Intrasellar gangliocytomas

Report of four cases

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Four cases of intrasellar gangliocytoma are reported because of their close clinical resemblance to tumors of the pituitary gland. Similar cases from the literature are discussed.

Key Words: intrasellar gangliocytoma · pituitary tumor · sella turcica

Among the tumors of the sella turcica, pituitary adenomas and craniopharyngiomas are the best known. Other abnormal space-occupying processes in this region, whether blastomas or not, are extremely rare and are usually not diagnosed preoperatively. We are presenting four cases of tumors that appeared to be pituitary adenomas but were identified histopathologically as gangliocytomas.

Analysis of Cases

General Findings

Clinical symptoms, endocrinological, radiological, intraoperative, and histological findings are summarized in Table 1. Case 1 exhibited total insufficiency of the anterior lobe of the pituitary. In Cases 2 and 4, acromegaly was prominent while Case 3 showed a fully developed Cushing's syndrome. In each case an enlarged pear-shaped sella with demineralization of the dorsum sellae combined with the endocrinological findings to suggest a tumor of the pituitary gland.

At operation the whitish aspect of the tumor, its atypical meaty consistency, and the absence of a capsule were noted. In all four cases the diaphragm and basal dura of the sella were intact.

In each case histological study revealed large nerve cells that were sometimes pyramidal with processes arising from pericaryon that could be traced into the surrounding tissue. These cells varied in size, but most were large with a small quantity of cytoplasm and large vesicular nuclei. Nissl granules were noted at the cell margins. These nerve cells were frequently multinucleated. In addition, there were smaller cells that also had abundant cytoplasm and vesicular nuclei. It was not possible to determine positively whether these cells were of pituitary or ganglionic origin. In three of the four cases the material removed at operation showed
### TABLE 1

*Summary of four patients with intrasellar gangliocytomas*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Symptoms at Admission</th>
<th>Duration of Symptoms</th>
<th>Endocrine* Studies</th>
<th>Radiology &amp; Brain Scan</th>
<th>Operative Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>41 F</td>
<td>headache, amenorrhea, bilateral upper quadrant, anopsia</td>
<td>2 yrs</td>
<td>no gonadotropin</td>
<td>tumor of the pituitary with suprasellar expansion</td>
<td>rhinosseptal approach; meaty tumor with fibrous tracks growing by expansion; diaphragm &amp; basal dura of sella intact; total removal</td>
</tr>
<tr>
<td>2</td>
<td>34 F</td>
<td>headache, amenorrhea, diabetes mellitus, galactorrhea, increasing acromegaly</td>
<td>2 1/2 yrs, 8 mos after x-ray therapy</td>
<td>STH 100/94 ng/ml plasma</td>
<td>pear-shaped sella, dorsum sellae intact</td>
<td>transsphenoidal approach; soft whitish tumor displacing adjacent tissues; diaphragm &amp; basal dura of sella intact; total removal</td>
</tr>
<tr>
<td>3</td>
<td>37 F</td>
<td>weakness, fatigue, obesity, osteoporosis, Cushing's syndrome</td>
<td>2 1/2 yrs</td>
<td>hypocalcemia, alkalosis, PBI 8.2% plasma-cortisol 41.58%, 17 hydroxysteroids 35.5 mg/24 hr, 93.0 mg/24 hr after Metopirone, 17 keto- steroids 8.9 mg/24 hr, 14.2 mg/24 hr after Metopirone</td>
<td>sella turcica normal, os sphenoidale destroyed, no suprasellar growth</td>
<td>transsphenoidal approach; tumor lying only in cavum sinus sphenoidalis; diaphragm &amp; basal dura intact; total removal</td>
</tr>
<tr>
<td>4</td>
<td>56 M</td>
<td>headache, acromegaly, fatigue, bitemporal heteronymous hemianopsia, diabetes mellitus</td>
<td>15 yrs, 1 yr after x-ray therapy</td>
<td>cavum sellae enlarged, cranial dislocation of diaphragm</td>
<td></td>
<td>transfrontal approach; soft whitish-blue expanding tumor; diaphragm &amp; basal dura intact; total removal</td>
</tr>
</tbody>
</table>

* STH = somatotropin; PBI = protein bound iodine. Metopirone is made by CIBA Pharmaceutical Company, Summit, New Jersey 07901.
pituitary tissue while in one case there was a small pituitary adenoma in addition to the larger gangliocytoma.

Histological Findings

Case 1. Fibrillary tissue showed nest-like accumulations of large round, oval-shaped, or pyramidal cells with one or more axons. These cells had large vesicular nuclei with pointed nucleoli and were sometimes bi- or multinucleated. In addition, there were cells without clearly visible cytoplasm, also with round or oval-shaped nuclei. Some pituitary tissue showed infiltration by tumor cells. The diagnosis was gangliocytoma.

Case 2. Besides small, mostly agranular cells, there were small cells with granules selectively stained by Orange G and islands of voluminous partly pyramidal mono- or multipolar cells with typical vesicular nuclei and well-defined nucleoli. Nissl granules could be seen at the cell margin in the cytoplasm. Two tumors, pituitary adenoma and gangliocytoma, were diagnosed.

Case 3. Rhythmic structures with microcysts, sporadic pseudopapillae, and bi- or multipolar ganglionic cells were seen; these cells were uni- or multinucleated with large vesicular nuclei and Nissl granules in the cytoplasm (Fig. 1). In addition, there were small cells with vesicular nuclei without certain proof of pituitary or ganglionic origin. A gangliocytoma was diagnosed.

Case 4. Pituitary tissue was removed from anterior and middle lobe. In the posterior lobe there were nest-like accumulations of uni- or multipolar cells with vesicular nuclei and large nucleoli. Nissl substance was located at the margin of the cytoplasm. These nerve cells were often multinucleated. In addition, tightly packed small cells also with large nuclei and nucleoli could be seen. These cells sometimes showed atypical mitosis (Fig. 2). The tumor was diagnosed as a gangliocytoma.

Discussion

Gangliocytomas have been reported in the tuber cinereum and third ventricle, temporal lobe, frontal and parietal lobes, cerebellum, and medulla oblongata in that order of incidence. Our four cases of gangliocytoma of the sella turcica, together with the seven we found reported else-
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Fig. 2. Case 4. Photomicrographs showing large, sometimes pyramid-shaped cells that are often multinucleated and undergoing atypical mitosis. H & E, × 500.

where,1,3,8,12,15,16 represent a relatively rare location. There was evidence in our Cases 1, 2, and 4 that the tumor originated from the region of the pituitary gland, while in Case 3 it may have originated from the fossa nasalis, as described by Tonelli,19 Duchini,6 and Grimaud, et al.9

The ages of our patients ranged from 34 to 56 years, in agreement with previous reports of gangliocytomas in the same region. Patients with gangliocytomas in other locations were younger, usually in the second decade,21,22 they also were usually male,21 while in our series three of four patients were female.

As in other reports,1,8,15 all our patients showed symptoms of endocrine over- or underproduction. We agree with the suggestion of Angelstein1 that the endocrinological disturbances may be produced either by destruction of the pituitary gland or by central disturbances without change in the pituitary. In fact, it seems that intrasellar gangliocytomas cause endocrinological symptoms not by themselves but by secondary involvement of the surrounding tissue.

In each of our cases the diaphragm and the basal dura of the sella turcica were intact. Thus, the typical alterations described by Driggs and Spatz,4 Schmidt, et al.,18 and Lange-Cosack13,14 for hamartomas of the pituitary stalk or hypothalamus cannot be held responsible for the endocrinological changes in our cases. These infundibular gangliocytomas contain endocrinologically active cells.

The characteristic maturity of the cells in all our cases is consistent with the long duration of symptoms before hospitalization and the relatively good prognosis after operation. Since the research of Romeis,17 we know that there are ganglionic cells in the fetal posterior lobe of the pituitary gland which are reduced during further maturation. There still may be residual nerve cells in the adult human neurohypophysis10,11,17 from which gangliocytomas of the pituitary gland may originate, as Bielschowsky and Simons2 have postulated for other locations. Similarly, the infundibular stalk tumors probably arise from the nucleus of Cajal.4

References


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