Considering the Ectopic Pituitary Gland in Evaluation of the Nasopharyngeal Mass

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Nonadenoidal nasopharyngeal masses in children are rare and are usually benign lesions that arise from congenital inflammatory or neoplastic processes. Results of imaging studies can suggest the etiology but a biopsy is often required for definitive diagnosis. This case report demonstrates the need to carefully consider the potential origin of nasopharyngeal lesions and to recognize the rare risk of removing ectopic and functional pituitary tissue.

Report of a Case
A 13-month-old girl was referred for evaluation of an asymptomatic, solid nasopharyngeal mass found incidentally on magnetic resonance imaging (MRI) results obtained for evaluation of a morning glory disc anomaly at age 11 months. Results of the MRI revealed a minimally enhancing rounded 5-mm soft-tissue mass along the roof of the right nasopharynx, distinct from the adjacent adenoid tissue (Figure 1). The sella turcica was small and abnormally shaped, and a persistently patent craniopharyngeal canal was evident, measuring 2 mm superiorly. Additional tissue was evident inferiorly within the craniopharyngeal canal, suspicious for anterior pituitary tissue. A direct connection between the craniopharyngeal canal and the nasopharyngeal mass was not apparent.

Flexible fiberoptic nasopharyngoscopy through the nares revealed a smooth, pedunculated lesion predominantly obstructing the right choana and adherent to the posterior pharyngeal wall by a stalk. The pedunculated mass appeared distinctly separate from adjacent sessile adenoid tissue. The clinical and radiographic findings suggested the lesion might be ectopic pituitary tissue. Results of endocrine testing and normal levels of insulin-like growth factor 1, thyroid-stimulating hormone, free thyroxine, and adrenocorticotropic hormone excluded diagnosis of a secreting pituitary adenoma. Although histologic confirmation was desirable, we could not rationalize performing a biopsy since the lesion appeared benign and was asymptomatic, and the concern of it being ectopic pituitary tissue was justifiable given the abnormal size and shape of the child's sella turcica. Close observation with repeat fiberoptic examination and MRI were consequently advocated rather than biopsy.

Fiberoptic nasopharyngoscopy 6 months later revealed the mass ball-valving into the right choana and partially obstructing the left choana (Figure 2). Results of repeated MRI showed no significant growth of the mass and repeated testing to measure hormone levels were normal. Results of a follow-up examination a year later were similar, although there was an increase in surrounding adenoid tissue in this now 2½-year-old child.

Discussion
Pituitary Gland Embryologic Origins
Ectopic location of the pituitary gland in the roof of the nasopharynx is a rare congenital lesion that results from aberrant

IMPORTANCE Extracranial pituitary ectopia is an uncommon finding characterized by all or a portion of the pituitary gland situated in an aberrant location. Often these lesions come to clinical attention only once they begin to function abnormally or increase in size significantly.

OBSERVATIONS We describe an incidentally noted, asymptomatic pedunculated nasopharyngeal lesion in a 13-month-old girl that may be an extracranial pituitary gland. Consideration of this rare anomaly cautioned against performing a biopsy and may have prevented inadvertent removal of functional pituitary tissue. We describe the embryologic origins of ectopic pituitary tissue, discuss the differential diagnosis for nasopharyngeal lesions in children, and emphasize physical and radiologic findings suggestive of ectopic pituitary tissue to prevent potential inadvertent removal of this rare anomaly.

CONCLUSIONS AND RELEVANCE We report this unusual case to review embryologic origins of ectopic pituitary tissue and to alert otolaryngologists of the need to consider ectopic pituitary tissue in the diagnosis of nasopharyngeal lesions in children.

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The pituitary gland arises from 2 distinct structures: downward evagination of diencephalic neuroectoderm as well as upward extension of oral ectoderm, also known as Rathke pouch. The craniopharyngeal canal, which provides the opening for oral ectoderm to migrate superiorly into the sella, involutes early in development, leaving a bony layer separating the intracranial sella turcica from the extracranial sinuses and nasopharynx. Pituitary tissue has been previously described at various locations along this migratory path.3

Differential Diagnosis of Nasopharyngeal Masses in Children

Benign adenoid hypertrophy is the most common etiology of nasopharyngeal tissue; however, the differential diagnosis also includes congenital, inflammatory, and neoplastic lesions (Box). A brief review of the etiology of nasopharyngeal masses is presented here. Our patient’s lesion appears most consistent with ectopic pituitary tissue.

Cystic lesions of the nasopharynx include encephaloceles, Rathke cleft cysts, Thornwaldt cysts, and branchial cysts, among others. Often these benign cystic lesions are discovered incidentally on endoscopy or during routine imaging, similar to the lesion seen in our patient, although our patient’s le-
sion did not appear cystic. They may rarely present with sinonasal symptoms, including nasal obstruction, cerebrospinal fluid rhinorrhea, or visual disturbances. Magnetic resonance imaging is the initial diagnostic study of choice, although computed tomography is sometimes also indicated to better define the bony anatomy of the skull base.2

A nasopharyngeal encephalocele is a neural tube defect best visualized by nasal endoscopy or imaging. These slow-growing defects are often clinically silent but can present with nasal obstruction or visual disturbances. There is an association between basal encephaloceles and the suspected morning glory syndrome, which prompted the need for brain MRI in our patient.4 The morning glory disc anomaly consists of an enlarged optic disc at the center of chorioretinal pigmentation and a white tuft of glial tissue that overlies the central portion. Morning glory discs are usually unilateral and occur more commonly in African American females; visual acuity with morning glory discs is typically 20/200 OU. Magnetic resonance imaging is the diagnostic tool of choice for transsphenoidal encephaloceles.5 Endoscopically, the nasopharyngeal encephalocele appears as a nondistinct sac-like protrusion and may be associated with a persistent basilar skull bony defect. Magnetic resonance imaging is the initial imaging study of choice because it allows for the greatest detail to visualize dural sinuses seen in children.1 These lesions may not have a characteristic appearance; however, they are differentiated clinically and radiographically by their size, progression, lobular appearance, and bony destruction.

Extracranial ectopic pituitary tissue is rare and despite being congenital in origin has only been previously reported in adults.3 Nearly 80% of reported cases occur in the sphenoid or cavernous sinus, and 20% in the clivus and nasopharynx.9 Only 1 biopsy-proven case was reported exclusively in the nasopharynx, a thyroid-stimulating hormone–secreting adenoma in a 50-year-old woman with normal pituitary tissue who presented with symptoms of hyperthyroidism.9 Ectopic pituitary tissue has the potential to function normally, not function, or become a nodule for a pituitary adenoma. Often these lesions only come to clinical attention once they begin to function abnormally or increase in size significantly. It is for this reason that most of the literature on extracranial pituitary tissue focuses on adenomas; in fact, 62% of the cases reported are in women with a mean age of 50 years.9 These demographic and clinical factors make it difficult to find comparable cases in the literature9-15 (Table). Morning glory syndrome commonly occurs as an isolated ocular abnormality, but has rarely been associated with midline craniofacial defects or posterior pituitary ectopia and hypopituitarism.5

Clinical findings and imaging studies are of paramount importance in determining the etiology of a nasopharyngeal lesion. Computed tomography and MRI are both useful, but MRI is our preferred initial diagnostic study in pediatric patients as it is not associated with ionizing radiation. The MRI results in this case demonstrate the hypophyseal (craniopharyngeal) canal well. It is funnel shaped, measuring 2 mm superiorly and
1 mm inferiorly. Any persistence of a hypophyseal canal extending from the floor of the sella to the nasopharynx is a rare congenital defect. Even rarer is a canal with a diameter greater than 1.5 mm, which is also much more likely to be associated with an ectopic pituitary gland, encephalocele, or tumor.6 The pedunculated nature of the mass presented here, the smooth appearance of its surface, the absence of bony destruction, and the lack of significant growth across time make a neoplastic appearance of its surface, the absence of bony destruction, and the finding of a small sella turcica, a patent craniopharyngeal canal, and a stalk of tissue protruding from the pituitary into the patient’s cranioophyseal canal support the etiology of ectopic pituitary tissue.

Potential risks of performing a tissue biopsy to definitively determine the etiology of our patient’s mass include hemorrhage, infection, or hypopituitarism. Hypopituitarism is a highly variable group of disorders with heterogeneity of hormonal deficiencies in regard to both the specific hormones and the degree of paucity. However, regardless of the specific cause or cohort examined, all-cause mortality in patients with hypopituitarism is increased when compared with age- and sex-matched controls, even with hormone therapy.7

Without obtaining a biopsy we cannot confirm that the nasopharyngeal lesion is pituitary; however, the presence of a patent craniophyseal canal and a small and abnormally shaped sella suggests that the pituitary is abnormally developed and that the gland, or portion of the gland, is not in its usual location. The pedunculated mass is suspicious for pituitary tissue but the risk to benefit ratio of biopsying the lesion and possibly affecting the patient’s pituitary function is too great to accept, so biopsy was deferred. Future management includes serial examinations, interval imaging, and laboratory studies. Biopsy may never be indicated if clinical symptoms of obstruction or hormone excess do not develop.

Characteristics that would warrant a biopsy to exclude a malignant nasopharyngeal lesion include bony destruction, invasion into adjacent structures, or rapid growth.8 We recommend preoperative MRI prior to biopsy of nasopharyngeal lesions to exclude an intracranial communication or abnormal pituitary development that may indicate the presence of functional ectopic pituitary tissue.

Conclusions

We describe a rare case of a nasopharyngeal mass in a 13-month-old girl that clinically and radiographically is consistent with an ectopic anterior pituitary gland, presumed to be functional tissue given the child’s small sella turcica. Although most nasopharyngeal masses in the pediatric population are benign, biopsy or removal is sometimes indicated and should be preceded by imaging studies to consider the possibility of ectopic pituitary tissue. Deferring biopsy may prevent hypopituitarism caused by inadvertent removal of functioning normal but ectopic pituitary tissue.

Table. Previously Reported Cases of Nasopharyngeal Ectopic Pituitary Tissue*

<table>
<thead>
<tr>
<th>Source</th>
<th>Patient Age, y/Sex</th>
<th>Location of Tissue</th>
<th>Presenting Symptoms</th>
<th>Biopsy Performed</th>
<th>Sellar Involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chessin et al, 12, 1976</td>
<td>64/F</td>
<td>Sphenoid and nasopharynx</td>
<td>Nasal congestion</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Rasmussen and Lindholm, 13, 1979</td>
<td>36/M</td>
<td>Middle meatus</td>
<td>Visual disturbance</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Anand et al, 14, 1993</td>
<td>58/F</td>
<td>Clivus and nasopharynx</td>
<td>Nasal obstruction, anosmia, visual disturbance</td>
<td>Yes</td>
<td>Not identified</td>
</tr>
<tr>
<td>Slonim et al, 15, 1993</td>
<td>76/F</td>
<td>Pharynx</td>
<td>Cushing syndrome</td>
<td>Yes</td>
<td>Not identified</td>
</tr>
<tr>
<td>Collie and Collie, 9, 2005</td>
<td>50/F</td>
<td>Nasopharynx</td>
<td>Hyperthyroidism</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Hong et al, 10, 2012</td>
<td>48/M</td>
<td>Sphenoid</td>
<td>Acromegaly</td>
<td>Yes</td>
<td>Empty sella</td>
</tr>
<tr>
<td>Rabelink et al, 11, 2012</td>
<td>38/F</td>
<td>Nasopharynx</td>
<td>Galactorrhea</td>
<td>No</td>
<td>Sella not recognized</td>
</tr>
</tbody>
</table>

* All cases presented symptomatically in adulthood.


