

Case Report

MRI Findings of Infundibular Craniopharyngioma: Two Case Reports

İnfundibular Kraniofaringiomanın MR Görüntüleme Bulguları:
İki Olgu SunumuÖzkan Ünal¹, Salih Çırak¹, Ercan Bal²¹Ankara Dr Abdurrahman Yurtaslan Oncology Training and Research Hospital,
Radiology Department, Ankara, Turkey²Ankara City Hospital, Department of Neurosurgery, Ankara, Turkey

ABSTRACT

Primary infundibular craniopharyngioma is a relatively unusual disease due to its location, which usually results in late diagnosis. Two female patients are referred to the radiology clinic because of bitemporal hemianopsia detected in visual assessment at different times.

Infundibular lesions were detected with MRI both patients and craniopharyngioma was considered as the primary diagnosis. Both masses are operated and the diagnosis of craniopharyngioma is proven pathologically.

Keywords: MRI, infundibular Craniopharyngioma, Pituitary infundibulum

ÖZET

Primer infundibular kraniofaringiyoma, konumu nedeniyle nispeten nadir görülen ve genellikle geç tanı alan bir hastalıktır. Farklı zamanlarda norolojik muayenelerinde bitemporal hemianopsi tespit edilen iki hasta MRG incelemesi için radyoloji kliniğine refere edildi. Her iki hastada da MRG ile infundibüler yerleşimli lezyonlar tespit edildi ve kraniofaringiyom birincil tanı olarak düşünüldü. Her iki kitle de ameliyat edildi ve kraniofaringiyom tanısı patolojik olarak doğrulandı.

Anahtar Kelimeler: MRG, İnfundibular Kraniofaringioma, Hipofizer İnfundibulum

Introduction

Craniopharyngiomas account for 1–4% of all intracranial tumours and 20% of the tumours of the sellar and chiasmatic region [1]. The tumor has two age peaks, the one occurring in children and the other one in adults between the 4th and 6th decades despite it's more common in childhood than adulthood period. Craniopharyngiomas arise from remnants of embryonic canal extending from the oropharynx to the median eminence and infundibulum. According to this theory, any place along this canal may serve as a site of tumor origin [2]. Although the epicenter of the lesions is usually suprasellar (90%), sellar or infrasellar region; anterior, middle and

posterior cranial fossas, retroclival region, sphenoid bone, nasopharynx, cerebello-pontine angle and even pineal gland are other rare sites of the tumor development [3]. Infundibulum along the embryonic canal is another potential site for tumor origin. However, primary infundibular craniopharyngioma is a rare disease and uncommonly early diagnosed until it grows towards the suprasellar or parasellar regions. It usually presents with neurological complications like headache, visual disturbance and symptoms related with hypothalamic or hypophyseal gland dysfunction [4].

Here we report unusual infundibular cranio-

pharyngioma cases which is pathologically confirmed and primarily arising from the infundibulum. We want to discuss imaging findings and differential diagnosis of infundibular craniopharyngioma.

Case Report

Case 1: A 32-year-old female patient has been admitted to the neurology outpatient clinic with complaints of persistent vision problems and headache for one month. Bitemporal hemianopsia is detected in the visual examination.

CT showed a hypodense mass with punctate peripheral calcifications in the suprasellar area. MRI revealed that the mass was originating from the pituitary infundibulum and containing cystic and solid areas, approximately 20x20 mm in size. The lesion compresses the optic chiasm posteriorly and is closely adjacent to vascular structures. The pituitary gland had a normal appearance. Based on these findings, radiological diagnosis was considered to be infundibular craniopharyngioma (Fig 1a-c). Infundibular craniopharyngioma was confirmed histopathologically. On the 2nd year follow-up MRI, there was a mass lesion measuring 15 x 10 mm consistent with residual or recurrent tumor (Fig 1d).

Case 2: A 61-year-old woman suffering from visual disturbance for 2 months admitted to the hospital. Visual examination of the patient revealed bitemporal hemianopsia. An infundibular mass was demonstrated as intermediate signal intensity on T1-weighted MR images and high signal intensity similar to CSF corresponding to cystic changes on T2-weighted images. It showed peripheral and nodular enhancement pattern on postcontrast T1-weighted images. No restriction of diffusion is noted on diffusion weighted images. The tumor was extending from hypophysial infundibulum to hypothalamus

and compressing optic chiasm. (Figure 2 a-c) The initial radiological diagnosis of the lesion was craniopharyngioma, and then the lesion was decided to be operated on. Histopathologic results confirmed infundibular craniopharyngioma. Follow-up MRI images two months after the operation showed no recurrence or residual tumor.

Discussion

Craniopharyngiomas are histologically benign tumors originating from squamous epithelial cells of Rathke's cleft. They usually involve sellar-suprasellar region and invade or extend to clinically important structures as optic chiasm and hypothalamus. It's more common in childhood than adulthood period. However our two patients were adult. Although craniopharyngiomas usually arise from Rathke's cleft, they can also arise from embryonic cells located anywhere along the craniopharyngeal canal. [5].

Infundibular involvement may be seen in many conditions as well as in craniopharyngiomas. It's important to make a differential diagnosis of infundibular diseases in order to treat appropriately. Infundibular lesions are generally classified into three categories as neoplastic, inflammatory-infectious and congenital-developmental. Neoplastic lesions include astrocytoma, ependymoma, germinoma, pleomorphic xanthoastrocytoma, meningioma, prolactinoma, metastasis and craniopharyngioma. The first diagnosis to be considered in the absence of primary tumor in isolated stalk masses is craniopharyngioma. The size of infundibulum usually should be lesser than basilar artery at clivus level. More than this size might be a remarkable point for infundibular lesions. In many cases, hypothalamus is involved together with the infundibulum. The lesions involving both pituitary stalk and hypothalamus may result in diabetes insipidus.

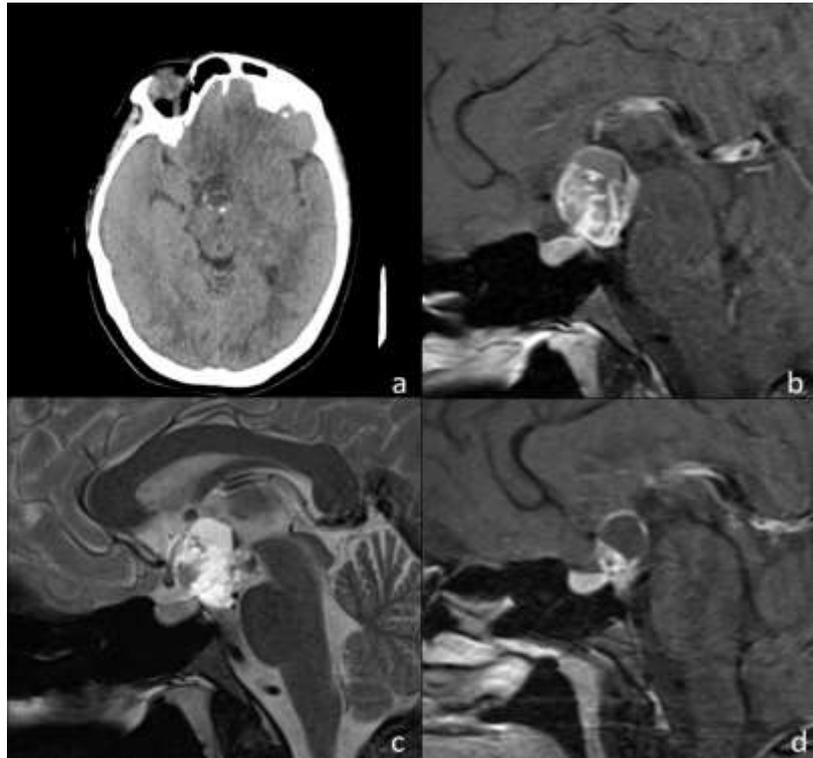


Figure 1(a-d): Non-contrast CT shows a mass with peripheral punctate calcifications in the suprasellar area.

(a). Solid lesion of stalk with cystic-necrotic component is shown on contrast enhanced T1-weighted sagittal image (b) and T2-weighted sagittal image (c). The mass compresses the optic chiasm from the posterior and causes visual symptoms (c). The remaining mass is demonstrated in the postop contrast-enhanced MR image taken two years after surgery (d).

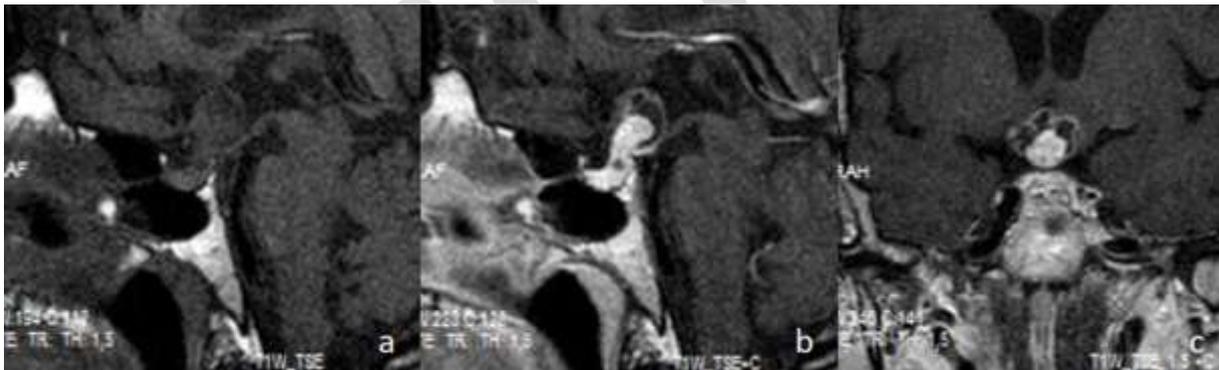


Figure 2(a-c). Sagittal non-contrast T1-weighted MRI shows heterogeneous suprasellar mass lesion (a). Contrast-enhanced solid lesion of stalk with superior cystic-necrotic component demonstrates on contrast enhanced T1-weighted sagittal (b) and coronal (c) images.

Pituitary adenomas may cause other endocrinologic problems due to excessive secretion of hormones. Germinomas which usually present as a tumor of pineal gland or hypothalamus, can also be seen in primarily infundibulum [6]. Metastasis to infundibulum are usually from breast and lung cancers. In the presence of primary tumor, metastasis can be considered primarily, but neither of our patients had primary tumors. Leukaemia,

lymphoma and atypical/malignant meningiomas which are aggressive tumors of hypothalamic-suprasellar region are other rare tumors can involve infundibulum [1]. Rathke's cleft cysts should be taken into account if there's a cystic tumor of infundibulum. All of these tumors are in the differential diagnosis of infundibular craniopharyngiomas. However, there are some granulomatous diseases of the infundibulum that may

resemble neoplasms. Sarcoidosis, tuberculosis, langerhans cell histiocytosis are such kind of granulomatous diseases that involve infundibular stalk. Therefore, MRI is an important imaging method in the differential diagnosis of infundibular lesions.

In our cases, infundibular involvement is due to a true neoplastic pathology called infundibular craniopharyngioma. Visual impairment is one of the major and earliest presenting symptoms of patients harboring craniopharyngiomas and also a potential complication of the surgical treatment [7]. The first symptom in our patients was visual problems. Also it is usually diagnosed late unless complains of headache, polyuria and polydipsia or complains due to compression of optic chiasm consist. After diagnosis of craniopharyngioma, the next step is to choose appropriate treatment modality. There is a

therapeutic dilemma for craniopharyngioma, so the approach is important when deciding on conservative or aggressive treatment. Conservative treatment may be an alternative management for early diagnosed small tumors especially for the tumors which are not extending to optic chiasm. However, there is no consensus on treatment options of larger lesions extending to optic chiasm. Complete surgical resection is usually indicated because it's believed that craniopharyngiomas are curable tumors. It's important to conserve infundibulum but it's sometimes sacrificed for total resection. Another alternative treatment modality for some authors is limited surgery followed by radiotherapy [8,9]. Surgical treatment was preferred in both of our patients. While total resection was achieved in our second case, our first case is being followed due to a residual mass.

REFERENCES

1. Figueiredo EG, Welling LC, de Faria JW, Teixeira MJ. Pituitary stalk craniopharyngioma. *BMJ Case Rep.* 2011; bcr0620103097.
2. Songtao Qi, Yuntao Lu, Jun Pan, Xi'an Zhang, Hao Long, and Jun Fan. Anatomic relations of the arachnoidea around the pituitary stalk: relevance for surgical removal of craniopharyngiomas. *Acta Neurochir (Wien).* 2011; 153(4): 785–796.
3. Sipos L, Vajda J. Craniopharyngioma of the third ventricle. *Acta Neurochir (Wien)* 1997; 139: 92–3.
4. Pusey E, Kortman KE, Flannigan BD, Tsuruda J, Bradley WG. MR of craniopharyngiomas: tumor delineation and characterization. *AJR Am J Roentgenol.* 1987; 149 (2): 383-8.
5. Mehta V, Black PM. Craniopharyngioma in the adult. In: Winn RH, ed. *Youmans Neurological Surgery.* Fifth edition Philadelphia, PA: Saunders Elsevier; 2004:1207–21.
6. Rupp D, Molitch M. Pituitary stalk lesions. *Curr Opin Endocrinol Diabetes Obes* 2008; 15: 339–45.
7. Prieto R, Pascual JM, Barrios L. Optic chiasm distortions caused by craniopharyngiomas: clinical and magnetic resonance imaging correlation and influence on visual outcome. *World Neurosurg* 2015; 83:500 –29.
8. Nishizawa S, Ohta S, Oki Y. Spontaneous resolution of diabetes insipidus after pituitary stalk sectioning during surgery for large craniopharyngiomas. *Neurol Med Chir (Tokyo)* 2006; 461: 126–135.
9. Rahmathulla G, Barnett GH. Minimally invasive management of adult craniopharyngiomas: An analysis of our series and review of literature. *Surg Neurol Int.* 2013; 20; 411-21..

Corresponding author e-mail: drozkanunal@yahoo.com

Orcid ID:

Özkan Ünal 0000-0001-7627-9317

Salih Çırak 0000-0001-7053-9130

Ercan Bal 0000-0003-0138-2090

Doi:

proof