Prolactin-Secreting Clival Ectopic Pituitary Adenoma

Lydia A Leavitt, BS*

University of Illinois College of Medicine, Rockford, Illinois, USA

*Corresponding author: Lydia A Leavitt, BS, University of Illinois College of Medicine, Rockford, Illinois, USA, Tel: 608-345-8349

Abstract
Background: Ectopic pituitary adenomas are an extremely rare subset of pituitary tumors, but when present, are often misdiagnosed. This particular case illustrates a unique presentation of an already uncommon disease.

Case presentation: A 69-year-old female with a 30-year history of hyperprolactinemia symptomatically controlled with dopamine agonist therapy presented with complaints of worsening headaches. Findings on physical examination and endocrine studies were unremarkable. Magnetic resonance imaging of the brain revealed a mass in the clivus. The patient underwent surgical excision of the lesion and the resected tissue was consistent with an ectopic prolactin-producing pituitary adenoma. Her dopamine agonist therapy was discontinued following surgery and her endocrinologic function normalized with no further therapy needed.

Conclusion: Although a prolactin-secreting ectopic pituitary adenoma in the clivus is uncommon, it should be part of the differential diagnosis for clival lesions—especially when the patient has a known history of endocrine dysfunction. Surgery is not always the first-line treatment option for pituitary adenomas. Dopaminergic therapy is capable of controlling symptoms of hyperprolactinemia, but surgery is necessitated when symptoms of a space-occupying mass lesion occur. Resection was curative for this patient.

Keywords
Ectopic pituitary adenoma, Pituitary adenoma, Prolactinoma, Hyperprolactinemia

Introduction

Pituitary tumors are common intracranial neoplasms accounting for 10-15% of intracranial tumors [1,2]. 90% of pituitary tumors are adenomas, which are typically confined to the sella turcica. However, on rare occasions, the adenoma resides exclusively outside of the sella turcica—called an ectopic pituitary adenoma (EPA) [1]. EPAs are benign neoplasms found in a variety of intra- and extracranial locations. Current knowledge about EPAs comes from the relatively few single case reports found in medical literature. Additional studies of EPAs are needed to understand clinical presentation patterns, associated morbidities, and treatment options.

Case Study

MG is a 69-year-old female with a past medical history of asthma and hypertension who initially presented about 30 years ago with symptoms of hyperprolactinemia—bilateral nipple discharge, menstrual irregularities, and vaginal dryness. She affirmed that laboratory results at that time showed an elevated prolactin level, but all other labs were unremarkable. MG also reported that CT scans of the thorax, abdomen, and pelvis were negative for masses, and brain MRI was similarly unremarkable. Our treatment team, however, was unable to locate the patient's initial laboratory data and imaging. MG's physician at that time thought her symptoms were likely secondary to a microprolactinoma too small to be visualized on pituitary imaging. She was treated with dopamine agonist therapy with successful control of her symptoms, remaining asymptomatic for 30 years.

MG presented to our clinic with a primary complaint of headache. She began experiencing occasional mild headaches several months ago, but the headaches progressively became more frequent and severe. MG also reported a several-month history of diminished taste, but otherwise denied any other symptoms, including visual changes, diplopia, facial pressure, or facial pain. Her physical examination was unremarkable, including fully intact extraocular movements and no afferent pupillary defect. Follow-up MRI imaging revealed a...
uniform contrast-enhancing clival mass extending laterally to the petroclival junction without invasion into the cavernous sinuses. All laboratory results, including endocrine testing, were unremarkable.

Our top differential diagnoses included ectopic pituitary adenoma (EPA), chondroma, meningioma, astrocytoma, and metastatic tumor. Given the patient’s history of hyperprolactinemia, an EPA was thought to be the most likely diagnosis. The patient agreed to surgical resection via an endoscopic endonasal approach. Intraoperatively, it was confirmed that the sellar dura was completely intact and the mass originated from the clivus. Gross-total resection, which was confirmed with postoperative imaging, was achieved without complication. Frozen-section pathology was consistent with postoperative imaging, was achieved without surgical confirmation, and involved no further pharmacologic therapy. Following the resection, the patient discontinued her dopamine agonist therapy. The patient was discharged on postoperative day two with normal endocrine function and required no further pharmacologic therapy.

Discussion

EPAs are rare tumors almost exclusively found in locations along the pituitary migratory path and are thought to arise from embryological remnants of the pituitary gland [2]. The most common locations for EPAs are in the sphenoid sinus and suprasellar region, which together account for approximately 60% of EPAs [2,3]. Other relatively common locations include the clivus, nasal cavity, cavernous sinus, parasellar region, and sphenoid wing. EPAs have also been reported in sites beyond the pituitary migratory path including within the petrosal temporal bone, superior orbital fissure, third ventricle, and temporal lobe [3].

Clinical manifestations of EPAs typically arise secondary to hormonal derangements and involvement of adjacent structures causing symptoms of mass effect [1-5]. Common hormonal derangement presentations include Cushings syndrome secondary to hypercortisolism, acromegaly secondary to growth hormone excess, and hypogonadism and/or galactorrhea secondary to hyperprolactinemia [1,2]. Other presentations include Cushing’s syndrome secondary to hypercortisolism, acromegaly secondary to growth hormone excess, and hypogonadism and/or galactorrhea secondary to hyperprolactinemia [1,2]. Other presentations include Cushing’s syndrome secondary to hypercortisolism, acromegaly secondary to growth hormone excess, and hypogonadism and/or galactorrhea secondary to hyperprolactinemia [1,2]. Other presentations include Cushing’s syndrome secondary to hypercortisolism, acromegaly secondary to growth hormone excess, and hypogonadism and/or galactorrhea secondary to hyperprolactinemia [1,2]. Other presentations include Cushing’s syndrome secondary to hypercortisolism, acromegaly secondary to growth hormone excess, and hypogonadism and/or galactorrhea secondary to hyperprolactinemia [1,2].

EPA pathogenesis

EPAs are thought to arise from neoplastic transformation occurring within ectopic pituitary tissue that remains along the embryonic pituitary gland migratory path [1-5]. To understand the proposed pathologic transformation that occurs, it is beneficial to briefly review pituitary gland embryogenesis and anatomy.

Pituitary gland embryogenesis [7,8]

The pituitary gland arises from Rathke’s pouch cells, which arise from the surface ectoderm of the pharyngeal roof. During the fourth week of embryogenesis, the ectoderm cells migrate from the oral cavity superiorly towards the floor of the diencephalon to form Rathke’s pouch. Between weeks six and eight of embryogenesis, the connection between Rathke’s pouch and the pharynx degenerates, while a connection between Rathke’s pouch and an inferior invagination of the neuroectoderm is formed. The creation of this connection positions the pituitary gland in the sella turcica of the sphenoid bone.

Pituitary gland anatomy [7-9]

The pituitary gland is divided into the neurohypophysis (posterior lobe) and the adenohypophysis (anterior lobe). The neurohypophysis is comprised of the pars nervosa, which serves as the storage site for oxytocin and vasopressin, and the infundibular stalk, which is the connection between the pituitary and hypothalamus. The continuity of the infundibular stalk is essential for adequate development and proper functioning of the pituitary gland.

The adenohypophysis has three divisions—the pars distalis, pars intermedia, and pars tuberalis. The pars distalis is the secretion site for the pituitary hormones ACTH, TSH, FSH, GH, LH, and prolactin. The pars intermedia forms a boundary between the adenohypophysis and neurohypophysis. The pars tuberalis is an extension of the pars distalis and surrounds the infundibular stalk of the neurohypophysis.
Current theories of EPA pathogenesis are as follows [5]

1. During migration, remnant cells of Rathke’s pouch localize outside of adenohypophysis and become progenitor cells of EPAs.
2. Suprasellar EPAs arise from pituitary cells of the subdiaphragmatic portion of the pars tuberalis.
3. During embryonic development, an abnormal fusion of the post-sphenoid cartilage results in craniofaryngeal duct that extends from the sella turcica to the nasopharynx, which allows for aberrant migration of pituitary cells.
4. EPAs result from a lack of hypothalamic regulation of pituitary development during embryogenesis and subsequent impaired dopamine delivery.

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Conflict of Interest

The author does not have conflicts of interest to disclose.

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