of the right side of his mouth. Two weeks before entry, he developed increasingly severe headache with nausea and vomiting. At about that time he noticed motor weakness on the left side.

Examination. He was a well developed, well nourished man. Ophthalmoscopic examination revealed bilateral papilledema. He had a complete right sixth-nerve palsy and vertical nystagmus on looking upward. Hypoesthesia and hypesthesia were noted in the distribution of all divisions of the right fifth nerve, with absence of corneal reflex, atrophy of the right masseter and temporal muscles, and deviation of the jaw to the right on opening the mouth. There was a flattening of the right nasolabial fold. Hearing was normal. A left hemiparesis was noted. On testing for rapid alternating movement, motion of the right arm was slow and irregular. The deep-tendon reflexes were active and equal, with a positive Babinski sign on the left. Routine hematological and chemical studies were normal.

Plain skull films were normal. Laminograms of the temporal bones were normal. A CT scan demonstrated a lesion with faintly increased absorption in the right cerebellopontine angle (Fig. 1 left). Postcontrast CT revealed two discrete areas of homogeneous enhancement (Fig. 1 center). The mass extended rostrally into the pontine cistern and slightly to the right of the midline. The fourth ventricle was displaced to the left and there was a moderate degree of hydrocephalus. A retrograde brachial angiogram showed elevation and posterior displacement of the right superior cerebellar artery. The petrous vein was not visualized despite filling of the right posterior inferior cerebellar artery.

Operation. A right suboccipital craniectomy was performed. The dura appeared normal. On elevation of the cerebellar hemisphere, a tumor was exposed in the right cerebellopontine angle. It was yellowish, firm, and well encapsulated. The tumor capsule was opened and the dry, cheesy, yellow intracapsular contents were removed. The tumor was located anterior
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FIG. 1. Computerized tomography scans. **Left:** Precontrast scan showing a lesion of faintly increased density in the right cerebellopontine angle with displacement of the fourth ventricle to the left. **Center:** Postcontrast scan demonstrating two discrete areas of homogeneous enhancement. **Right:** Scan 5 months after operation reveals a homogeneously enhanced lesion in the brain stem.

to the right seventh- and eighth-nerve complex, displacing the superior cerebellar artery superiorly and enveloping the fifth nerve which was resected. The medial part of the tumor was firmly adherent to the inferolateral aspect of the pons so that it was left intact. Histological examination of the removed tissue showed squamous cell carcinoma with laminated keratin formation.

**Postoperative Course.** The patient’s postoperative course was complicated by aseptic meningitis and communicating hydrocephalus which required a ventriculoperitoneal shunt. He was given a course of physical therapy, but slowly developed progressive signs of brain-stem involvement. A CT scan 5 months after operation showed a contrast-enhanced lesion in the brain stem (Fig. 1 right). He died 7 months after operation.

**Postmortem Examination.** Significant findings were confined to the intracranial cavity. A multilobulated, encapsulated, grayish-white tumor occupied the right cerebellopontine angle, encasing the right seventh and eighth nerves, and compressing the right inferolateral pons. The pons was irregularly swollen due to direct infiltration by the tumor. On transverse sections through the pons and cerebellum, a large part of the pons was replaced by a sharply demarcated tumor tissue which extended into the fourth ventricle (Fig. 2). The aqueduct of Sylvius was

FIG. 2. Photograph of the sharply demarcated tumor invading the pons, and extending into the fourth ventricle.

FIG. 3. Photomicrograph showing squamous-cell carcinoma with acellular laminated keratin. H & E, × 75.
completely obstructed. There was no evidence of tumor elsewhere in the brain and meninges. Microscopically, the tumor consisted of a large amount of desquamated keratinized material which was lined by squamous-cell carcinoma (Fig. 3). The degree of differentiation of the tumor varied considerably; in some areas it was poorly differentiated, and in other areas squamous cells could be recognized (Fig. 4). The central part of the tumor was necrotic with no massive hemorrhage. In no area was any obviously benign squamous epithelium seen.

Discussion

Epidermoid cysts are congenital tumors, and account for 0.2% to 1.0% of all central nervous system neoplasms. Intradural epidermoid cysts arise in one of several preferred sites. The cerebellopontine angle is the most common location of origin in many reported series. The cysts contain structureless, waxy, keratinous material, with walls formed by a thin layer of squamous epithelium. Malignant change in the lining of the squamous epithelium is rare but well documented; 13 cases have previously been reported in the literature. The most common site for the carcinoma is the parapontine or cerebellopontine angle, the location of the tumor in eight of the 14 cases, including ours. The age of the patients varied from 4 to 85 years, and the peak incidence was in the fifth and sixth decades. There is a male predominance of 10 to 4. Wong, et al., reviewed the world literature and stated that the incidence, symptoms, and duration of symptoms of parapontine epidermoid carcinomas arising from an epidermoid cyst closely mimic those of benign epidermoid cysts of the cerebellopontine angle or parapontine.

The CT appearance of epidermoid cysts has been reported to be of low absorption value with or without a calcified capsule. The cysts do not enhance following infusion of contrast material. Since the advent of

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FIG. 4. Photomicrographs of the tumor tissue. H & E, × 160. Left: Section showing squamous-cell carcinoma. Right: The tumor cells are poorly differentiated with hyperchromatic, pleomorphic nuclei.
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CT, there has been only one report of a malignant epidermoid cyst, the case of a 59-year-old man in whom a CT scan demonstrated a homogeneously enhanced lesion in the right cerebellopontine angle after injection of contrast material. In our case, plain CT scan demonstrated an area of faintly increased density that was then enhanced homogeneously with contrast material, and showed a lobulated contour. From these two cases, it appears that a malignant epidermoid cyst is isodense or of faintly increased absorption value, and is enhanced homogeneously throughout the lesion. This type of CT appearance is seen in neurinomas and meningiomas. In our experience, it is impossible to distinguish the carcinoma from these two tumors. In the limited data available, malignant epidermoid cysts showed a variable degree of vascular displacement on angiogram because of their avascular nature.5,4,6,10

Epidermoid cysts are slow-growing benign tumors, but may have grown too large to be resected completely, resulting in recurrence after long intervals. Fox and South4 reported the case of a 43-year-old man in whom a benign intraventricular epidermoid cyst underwent malignant changes over a period of 7 years. Their patient died 6 months after the diagnosis was made. The cases described by Toglia, et al.9 and Haig5 were found to have malignant epidermoid tumors 13 and 15 months, respectively, after the initial operation, at which time a benign epidermoid cyst was diagnosed. The majority of the previously reported cases were fatal in less than 12 months after the onset of symptoms or establishment of the diagnosis.4,6,9,10 Occurrence of malignant change should be suspected in any patient with a known epidermoid cyst who does not show the expected recovery after surgical removal of the cyst's contents, or whose condition rapidly deteriorates. Malignant transformation may be considered when postoperative CT scans show contrast enhancement in the previous operation site.

Carcinoma arising from these cysts has been described as keratinizing and highly infiltrative. Metastatic squamous-cell carcinomas should be considered in the differential diagnosis. The gross encapsulation of this tumor and the absence of any primary tumor elsewhere at autopsy indicate a primary lesion. The present case was found to have invaded the brain stem in spite of the absence of widespread leptomeningeal infiltration.

References

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