CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 9 (2015) 57–60

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Primary ectopic frontotemporal craniopharyngioma

Luis Alberto Ortega-Porcayo[a], Juan Antonio Ponce-Gómez[a], Mauricio Martínez-Moreno[a], Lesly Portocarrero-Ortíz[b], Martha Lilia Tena-Suck[c], Juan Luis Gómez-Amador[a,*]

[a] Department of Neurosurgery. National Institute of Neurology and Neurosurgery "Manuel Velasco Suarez" Mexico City, Mexico
[b] Department of Neuroendocrinology. National Institute of Neurology and Neurosurgery "Manuel Velasco Suarez", Mexico City, Mexico
[c] Department of Neuropathology. National Institute of Neurology and Neurosurgery "Manuel Velasco Suarez" Mexico City, Mexico

ARTICLE INFO

Article history:
Received 23 September 2014
Received in revised form
22 December 2014
Accepted 27 December 2014
Available online 8 January 2015

Keywords:
Ectopic
Craniopharyngioma
Endoscopic
Keyhole

ABSTRACT

INTRODUCTION: Primary ectopic craniopharyngiomas have only rarely been reported. Craniopharyngiomas involve usually the sellar and suprasellar region, but can be originated from cell remnants of the obliterated craniopharyngeal duct or metaplastic change of andenohypophyseal cells. We present the first case of a primary ectopic frontotemporal craniopharyngioma.
PRESENTATION OF CASE: A 35-year old woman presented with a one-year history of headache and diplopia. MRI showed a large frontotemporal cystic lesion. Tumor resection was performed with a keyhole endoscopic frontal lateral approach. The pathological features showed an adamantinomatous craniopharyngioma with a cholesterol granuloma reaction.
DISCUSSION: There have been reported different localizations for primary ectopic craniopharyngioma. Our case presented a lobulated frontotemporal cystic mass formed by a dense eosinophilic proteinaceous material dystrophic calcifications and cholesterol crystals, with epithelial remnants. No tumor regrowth was observed in the magnetic resonance image 27 months postoperatively.
CONCLUSION: Primary ectopic craniopharyngioma is a rare entity with a pathogenesis that remains uncertain. This is an unusual anatomic location associated with unique clinical findings.

1. Introduction

Craniopharyngioma is an epithelial tumor that arise from the remnants of Rathkés pouch, they often involve the sellar/suprasellar region; but primary ectopic [1–10] localizations had been reported since 1924 when Bock described an isolated infracranial tumor [11]. They arise from epithelial remnants anywhere along the obliterated craniopharyngeal tract from Rathkés cleft to the floor of the third ventricle. Two common patterns of ectopia have been described: contamination with tumor cells along the surgical tract and spreading via CSF and the subarachnoid space [12]. Retrostalk growth may extend to the posterior fossa and mismigrated cleft cells may originate tumor growth in other compartments [4].

2. Case report

2.1. Clinical findings and evaluation

A 35-year old woman presented with a one-year history of intermittent bilateral frontal headache of moderate severity, worsened with bending and improved with NSAIDís. Fifteen days before admission the severity increased associated with horizontal binocular diplopia and transient metamorphopsias. On examination she had papilledema, no visual field defects and left lateral rectus paresis. The pituitary hormonal test was normal.

MRI revealed a large frontotemporal cystic lesion with a rostral small solid nodule, isointense in T1, hypointense in T2, with heterogeneous contrast enhancement of the solid nodule (Fig. 1A and B). CT showed a frontotemporal cystic lesion with an irregular heterogenous calcified rostral nodule. The sellar and suprasellar region appeared intact without invasion.

2.2. Surgical technique

The patient was positioned supine with the head tilted to the contralateral side. We started with a 4 cm frontal curvilinear skin incision. The temporal muscle was transected and the fibers were retracted. A keyhole craniotomy of 3 × 2.5 cm was performed and the dura was opened in a curved fashion. A 7 mm

Abbreviations: CSF, cerebrospinal fluid; CT, computed tomography; KPS, karnofsky performance status scale; MRI, magnetic resonance imaging.

* Corresponding author. Tel.: +52 5556063822; fax: +52 5556063822.
E-mail address: jlga@neurocirugia-innn.com (J.L. Gómez-Amador).

http://dx.doi.org/10.1016/j.ijscr.2014.12.038

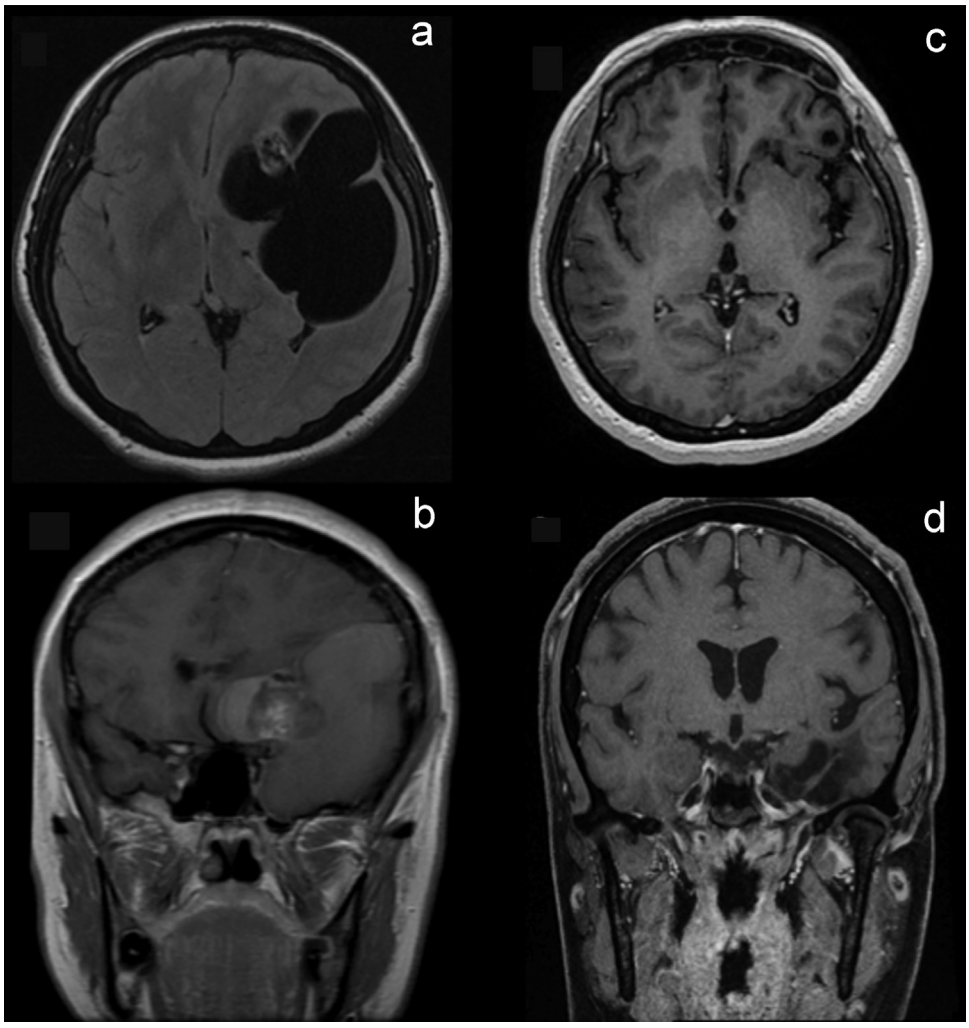

Fig. 1. Preoperative MRI (A and B) notice that midline structures and sellar compartment is not involved. Postoperative MRI (C and D) after 27 months revealed no residual tumor.

0° Karl Storz endoscope was introduced into the cystic cavity. A brownish fluid was drained and the natural cystic cavity was used for endoscopic navigation. Tumor resection was performed with endoscopic visualization with two-handed microsurgical technique with microsurgical dissectors, microscissors, aspirator and bipolar coagulation (see video).

2.3. Postoperative course

Left rectus paresis improved immediately after surgery. No diabetes insipidus, electrolyte imbalances or hormone deficiencies were documented during her hospital stay. KPS was 100 after surgery and during the follow up. MRI 27 months after surgery was negative to any residual tumor (Fig. 1C and D).

2.4. Pathology

The gross aspect of the tumor resected was a lobulated mass with spongy consistency and a cystic component of brownish thick liquid. Histologically we found a stratified epithelium remnant (Fig. 2D), immersed in dense, necrotic, eosinophilic and proteinaceous material (Fig. 2B), which formed cleft cholesterol (Fig. 2A) and dystrophic calcifications (Fig. 2C). No stellate reticulum was observed. Histopathology was suggestive of adamantinomatous variety of craniopharyngioma with a cholesterol granuloma reaction.

3. Discussion

3.1. Pathogenesis

The first tumor description of a craniopharyngioma was published by Mott and Barratt [13], in which they proposed that craniopharyngiomas might arise from the hypophyseal duct. In 1904, the pathologist Jakob Erdheim [14] was surprised to find small clusters of squamous epithelium above the cyst, at the junction of the infundibulum and pituitary. He proposed that craniopharyngioma originated from the embryonic squamous cell remnants of the obliterated craniopharyngeal duct. This theory explains the embryological reason for most of the adult craniopharyngiomas including the extracranial nasopharyngeal craniopharyngioma, unless they lacked of dural covering, intracranial extension or deformation on the sella; supporting the theory of ectopic infrasellar tissue [7]. The anterior, posterior and lateral extension has been reported extensively, but an ectopic craniopharyngioma does not have an intrasellar or suprasellar involvement. A primary ectopic craniopharyngioma is a tumor that arises for the first time without any previous surgery in any compartment with no midline involvement from remnants of the