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**Case Report**


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## Isolated petrous apex ectopic craniopharyngioma

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**ABSTRAK**


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*Kraniofaringioma ektopik primer jarang sekali dijumpai, dan hingga saat ini belum pernah ada laporan yang menyampaikan lokasinya yang ektopik di apeks tulang petrosus. Penelitian ini melaporkan satu kasus pria berusia 26 tahun yang mengeluhkan nyeri kepala dan pandangan ganda akibat kelemahan saraf kranialis VI (nervus abducent). Hasil pemeriksaan CT dan MRI memperlihatkan lesi kistik di dalam petrous apex kanan. Tidak terlihat tanda-tanda keterlibatan bagian sella maupun suprasella. Pasien menjalani operasi pengangkatan tumor via prosedur endoskopi endonasal transphenoid. Pemeriksaan histopatologi menunjukkan suatu kraniofaringioma adamantinomatous. Sejauh ini, kasus ini merupakan laporan kasus pertama kraniofaringioma ektopik primer yang terisolasi pada apeks tulang petrosus. Kraniofaringioma ektopik primer terjadi karena proses multifaktor yang diawali oleh kesalahan embriologi saat migrasi sel, dan laporan kasus ini bisa menjadi fakta pendukung terhadap pemahaman tersebut.*

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**ABSTRACT**


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Primary ectopic craniopharyngioma is a rare entity. Isolated petrous apex bone location has not been reported previously. This study reports a case of 26-year-old male with right abducent nerve palsy. CT and MRI imaging reveal right petrous apex cystic lesion. No sellar or suprasellar region involvement was found. Endoscopic endonasal transphenoid approach has been successfully performed. Histopathology examination confirms the diagnosis of adamantinomatous craniopharyngioma. So far, it's probably the first case report of primary ectopic craniopharyngioma isolated in the petrous apex. This case report supports the premise that primary ectopic craniopharyngioma is a multifactorial process that starts with an error from migrated embryological cells.

**Keywords:** petrous apex tumor, primary ectopic craniopharyngioma, transphenoid

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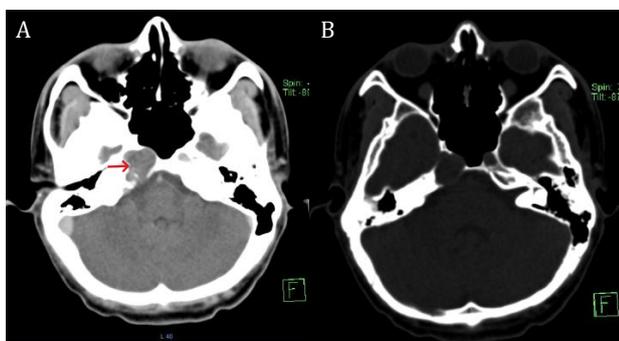
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Craniopharyngiomas are considered as benign tumors, but locally aggressive. It derived from epithelial remnants of Rathke's pouch. It accounts for 1.2% to 4.6% of all intracranial neoplasms.<sup>1-3</sup> Commonly it is located at the suprasellar region, either anterior or posterior to the optic chiasma.<sup>1-5</sup> Isolated infrasellar craniopharyngiomas without sellar involvement are extremely rare.<sup>4,6</sup> Isolated craniopharyngioma in the petrous bone has never been reported. This study reports an extremely rare case; histologically confirmed primary ectopic craniopharyngioma that arose at the apex of right petrous bone.

### CASE REPORT

A 26-year-old male came with a six months history of intermittent headache, and non-specific location. He usually took pain-killer drug (paracetamol), but in the last two months, the symptom worsened and the patient had double vision every time he looks to the right side.

Right abducent nerve palsy was found through physical examination. Laboratory examination was normal. Computed tomography (CT) scan showed an isolated cystic hypodense mass (2 cm) at the right petrous apex bone isolated from sella turcica which did not enhance after injection of contrast medium. The mass slightly protruded to the sphenoid sinus, prepontine cistern, carotid canal, and Meckel's cave (Figure 1). The magnetic resonance imaging (MRI) revealed a 22 mm x 29 mm x 30 mm cystic lesion on the apex of the right petrous bone which appeared to be hyperintense on both

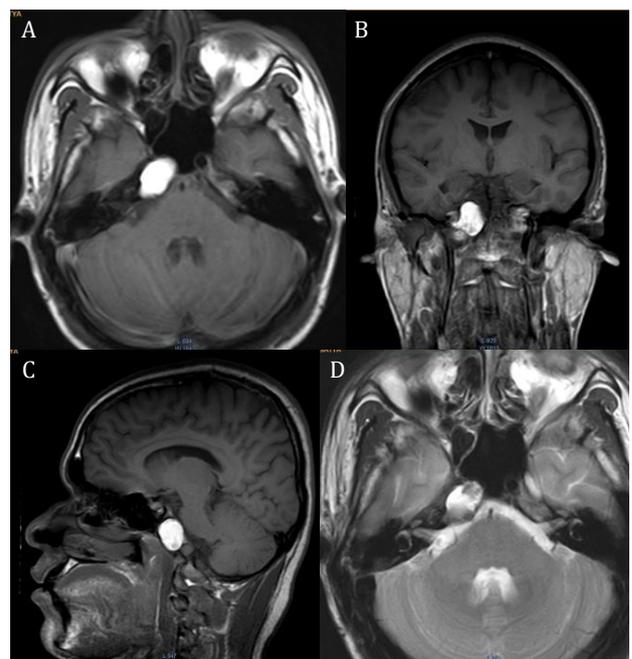


**Figure 1.** Axial brain window with contrast (A) and bone window (B) CT scan showed osteolytic cystic lesion in the apex of right petrous bone, which did not enhance with contrast administration.

MRI T1 weighted without contrast and MRI T2 weighted images (Figure 2). The differential diagnosis included an epidermoid cyst, cholesterol granuloma and primary ectopic craniopharyngioma.

The patient underwent endoscopic endonasal transphenoid surgery because this approach provides direct view to the lesion. In the sphenoid sinus, the lesion was seen as a protrusion of petrous apex bone, at the right side of clivus (Figure 3). The protruded bone was drilled using high speed drill, with a 4 mm diamond drill head. When the tumor capsule was opened with telescopic knife, the classical appearance of brownish "Machinery oil" fluid was drained from the cyst. Most part of the capsule was removed during surgery, and the capsule was sent to histology for review. After the surgery, the double vision and headache were improved remarkably; the patient was discharged on day three after surgery without any major post-surgical complications. Histopathological examination confirmed the diagnosis of adamantinomatous craniopharyngioma (Figure 4).

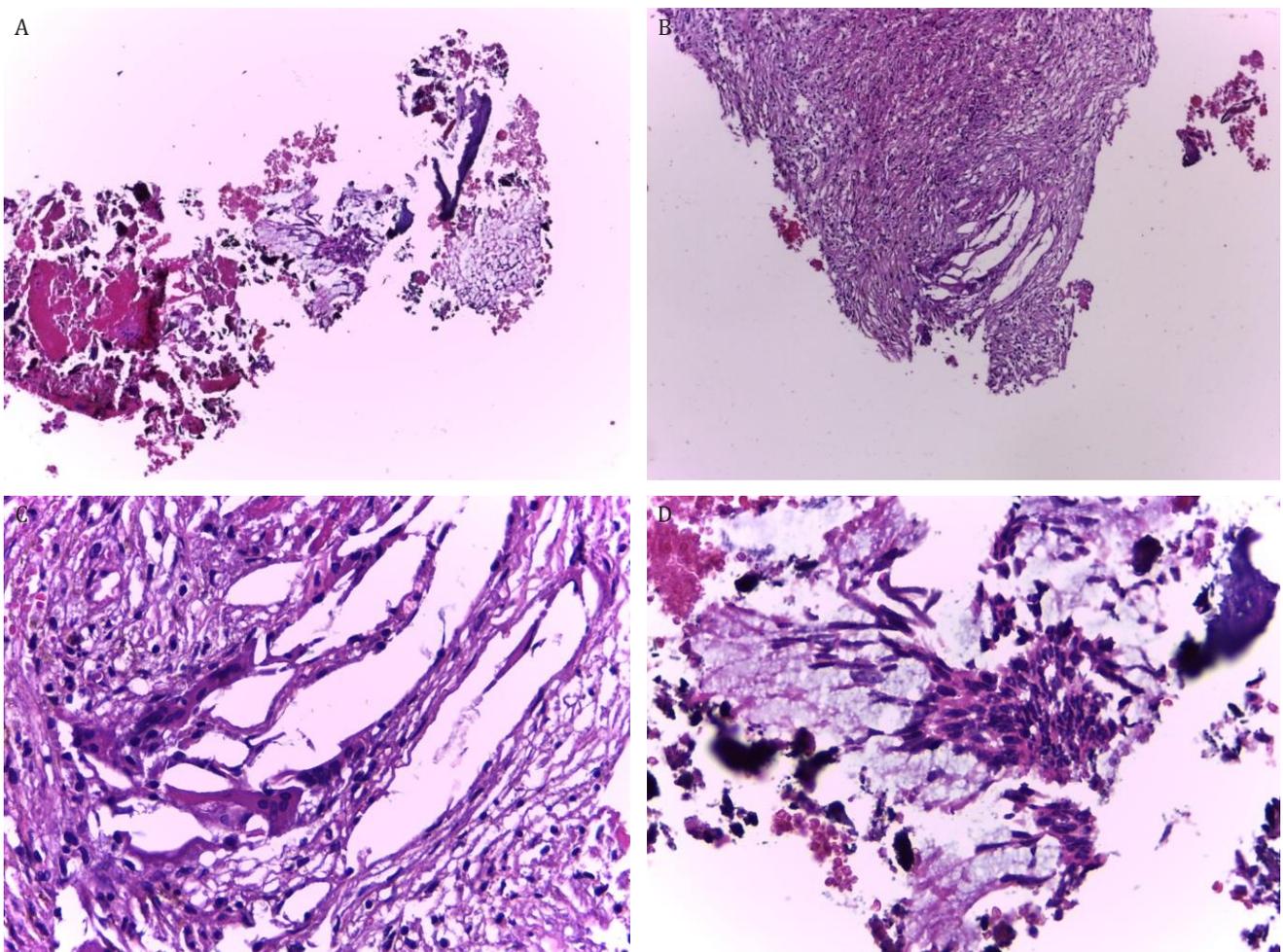
To complete the treatment, gamma knife surgery was done two weeks after the surgery.



**Figure 2.** Axial (A), coronal (B), sagittal (C) T1-weighted, and axial T2-weighted (D) MR images demonstrating a hyperintense cystic lesion in the apex of right petrous bone that protruded to sphenoid sinus and prepontine cistern.



**Figure 3.** Intraoperative photo showing: (A) bony protrusion on the right side of clivus towards the sphenoid sinus. (B) Yellow capsule was revealed after drilling of the posterior wall of the sphenoid sinus. (C) "Machinery oil" with cholesterol crystal was drained from the cyst



**Figure 4.** Photomicrographs light microscopy studies of surgical specimen showing: (A) Overview of the specimen. (B and C) Clefts of cholesterol crystals with foreign body giant cell reactions. (D) A cluster of epithelial cells in the center of the field. H & E. Original magnification x100 (A and B), and x 400 (C and D)

## DISCUSSION

Craniopharyngioma is an intracranial benign tumor believed to originate from the epithelium

of Rathke's pouch. An ectopic craniopharyngioma means the tumor does not have an intrasellar or suprasellar involvement.<sup>7</sup> The post-operative occurrence of ectopic craniopharyngiomas by direct mechanical transplanted or meningeal

seeding is a recognized phenomenon.<sup>3,8</sup> Nevertheless, histologically confirmed primary craniopharyngioma at an ectopic location is rare, although cases of epidural,<sup>5</sup> frontotemporal,<sup>8</sup> posterior fossa,<sup>2,6,9</sup> and infrasellar craniopharyngiomas<sup>1,4,10,11</sup> have been reported. On the other hand, primary isolated petrous apex craniopharyngioma has not been previously reported.

Two hypotheses have been proposed for the development of a craniopharyngioma. The first was proposed by Erdheim, in which they may arise from any location along the craniopharyngeal duct. The second supports the squamous cell nests that may derive from metaplastic change of adenohypophyseal cells.<sup>1,7</sup> It has been accepted that aberrations in the sequence of origin during embryogenesis result in the formation of craniopharyngiomas and their variable location. In the beginning of embryogenesis processes, glandular part arises from the out pouching of the ectodermal epithelium covering the roof of stomadeum, called the Rathkes diverticulum. The Rathkes diverticulum then invaginates and penetrates the mesenchyme in the direction of the diencephalon and forms the Rathkes pouch. The cell of Rathkes pouch rapidly proliferates and follows the sphenoid bone development until it get to the sella turcica. The Rathkes pouch comes together with neuroectoderm of neurohypophysis, which descends from third ventricle of embryonic brain, and finally become adenohypophysis. The extracranial passage is extending from the junction between the posterior nasal septum and palatum to the anterior part of the sphenoidal hypophyseal fossa.<sup>1,4</sup> In this case, the tumor was located in the right petrous apex with no connection to the craniopharyngeal duct. So far, there is no consensus for a primary ectopic craniopharyngioma. The cause for primary ectopic craniopharyngioma was believed as multifactorial and it starts with an error in mismigrated embryological cells in addition to mutations of oncogenes and tumor suppressor genes in which different endocrinological growth and vascular factors produce an ideal environment.<sup>7</sup>

The clinical manifestations are defined by the tumor's location that compressing the surrounding neurovascular structures. Generally, the symptoms could consist of visual, endocrine, or cognitive disturbance, and all the symptoms related with high intracranial pressure.<sup>1-11</sup> In

this case, the symptom was only double vision with esotropia (in-turning eye) on examination. We believed that it was caused by compression on the sixth nerve by tumor protrusion to the prepontine cistern and the direct compression to the Dorello's canal. It's supported by the fact that the patient's symptoms were improved remarkably after surgery, the patient was sent home on the third day after surgery with almost no double vision and headache.

CT scan and MRI are preferred when diagnosing craniopharyngiomas.<sup>1,4,6</sup> CT scans play an important role in identifying bony anatomy and calcifications. MRI will show tumor extension and its consistency, both solid or cystic part, enhanced scanning range and the relationship to adjacent neurovascular structures. Although CT scans are more specific to display calcified tissue, MRI can provide detail measurement of tumor range for the surgical approach. It is not difficult to diagnose typical craniopharyngioma, but for the non-typical location and imaging manifestations, the combination of both CT and MRI would be very helpful. The differential diagnosis would be epidermoid cyst, but usually the diffusion MR would be able to differentiate epidermoid from other lesion.

The direct surgical approach through sphenoid sinus is very simple and provides direct view for the surgeon. When the tumor capsule was opened, "machinery oil" fluid with shimmering cholesterol crystals came out which suggested craniopharyngioma. It was confirmed adamantinomatous craniopharyngioma. Adamantinomatous type seen mainly in children with poor overall outcome and a squamous papillary type which is more common in adults with a better prognosis. The cystic components in adamantinomatous type are often described as having a characteristic "machinery oil" interior, containing desquamated squamous epithelium and comprised mainly of keratin and cholesterol.<sup>12</sup>

Although during surgery we believe that we had removed most part of the tumor capsule, we still feel it is necessary to apply radiation to the tumor bed to prevent recurrence.

Primary ectopic craniopharyngioma, especially isolated in the apex petrous bone is a very rare case.

In conclusion, to the best of author's knowledge, it's probably the first case report of primary ectopic craniopharyngioma isolated in the petrous apex. This case report supports the premise that primary ectopic craniopharyngioma is a multifactorial process that starts with an error from mismigrated embryological cells.

### Conflicts of interest

The authors affirm no conflict of interest in this study.

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