Squamous cell carcinoma arising in a suprasellar epidermoid cyst

Case report

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A case is presented in which a squamous cell carcinoma developed in an intracranial epidermoid cyst. The patient was a 54-year-old woman with a 3-year history of depression and amblyopia; no focal findings were noted and she was diagnosed as having psychiatric disorders. On her final admission she showed clinical evidence of a rapidly growing intracranial mass. Computerized tomography (CT) identified a right parasellar and temporal lesion which was then incompletely removed. The literature on primary intracranial squamous cell carcinoma is reviewed, and the role of CT scanning in preoperative diagnosis of this lesion is discussed.

KEY WORDS: brain tumor □ parasellar tumor □ epidermoid cyst □ computerized tomography □ squamous cell carcinoma

INTRACRANIAL epidermoid cysts are histologically benign lesions which may nonetheless produce complications because of their insinuating expansile patterns of growth. There are several reports of apparently primary intradural squamous cell carcinoma, most often in association with a preexisting epidermoid cyst.4-7,10,12-17,20-25 We are reporting another such case, which presented acutely as an expanding suprasellar mass. At craniotomy a malignant lesion was found in the medial right temporal lobe, which proved to be squamous cell carcinoma. A subjacent epidermoid cyst extended inferomedially anterior to the brain stem and into the right cerebellopontine angle.

Case Report

This 53-year-old woman was admitted with increasing confusion, drowsiness, right hemiparesis, and headache and vomiting. Three years earlier she had been placed in a nursing home because of inability to care for herself while living alone; she had profound "depression" and "hysterical amblyopia."

Examination. On admission, her pupils were equal and reactive to light, and she had bilateral papilledema and sixth nerve palsies, slight weakness of the right side of her face and right arm, and right hyperreflexia with a Babinski response. She was fluent dysphasic and comprehension was impaired. Computerized tomography (CT) was performed (Fig. 1). The unenhanced scan showed a low-density lesion in the suprasellar and right parasellar regions (mean 12 Hounsfield units) extending to the right cerebellopontine cistern and across the midline to the left Sylvian fissure. These lesions did not enhance with contrast material, but were in direct contact with a slightly dense lesion in the right temporal lobe which enhanced dramatically to 84 Hounsfield units. A low-density area around the enhancing component suggested edema. There was a mild midline shift to the left, and moderate hydrocephalus.

Operation. Piecemeal removal of the anterior 6 cm of the right temporal lobe revealed quite vascular tumor material in its medial part. Below and continuous with this tumor was a pearly white mass, fingers of which extended anteriorly to the brain stem and into the right cerebellopontine angle. Vascularity precluded total removal of the intratemporal lesion, which extended further posteriorly. The patient did well initially, but deteriorated quickly with increasing hydrocephalus and...
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Fig. 1. Left: Unenhanced computerized tomography scan. The suprasellar low-density area has a mean value of 12 Hounsfield units. This low-density area extends into the left Sylvian fissure. Right: Enhanced scan (slightly lower cut). The enhancing component has a mean value of 84 Hounsfield units and abuts on the low-density suprasellar component. Note the extension of this low-density area into the right cerebellopontine cistern and left Sylvian fissure.

tumor recurrence. She died several weeks after the operation, and autopsy was not permitted.

Pathological Examination. The malignant tumor consisted of sheets and clumps of cells with multifocal necrosis and moderate stroma. It infiltrated brain tissue as blunt, well defined tumor masses, provoking moderate astrocytic reaction (Fig. 2). Tumor nuclei were large and rounded with one to three irregular nucleoli, fine chromatin, and a delicate membrane. There was moderate mitotic activity. Cytoplasm was usually scanty and poorly defined, slightly granular, and basophilic. Occasional better differentiated foci included large polygonal cells with pale eosinophilic cytoplasm, often containing dense keratin-like flakes. Prickle cells were not seen. Cells contained some dispersed glycogen, but no mucus. The stroma contained patchy glial fibrillary acidic protein, presumably reflecting trapped astrocytes. Electron microscopy showed scanty tonofilbrils and desmosomes and was characteristic of squamous cell carcinoma. Sections from the pearly white mass showed benign squamous epithelium with masses of keratin (Fig. 3); the transition to malignancy was not seen in available material.

Discussion

Survey of the literature shows 18 previously reported cases of primary intradural squamous cell carcinomas, which are summarized along with the present case in

Fig. 2. Photomicrographs of the malignant tumor. Left: Infiltration of the brain tissue by malignant tumor is shown. H & E, × 75. Right: Moderate differentiation can be seen in the squamous cell carcinoma. H & E, × 460.
TABLE 1

Summary of 19 cases of primary intradural squamous cell carcinoma*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors, Year</th>
<th>Age (yrs), Sex</th>
<th>Symptom Duration</th>
<th>Tumor Origin</th>
<th>Location of Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Ernst, 1912</td>
<td>52, M uncertain</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>EC</td>
<td>cerebellopontine angle</td>
</tr>
<tr>
<td>2</td>
<td>Hug, 1942</td>
<td>49, M</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>EC</td>
<td>parapontine</td>
</tr>
<tr>
<td>3</td>
<td>Henkel, 1951</td>
<td>49, M</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>EC</td>
<td>parapontine</td>
</tr>
<tr>
<td>4</td>
<td>76, M</td>
<td>uncertain</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>EC</td>
<td>parapontine</td>
</tr>
<tr>
<td>5</td>
<td>Yamanaka, et al., 1955</td>
<td>57, M</td>
<td>uncertain</td>
<td>EC basal</td>
<td>cerebellopontine angle</td>
</tr>
<tr>
<td>6</td>
<td>Kahn, 1955</td>
<td>37, F</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>EC rt frontal</td>
<td>rt frontal</td>
</tr>
<tr>
<td>7</td>
<td>Davidson &amp; Small, 1960</td>
<td>46, M</td>
<td>&lt; 12 mos</td>
<td>EC rt frontal</td>
<td>rt frontal</td>
</tr>
<tr>
<td>8</td>
<td>Landers &amp; Danielski, 1960</td>
<td>73, F</td>
<td>&lt; 12 mos</td>
<td>EC cerebellum, 4th ventricle</td>
<td>?</td>
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<tr>
<td>9</td>
<td>Gluszczy, 1962</td>
<td>52, M</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>DC</td>
<td>rt frontal</td>
</tr>
<tr>
<td>10</td>
<td>Komjatszegi, 1964</td>
<td>45, F</td>
<td>&lt; 12 mos</td>
<td>EC basal</td>
<td>rt frontal</td>
</tr>
<tr>
<td>11</td>
<td>Fox &amp; South, 1965</td>
<td>43, M</td>
<td>&gt; 12 mos, known EC</td>
<td>EC</td>
<td>rt frontal</td>
</tr>
<tr>
<td>12</td>
<td>Toglia, et al., 1965</td>
<td>53, F</td>
<td>&gt; 12 mos, known EC</td>
<td>EC</td>
<td>rt frontal</td>
</tr>
<tr>
<td>13</td>
<td>Salver &amp; Carter, 1973</td>
<td>58, F</td>
<td>&lt; 12 mos</td>
<td>EC</td>
<td>rt frontal</td>
</tr>
<tr>
<td>14</td>
<td>Wong, et al., 1976</td>
<td>4, M</td>
<td>&lt; 12 mos</td>
<td>ISC</td>
<td>sellar</td>
</tr>
<tr>
<td>15</td>
<td>Scully, et al., 1977</td>
<td>59, M</td>
<td>&lt; 12 mos, no known focal lesion</td>
<td>?</td>
<td>parapontine</td>
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<tr>
<td>16</td>
<td>Kompf &amp; Menges, 1977</td>
<td>57, F</td>
<td>&gt; 12 mos, no known focal lesion</td>
<td>?</td>
<td>cerebellopontine angle</td>
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<tr>
<td>17</td>
<td>Nosaka, et al., 1979</td>
<td>46, M</td>
<td>&lt; 12 mos</td>
<td>?</td>
<td>cerebellopontine angle</td>
</tr>
<tr>
<td>18</td>
<td>Dubois, et al., 1981</td>
<td>53, M</td>
<td>&lt; 12 mos</td>
<td>EC</td>
<td>4th ventricle</td>
</tr>
<tr>
<td>19</td>
<td>Lewis, et al., 1983</td>
<td>53, F</td>
<td>&lt; 12 mos</td>
<td>EC</td>
<td>parasellar</td>
</tr>
</tbody>
</table>

* EC = epidermoid cyst; DC = dermoid cyst; ISC = intrasellar cyst; ? = preceding benign lesion not verified.

Table 2. A comparison with the reported features of benign epidermoid cysts (Table 2) suggests that the age and sex incidence, and the most sites of the malignant lesions, do not differ significantly from those of the benign cases. Of 16 cases in which the mode of presentation is known, nine patients had clinical neurological dysfunction for less than 1 year, with no hint of any preceding lesion. In seven cases there was a longer history (often of many years), frequently referable to cranial nerves or, as in the present case, considered to be functional. Only in two of these 16 cases 9,23 was a diagnosis of benign epidermoid cyst made prior to discovery of the malignant lesion. Nonetheless, 15 of the 19 cases are known to have originated either in an epidermoid cyst (in 13), a dermoid cyst (in one), or an intrasellar cyst (in one). In four cases no preceding lesion was identified.

The usual CT appearance of a benign intradural epidermoid mass is of a sharply defined low-density lesion that does not enhance following intravenous injection of contrast material. Attenuation values are often less than 0 Hounsfield units, 5,18 although high-density lesions measuring 80 to 120 Hounsfield units have been reported. 1,2,11

Intracranial epidermoid cysts may be intradural or extradural, and the former may be intra-axial or extra-axial. Extra-axial lesions tend to follow the subarachnoid pathways into the sulci and cisterns of the brain, as in the present case. The CT attenuation coefficients reflect the contents of the cyst which are mainly desquamated epithelial cells and cholesterol crystals. Fatty degeneration may account for the low densities below 0 Hounsfield units. Occasionally, escape of the fatty contents may produce fat-fluid levels in the cisterns and/or ventricles. 3 The cause of the high density of cyst contents is not certain, although it has been attributed to calcification in the desquamated and keratinized debris, saponification into calcium soaps, iron in the form of ferrocyanide complexes, hemoglobin, hemosiderin, or high protein content (Ito, et al., quoted by Handa, et al. 11). Peripheral calcification is uncommon and more often associated with dermoid cysts, although Handa, et al., 11 reported calcification in five of 13 patients with intradural epidermoid tumors, in one case quite dense.

Rim enhancement has been reported in only five cases of benign epidermoid cysts, 2,11,18,19 and generally there is no such enhancement. 2,5,8,18 The mechanism of enhancement in benign epidermoid cysts is not clear, since they generally have thin poorly vascularized capsules. Handa, et al., 11 suggest that peritumoral granulation tissue secondary to leakage of irritating cyst contents and chemical inflammation, together with
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...compressed vessels around the tumor, may account for the enhancement and simulate an enhancing capsule.

In case reports of either primary malignant epidermoid tumor or malignant change in a benign epidermoid cyst, CT scanning has demonstrated an area of contrast enhancement in the malignant part of the lesion.6,20,22 In the case of Dubois, et al.,6 both the benign cyst (low-density nonenhancing lesion) and the malignant component (enhancing lesion) were seen. In our case also, the two components (low-density benign suprasellar and malignant enhancing lesion) were evident. Before enhancement, the malignant part of the lesion is isodense or of slightly increased density relative to the normal brain. Low density adjacent to the enhancing component suggests some peritumoral edema. The finding of prominent enhancement adjacent to a lesion otherwise suggestive of an epidermoid cyst, although not specific, should raise the possibility of malignant change in the epidermoid cyst.

References


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