CASE REPORT

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Atypical Craniopharyngioma

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ABSTRACT

Magnetic resonance (MR) is the preferred method for diagnosing craniopharyngiomas. It best demonstrates the morphology of the tumor, its extent, and its association with surrounding structures. Variable appearance of craniopharyngiomas on MR images reflects diverse histologic appearance. We present a case of an atypical craniopharyngioma with high signal intensity in all MR sequences.

Keywords: Craniopharyngioma, parasellar region, magnetic resonance imaging

INTRODUCTION

A craniopharyngioma is a benign slow-growing dysontogenetic tumor of the central nervous system derived from the Rathke pouch epithelium and usually located in the sellar and parasellar regions. Craniopharyngiomas are rare and account for 3% of all intracranial tumors, with a rate of 0.5-2 cases per million per year. It has a classical bimodal age distribution: first peak incidence between 5 and 14 years of age and second peak incidence in adults older than 65 years. The incidence rate does not correlate with gender, race, or geographical location [1]. The association with medical conditions or genetic predispositions has not yet been verified. Magnetic resonance imaging (MRI) is the preferred method for identifying craniopharyngiomas and evaluating their location, tumor extension, and relationship with surrounding structures. Arriving at a correct diagnosis can be challenging because craniopharyngiomas have a very diverse appearance on MR scans because of their complex histological structure [2].

CASE PRESENTATION

A 49-year-old man presented with chronic headaches, nausea, and dizziness for 4 months. MRI demonstrated a 61 × 57 × 42 mm cystic mass with a lobulated configuration located in the parasellar region extending to the pontine cistern and the inferior part of the third ventricle. Contrast-enhanced T1-weighted image demonstrates heterogeneous enhancement of the solid parts of the lesion. The pons and midbrain were compressed and displaced posteriorly. The basilar artery was encased, and the circle of Willis was compressed by the tumor. The tumor appeared to have high signal intensity on all T1-, T2-, and FS T1-WI. Although the inferior part of the third ventricle was compressed, there was no evidence of obstructive hydrocephaly (Figure 1).

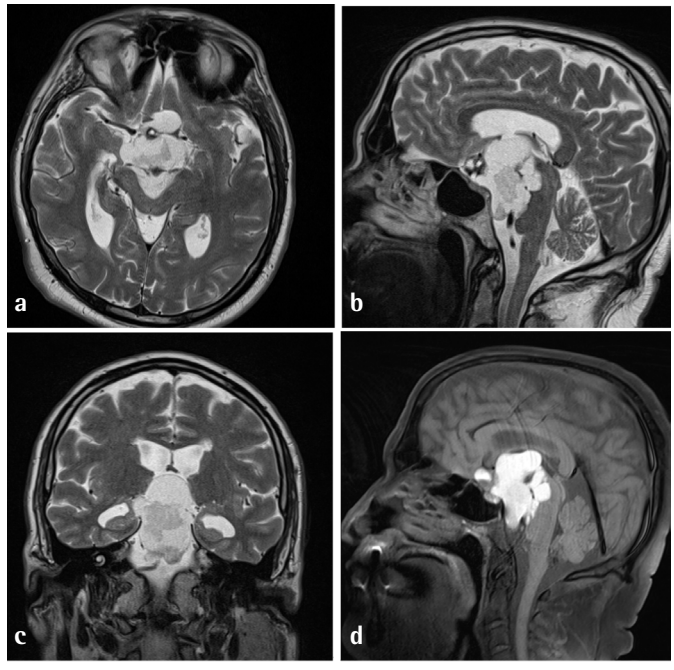

Figure 1. (a-c) Axial, sagittal, and coronal T2-weighted images. (d) Sagittal fat-saturated T1-weighted images demonstrate a large cystic mass in the parasellar region, which shows high signal intensity in all sequences

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DISCUSSION

There are two distinct pathologic types of craniopharyngiomas, each with its own theory of origin. The adamantinomatous type is thought to originate from remnant ectodermal cells of the craniopharyngeal duct, which are formed as a result of the migration of Rathke's pouch, a diverticulum arising from the embryonic buccal cavity that gives rise to the anterior pituitary gland (adenohypophysis). The origin of squamous papillary craniopharyngiomas is explained by metaplastic theory, which suggests that they develop because of metaplasia and further proliferation of residual embryonal adenohypophyseal cells of the pars tuberalis of the pituitary. The second type predominantly occurs in adults, whereas the first can affect all age groups [3]. Macroscopically, craniopharyngiomas are solid and/or cystic lesions; the adamantinomatous type often contains calcifications.

Although craniopharyngiomas are benign tumors the World Health Organization grade, they usually show a high recurrence rate and can be clinically aggressive depending on the location and size of the tumor [4]. Craniopharyngiomas are usually located in the sellar/parasellar region. The sellar region includes the sella turcica and pituitary gland. The parasellar region includes the cavernous sinuses, suprasellar (chiasmatic) cistern, hypothalamus, and ventral inferior third ventricle along with vessels, nerves, and meninges. Compression of the optic chiasm can lead to visual disturbances, such as bitemporal hemianopsia, diplopia, and optic nerve atrophy. If the tumor grows inferiorly, it can compress the pituitary gland or hypothalamus, which can cause a variety of problems related to endocrine dysfunction, such as growth retardation (in children), hypogonadism (in adults), neurohormonal diabetes insipidus, menstrual disorders, and hypothalamic obesity [5]. The most common symptoms are headaches, nausea, and vomiting due to increased intracranial pressure.

The diagnosis of craniopharyngioma is based on clinical examination along with radiological findings and is then confirmed by histopathological findings. Computed tomography is very useful in identifying calcifications that are seen in 90% of the cases, but MRI is the gold standard for sellar region assessment because it best demonstrates the morphology of the tumor, its localization, extent, and association with surrounding structures such as the hypothalamus. MR angiography helps to evaluate its involvement with the vessels and to differentiate a tumor from a possible arteriovenous malformation [6].

The wide range of histological appearances of craniopharyngiomas were reflected in their MR appearances. Adamantinomatous usually has cystic components and calcifications in 90% of cases. The papillary types of tumors are mostly solid. Solid parts and the cyst capsule are hypointense relative to the brain and show enhancement following gadolinium administration on pre-contrast T1-weighted images. The cystic fluid has variable signal intensity, depending on protein and cholesterol concentration, and is usually hypointense or hyperintense on T2-weighted images [7]. High intensity on T1-weighted images was noted in cystic lesions with high cholesterol, protein content , or methemoglobin levels.

Atypical craniopharyngiomas have diverse MRI appearances, which can complicate their diagnosis and differentiate them from other brain tumors. Therefore, a comprehensive evaluation, including clinical presentation, imaging features, and histopathological examination, is often necessary to make an accurate diagnosis and guide appropriate treatment for these tumors. The management of craniopharyngiomas is challenging because of their location, invasiveness, and proximity to adjacent neurovascular structures (pituitary, hypothalamus, optic chiasm, circle of Willis, third ventricle). This typically requires a multidisciplinary approach and should be individualized for each patient.

Ethics

Informed Consent: Informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.K., A.A., N.A., Concept: A.K., A.A., N.A., Design: A.K., A.A., N.A., Data Collection or Processing: A.K., A.A., N.A., Analysis or Interpretation: A.K., A.A., N.A., Literature Search: A.K., A.A., N.A., Writing: A.K., A.A., N.A.

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