International Journal of Advances in Medicine
Dhanju AS et al. Int J Adv Med. 2023 Dec;10(12):866-868
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20233575

Case Report

Craniopharyngioma in adults: a case report

Avtar Singh Dhanju, Nisha Narang, Manavdeep Kaur*, Rishabh Rikhye, Pankaj Jassal, Adab Alwinder Singh

Department of Medicine, Sri Guru Ram Das University of Health Sciences, Sri Amritsar, Punjab, India

Received: 24 October 2023
Revised: 14 November 2023
Accepted: 18 November 2023

***Correspondence:**
Dr. Manavdeep Kaur,
E-mail: mdeep2k7@gmail.com

ABSTRACT

We are discussing a case report of 70-year-old male patient with craniopharyngioma who presented with complaint of high grade fever and altered sensorium with presence of meningeal irritation signs and papilledema in fundus examination. MRI 3D brain showed elongated lobulated mass lesion in the suprasellar region. It mostly occurs in young age before 20 years of age with features of headache, projectile vomiting and signs of raised intracranial pressure but there is possibility of its occurrence in older age group. It arises from embryologic squamous epithelial remnants of the craniopharyngeal duct or Rathke's pouch. It develops near the hypothalamus near the pituitary gland that controls growth and many body functions. Usually, they are benign but can be malignant sometimes as they can cause serious problems by interfering with neuroendocrine structures or neuropsychological complications. There are only few cases of craniopharyngioma in old age patients worldwide.

Keywords: Craniopharyngioma, Rathke's pouch, Neuropsychological complications

INTRODUCTION

Craniopharyngiomas (CPs) are benign, suprasellar cystic masses which are relatively rare, accounting to only 3% of all intracranial tumours.[1] It arises from embryologic squamous epithelial remnants of the craniopharyngeal duct or Rathke's pouch.[2] It develops near the hypothalamus near the pituitary gland that controls growth and many body functions.[3] Usually, they are benign but can be malignant sometimes as they can cause serious problems by interfering with neuroendocrine structures or can cause neuropsychological complications.[4,5] Mostly patients present before 20 years of age but sometimes elder patients may also present with craniopharngioma.[6] Here we reported a case of 70 year male with suprasellar craniopharyngioma.

CASE REPORT

A 70-year-old male, presented with complaint of fever from four days associated with chills and rigors along with severe headache. Fever was high grade documented maximum upto 103 °F, it was associated with projectile vomiting. After four days patient had altered sensorium along with irrelevant talks and was drowsy and lethargic. On examination, patient was thin, lean and not oriented to time, place and person. The power in all muscle groups was 4/5 with deep tendon reflex 2+ and bilateral plantar were extensor. Patient had positive Kernig's sign, Brudzinski sign and neck rigidity was also present. Patient was expected to be suffering from viral encephalitis. On fundus examination-papilledema was present and MRI 3D brain was done which showed an elongated lobulated mass lesion in the suprasellar region, measuring approximately 2.7×2.3×1.4 cm in size. It appeared isointense on T1W images and hyperintense on T2W and FLAIR images and most of the part of suprasellar cisterns and showed hypointense area in it. On post contrast images, the lesion showed significant enhancement likely to be craniopharyngioma. Patient managed on the lines of viral encephalitis and improved. Detailed examination of the patient was done again after 4 days of admission. Patient

gave occasional history of headache but no history of vomiting, or cognitive impairment or weight gain or any vision loss. On examination visual acuity was normal and all other cranial nerve were normal. On motor examination all DTR were 2+ and bilateral plantar were flexor. Sensory examination was normal. On lab investigation patient's haemoglobin was 6.2 gm% with total leucocyte count 4100 cumm with platelet count of 2.03 lakh. Serum sodium was 110 mmol/l, serum potassium was 3.7 mmol/l. His serum prolactin was 25.40 ng/ml which was normal. Patient was referred to higher centre for transcranial or transsphenoidal resection of the tumour. Figure 1 (a and b) shows suprasellar enhancing lobulated mass lesions.

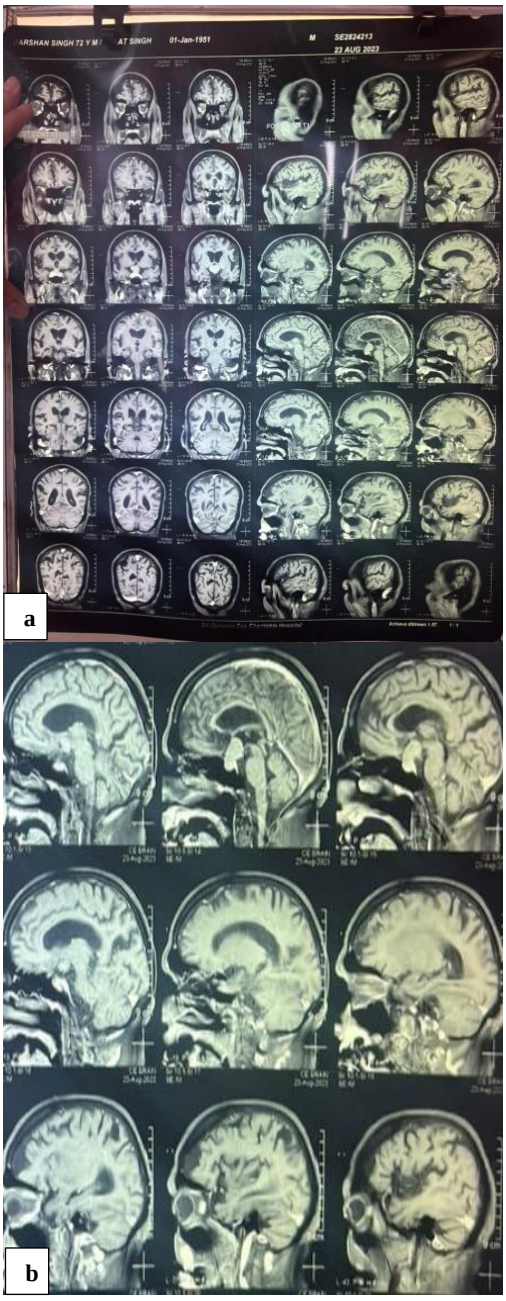

Figure 1 (a and b): Suprasellar enhancing lobulated mass lesions.

DISCUSSION

CPs are benign, suprasellar cystic mass lesion most commonly presenting before 20 year of age but rarely present in elder age. They are derived from squamous epithelial remnants Rathke's pouch and arise near the pituitary stalk extending into suprasellar cistern.[6] This pouch appears during the fourth gestational week. It transforms into a canal that obliterate itself and disappears by resorption during the seventh gestational week. The resorption begins at the middle of the canal.[7] This explains the presence of craniopharyngioma in the suprasellar area.[8] They are often large, cystic and locally invasive. These are made up of a mixture of calcium deposits, and cysts containing protein, lipids, and cholesterol pockets. There are two main types of CPs, adamantinomatous CP (ACP) which is more common in children, and papillary CP (PCP) which occur in adults.[9,10] Papillary CP are associated with activated BRAF V600E mutations. Adamantinomatous CPs are known to have β-catenin gene (CTNNB1) mutations.[11] Patient with craniopharyngiomas have symptoms of increased intracranial pressure such as headache, vomiting, papilledema and hydrocephalus along with visual field defects, cognitive impairment, changes in personality, cranial nerve damage, sleep difficulties and weight gain along with metabolic syndrome. Most of the children present with growth retardation and 90% of the patient had hypopituitarism and 10% had diabetes insipidus. MRI is diagnostic modality for diagnosis of CPs while CT scan is used to define calcification or evaluating the invasion into surrounding structures and sinuses.[6]

The main stay of treatment of CP is surgical resection followed by postoperative radiation of residual tumour. The surgical resection was done via transcranial or transsphenoidal method. As recurrence of tumour can occur because of adherence of tumour to vital structures or because of small tumour deposit in the hypothalamus or brain parenchyma radiotherapy was done. Without radiotherapy approximately 75% of the tumour reoccur. In patient with incomplete resection, radiotherapy improves 10-year survival to 70-90% but may be associated with secondary malignancies. Lifelong pituitary hormone replacement can be required in some patients. Patients with BRAF V600E mutations, use of BRAF inhibitors (dabrafenib or vemurafenib) either alone or in combination with MEK inhibitors (trametinib or cobimetinib) showed good results.[6]

CONCLUSION

As the CP is the tumour most commonly present in young age group, but it can be present in older age as well. The patient can present with atypical symptoms like high grade fever with altered sensorium along with signs of meningeal irritation but still MRI brain should be done to rule out the possibility of craniopharyngioma and patient should be thoroughly examined for visual field defects as it can compress on optic chiasma. MRI brain is an important diagnostic modality for diagnosing craniopharyngioma.

There are only few published case reports of the patients presenting with craniopharyngioma in old age.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Jane JA, Laws ER. Craniopharyngioma. Pituitary. 2006;9(4):323-6.
2. Cai X, Sun Z, Li Y, Shao D, Zheng X, Wang Y, et al. Primary ectopic parasellar craniopharyngioma: a case report. BMC Neurol. 2021;21:1-5.
3. Jastania RA, Saeed M, Al-Khalidi H, AlQuthami K, Nageeti TH, Al-Allaf FA, et al. Adamantinomatous craniopharyngioma in an adult: a case report with NGS analysis. Int Med Case Rep J. 2020:123-37.
4. Sterkenburg AS, Hoffmann A, Gebhardt U, Warmuth-Metz M, Daubenbuchel AM, Muller HL. Survival, hypothalamic obesity, and neuropsychological/psychosocial status after childhood-onset craniopharyngioma: newly reported long-term outcomes. Neuro Oncol. 2015;17(7):1029-38.
5. Feng Y, Ni M, Wang YG, Zhong LY. Comparison of neuroendocrine dysfunction in patients with adamantinomatous and papillary craniopharyngiomas. Exp Ther Med. 2019;17(1):51-6.
6. Kasper D, Fauci A, Hauser S, Longo D, Jameson J, Loscalzo J. Harrison's principles of internal medicine. 19th ed. New York, NY, USA: Mcgraw-Hill; 2015.
7. Goldberg GM, Esbaugh DE. Squamous cell nests of the pituitary gland as related to the origin of craniopharyngiomas. Arch Pathol. 1960;70:293-9.
8. Sangiovanni G, Tancioni F, Tartara F, Gaetani P, Rindi G, Passeri F, et al. Ectopic craniopharyngioma: presentation of a case arising from the corpus callosum. Acta Neurochir (Wien). 1997;139:379-80.
9. Muller HL. Craniopharyngioma. Endocr Rev. 2014;35(3):513-43.
10. Muller HL. Childhood craniopharyngioma–current concepts in diagnosis, therapy and follow-up. Nat Rev Endocrinol. 2010;6(11):609-18.
Sekine S, Shibata T, Kokubu A, Morishita Y, Noguchi M, Nakanishi Y, et al. Craniopharyngiomas of adamantinomatous type harbor beta-catenin gene mutations. Am J Pathol. 2002;161:1997-2001.

Cite this article as: Dhanju AS, Narang N, Kaur M, Rikhye R, Jassal P, Singh AA. Craniopharyngioma in adults: a case report. Int J Adv Med 2023;10:866-8.