Pituitary adenomas of adolescents

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A review of four cases of chromophobe adenomas in adolescents suggests that extracapsular extension with invasion of parasellar structures is more common in this age group than in adults. A syndrome of adolescent obesity, oculomotor palsies, and plain x-ray changes of the sella characterizes this group.

KEYWORDS • adolescents • chromophobe • adenoma • extracapsular extension

Pituitary adenomas in adolescents have been previously described but only in studies of the general population. The symptoms described are similar to those of adults, and no special features of these tumors in young people have been distinguished. A review of our cases and of the literature leads us to believe that there is a high incidence of extracapsular extension of this tumor in adolescents. We have seen 115 patients with pituitary adenomas during the past 15 years; of these, four were adolescents and all of them had invasive tumors. Two cases are presented in detail, and Table 1 summarizes the similarities of the four cases.

Case Reports

Case 1

This 15-year-old boy presented in 1961 with a 1-year history of decreased vision in the left eye and difficulty in using his right arm. Physical examination revealed mild obesity, left ptosis, right central seventh nerve palsy, and right hyperreflexia. Visual acuity on the left was reduced to finger counting and optic atrophy was evident; visual field was normal in the right eye, but the left eye revealed central scotoma and some depression in the superior temporal field. X-ray examinations revealed sellar enlargement, opacification of the sphenoid sinus with loss of the sellar floor, and expansion of the left superior orbital fissure. Carotid angiography demonstrated elevation of the carotid siphon and anterior choroidal artery consistent with a para- and suprasellar mass. Partial endocrine workup revealed a normal triiodothyronine (T₃), 17 hydroxy, and ketosteroids and metapyrone tests.

A left frontal craniotomy revealed a large tumor medial and lateral to the left optic nerve and extending into the middle cranial fossa. Pathological examination revealed some pleomorphism and cranial nerve invasion but the histological picture was consistent with a chromophobe adenoma. The immediate postoperative period was characterized by left third nerve palsy, mild right hemiparesis, and polyuria. The patient received postoperative radiation, a total of 5000 rads. At follow-up 1 year later, he had normal visual fields and acuity with no hemiparesis and normal adrenal pituitary axis. At present, the patient is fully employed as a surveyor.
TABLE 1
Clinical findings in four adolescents with pituitary adenoma*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age &amp; Sex</th>
<th>Obesity</th>
<th>Fifth Nerve Involvement</th>
<th>Brain Scan</th>
<th>Angiogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>15 M</td>
<td>mild</td>
<td>no</td>
<td>not done</td>
<td>supra- and parasellar mass</td>
</tr>
<tr>
<td>2</td>
<td>17 M</td>
<td>yes</td>
<td>yes</td>
<td>+</td>
<td>parapituitary mass</td>
</tr>
<tr>
<td>3</td>
<td>15 F</td>
<td>yes</td>
<td>yes</td>
<td>+</td>
<td>parapituitary mass</td>
</tr>
<tr>
<td>4</td>
<td>15 F</td>
<td>yes</td>
<td>yes</td>
<td>normal</td>
<td>parapituitary mass</td>
</tr>
</tbody>
</table>

* All four patients had oculomotor palsies and sellar changes on x-ray studies.

Case 4

This 15-year-old girl presented initially at the University of Minnesota Hospitals in 1971 for investigation of obesity. She weighed 250 lb; excessive weight gain had begun at the age of 8. Menarche occurred at age 13, with normal menstrual periods since. Physical examination revealed obesity, abdominal stria, and buffalo hump; the neurological examination was normal. Laboratory examinations were normal for diurnal cortisol variation, 17 ketosteroids, 17 ketogenic steroids, and 17 hydroxy corticosteroids. At that time, she was given a reducing diet and was discharged.

The patient returned in February, 1973, complaining of episodic headaches, double vision, and right facial numbness. Examination revealed obesity, right total ophthalmoplegia, and decreased sensation in the first and second divisions of the trigeminal nerve. Skull x-ray films, electroencephalogram, brain scan, lumbar puncture, and a right carotid angiogram were all normal. The patient was discharged with a tentative diagnosis of cavernous sinus thrombosis.

She returned again in January, 1974, complaining of irregular menstrual cycles and increased facial hair. Physical examination showed increased sensory deficit on the right side of the face and masseter paresis. Visual acuity was 20/80 on the left and 20/20 on the right. Visual fields were normal. Skull x-ray films at this time revealed mottling and sclerosis of the sella turcica and lesser wing of the sphenoid. Laboratory examinations showed an abnormal glucose tolerance test, no growth hormone response to levodopa and glucagon, elevated serum cortisol, 17 ketosteroids, and 17 hydroxy steroids, with partial suppression after a 5-day course of dexamethasone. Follicle stimulating hormone (FSH), total tetraiodothyronine (T4), free T4, thyroid stimulating hormone (TSH), and testosterone were all normal. Brain scan was also normal. Carotid angiogram demonstrated medial displacement of the internal carotid artery consistent with a parasellar mass.

A right transfrontal craniotomy revealed a completely extrasellar lesion extending into the superior orbital fissure and down the clivus. Histological examination was compatible with chromophobe adenoma. Postoperatively, the patient received 5000 rads. Her neurological status is unchanged; she has returned to her former level of activity and is enrolled in a secretarial school. She is receiving no endocrine replacement therapy.

Discussion

A review of previously reported series of pituitary adenomas in childhood failed to reveal any mention of the high incidence of extrasellar extension in this age group. However, in these previous series, a disproportionately large number of invasive adenomas occurred in adolescents. For example, two of five patients reported by Martins, et al., and three of 14 patients reported by Weinberger, et al., who had pituitary adenomas with parasellar extension were adolescents. In 15 years, we encountered four adolescents with invasive chromophobe adenomas, and had no patients under 20 years of age during this time with a purely intracapsular adenoma. The frequency of invasive adenomas in large series of pituitary tumors has been estimated at between 10% and 15%, so our experience would suggest a much greater occurrence rate in this age group.

Two other features also seem to occur frequently in adolescents who have invasive adenomas, namely, truncal obesity, and a relative preservation of normal pituitary function. All of our patients had varying...
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degrees of obesity and three of the four have not required replacement therapy. In one patient, the normal pituitary gland was seen by means of the operating microscope to be compressed against the posterior wall of the sella. Following removal of the tumor, the postoperative endocrine testing was normal.

A feature which is common to both adolescents and adults is the higher incidence of associated cranial nerve palsies. All four of our patients had such deficits and the tumors tended to involve the superior orbital fissure as well as the immediate parasellar area. Also in agreement with previous literature, there was no histological differentiation in these invasive tumors from the more typical intracapsular chromophobe adenomas. The term "invasive" simply represents involvement of contiguous structures and does not imply malignancy.

In conclusion, we suggest that pituitary adenomas in adolescents are characterized by early extrasella extension producing a clinical picture in which obesity and extraocular involvement are more frequent than visual field defects and hypopituitarism.

Addendum

Since the completion of this paper we have seen a 13-year-old child with an entirely intracapsular pituitary tumor.

References

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