CASE REPORT

CRANIOPHARYNGIOMA PRESENTING WITH BILATERAL OPTIC ATROPHY

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ABSTRACT

A young pre-pubertal female patient presented to the medical department with bilateral marked loss of vision. She also complained of facial and bipedal edema in the near past though other features of hypothyroidism were lacking in her history. She had a strong history of mental disorders in the family. General physical examination was unremarkable. Abdominal examination revealed hepatomegaly. Central nervous system examination showed bilateral optic atrophy and an up-going left plantar response. Baseline investigations were normal. Thyroid functions suggested sub-clinical hypothyroidism. MRI scan of the brain and orbits revealed a locally invasive cystic lesion with solid components, encasing the major structures consistent with craniopharyngioma. Partial excision of the lesion was carried out, and biopsy report confirmed it to be an adamantinoma variety of craniopharyngioma.

Key words: Optic atrophy, hypothyroidism, craniopharyngioma, adamantinoma.

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INTRODUCTION

Craniopharyngiomas are embryonic tumors that are benign on histopathology but behave aggressively like malignant tumors [1]. They most commonly arise from the pituitary stalk and project into the hypothalamus [2]. They extend in all directions along the path of least resistance. They can even extend extra-cranially as far as the cervical spine [3]. Craniopharyngiomas usually present as cystic lesions with gelatinous material that shines on exposure to light due to presence of cholesterol crystals [4]. Surgical resection is the usual treatment but recurrence after complete removal can occur 5. We hereby report a case of craniopharyngioma in a young patient presenting with bilateral optic atrophy.

CASE REPORT

A seven year old girl presented with a three months history of right hypochondrium pain and bilateral marked reduction in vision. She also had a history of frequent falls and was unable to walk independently; most likely attributable to her visual problem. She had suffered from bipedal edema and facial puffiness in the recent past. She was somnolent most of the times and had lost interest in her daily activities. There was no complaint of constipation, cold intolerance or weight gain. She was pre-pubertal. She had a family history of mental disorders and epilepsy in her siblings. One of the siblings was epileptic; another elder sister was also having some manic disorder, and one other elder sister had passed away a few years back; she was also reported to be having some mental disorder. The child was born to a non-consanguineous marriage.

On examination, she had a normal blood pressure and pulse. She was pale, but rest of the general physical examination was unremarkable, and there was no thyroid swelling. Her abdominal examination revealed hepatomegaly palpable up to two finger breadths below right hypochondrium. Central nervous system examination showed a left up-going plantar response. Power, reflexes and sensations were intact. Both the pupils were dilated, and the right pupil had very sluggish response to light. She had horizontal nystagmus. Visual perception of hand movements was there but patient could not count fingers. Fundoscopic examination of eyes revealed bilateral optic atrophy. Gower sign was also positive.

Her baseline investigations showed a hemoglobin of 11.2 gm/dl, White cell count = 5,900/cmm with a normal differential leukocyte count, and platelet count = 250,000/cmm. Her serum electrolytes were normal. Creatinine phosphokinase (CPK) measured 95 unit/liter (24-175 unit/liter). Random blood sugar was 60mg/dl. Renal profile was not deranged. Liver profile showed alanine transaminase = 82unit/liter (10-40unit/liter), serum bilirubin = 0.33mg/dl (0.1-1.5mg/dl) and alkaline phosphatase = 267unit/liter (child upto 625unit/liter). Serum calcium = 10.4 unit/liter (8-10 unit/liter). Thyroid function tests were suggestive of a sub-clinical hypothyroid state with TSH =

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11.4unit/liter (5-8unit/liter), T3 = 5.2 unit/liter (3-7 unit/liter), and T4 = 4.2 unit/liter (4-7unit/liter). An MRI brain and orbits was planned.

The MRI (Figures: 1 and 2) showed a large multi-septate predominantly cystic lesion measuring 4.5x4.1x3.5cm involving sella, supra-sellar cistern and extending into inter-peduncular cistern and right ambient cistern. The lesion had thick irregular septations and eccentric solid components which showed avid enhancement on post-contrast images. The lesion was closely applied to bilateral supra-clinoid parts of internal carotid artery and expanding the sella. The cystic chiasma was encased and distorted by the disease. Multiple small rounded cystic satellite nodules were seen in the left basal ganglia and right mid-brain. Disease was also closely applied to floor of 3rd ventricle and having mass effect on brainstem. In light of the MR findings, it was concluded that it is a locally inoperable neoplastic lesion, consistent with craniopharyngioma.

The patient was further referred to neurosurgery department, where she was operated upon, with partial excision of the mass. It could not be excised in total because of close approximation to vital structures, but to reduce its compressive symptoms, partial excision was carried out. The biopsy report returned confirming Adamantinoma type of craniopharyngioma, with palisades of small cells visible in the biopsy specimen, enclosing a loose, reticular zone and squamous cells. Few nodules of keratin were also seen in the specimen (Figure 3).

The patient was thus diagnosed as a case of craniopharyngioma of the Adamantinoma variety, and referred for radiotherapy after partial removal of the tumor mass. We hereby present her case to shed light on atypical presentation of craniopharyngioma with bilateral optic atrophy.

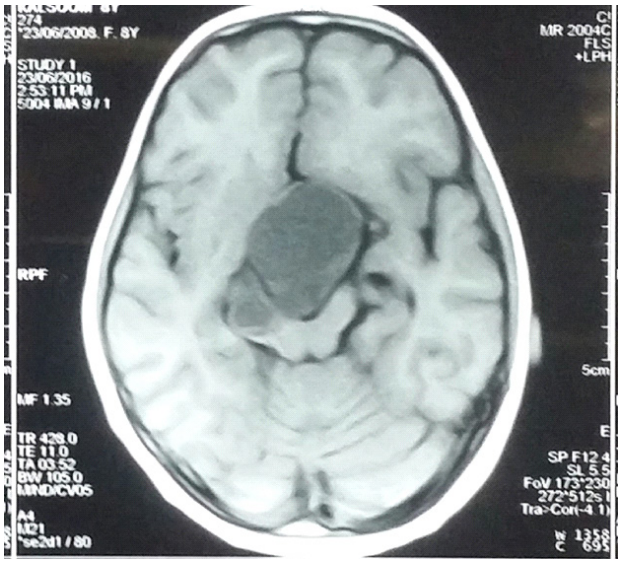

Fig 1: MRI Brain showing large craniopharyngioma

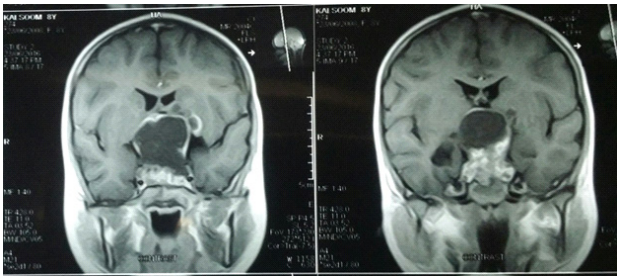

Fig 2: MRI Brain showing coronal view of craniopharyngioma

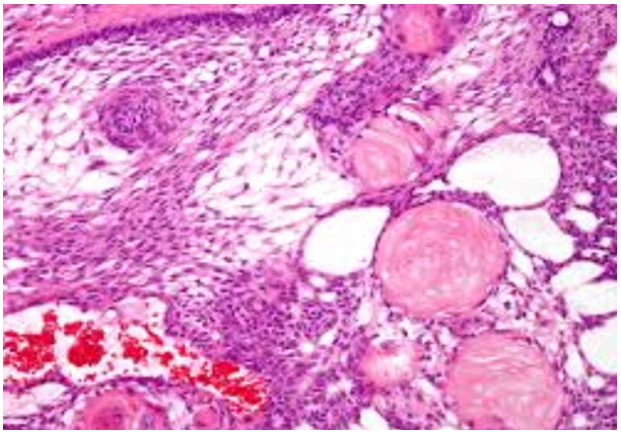

Fig 3: Histopathology of craniopharyngioma (magnification x100)

DISCUSSION

Craniopharyngiomas are slow-growing tumors that lead to symptoms when they grow beyond 3cm 6. They most commonly present with headaches, endocrine dysfunction and visual disturbances [7]. Our patient did not complain of headache but was having sub-clinical hypothyroidism and bilateral optic atrophy. Dysfunction of optic pathways occurs in 40-70% of the patients. Children become aware of visual problems only after almost complete damage to visual pathways. At this stage the loss is irreversible [8]. Craniopharyngioma is the most common cause of bilateral optic atrophy in population under 20 years of age [9].

Greater than 50% of the children with craniopharyngioma in one study presented with clinical features of raised intracranial pressure. However, one-third of these patients had optic atrophy on visual examination, instead of papilledema [10]. Our patient did not have raised intra-cranial pressure but had bilateral optic atrophy because of extension into and invasion of the visual pathway by the craniopharyngioma.

There are multiple non-surgical and surgical treatments available for the management of craniopharyngiomas. Bleomycin, radiotherapy, intracystic chemotherapy, cytokines, biomodulation and gross total or partial resection are the different approaches for management of craniopharyngioma. Total resection still has chances of

recurrence. Partial resection needs to be followed by radiotherapy to reduce risk of re-expansion of tumor mass [11, 12].

REFERENCES

1. Stamm AC, Vellutini E, Balsalobre L. Craniopharyngioma. Otolaryngol Clin North Am. Aug 2011;44(4):937-52, viii.
2. Müller HL. Craniopharyngioma. Handb Clin Neurol. 2014;124:235-53.
3. Jung TY, Jung S, Jang WY, Moon KS, Kim IY, Kang SS. Operative outcomes and adjuvant treatment of purely third ventricle craniopharyngioma after a transcallosal approach. Br J Neurosurg. 2012; 26(3):355-60.
4. Bunin GR, Surawicz TS, Witman PA, et al. The descriptive epidemiology of craniopharyngioma. J Neurosurg. Oct 1998; 89(4):547-51.
5. Fraioli MF, Moschettoni L, Catena E, Fraioli C. Cystic craniopharyngioma: trans-sphenoidal surgery and intra-cystic apposition of “bleomycin wax”. Acta Neurochir (Wien). 2010; 152(2):293-6.
6. Pereira AM, Schmid EM, Schutte PJ, et al. High prevalence of long-term cardiovascular, neurological and psychosocial morbidity after treatment for craniopharyngioma. Clin Endocrinol (Oxf). Feb 2005;62(2):197-204.
7. Waber DP, Pomeroy SL, Chiverton AM, et al. Everyday cognitive function after craniopharyngioma in childhood. Pediatr Neurol. Jan 2006;34(1):13-9.
8. Meuric S, Brauner R, Trivin C, et al. Influence of tumor location on the presentation and evolution of craniopharyngiomas. J Neurosurg. Nov 2005; 103(5 Suppl):421-6.
9. Menon V, Arya AV, Sharma P. An aetiological profile of optic atrophy. Acta Ophthalmol. 1992;70(6):725-29.
10. Kennedy HB, Smith RJ. Eye signs in craniopharyngioma. Br J Ophthalmology. 1975; 59(12):689-95.
11. Julow J, Lanyi F, Hajda M, Szeifert GT, Viola A, Balint K, et al. Stereotactic intracavitary irradiation of cystic craniopharyngiomas with yttrium-90 isotope. Prog Neurol Surg. 2007; 20:289-96.
12. Hukin J, Steinbok P, Lafay-Cousin L, Hendson G, Strother D, Mercier C, et al. Intracystic bleomycin therapy for craniopharyngioma in children: the Canadian experience. Cancer. May 15 2007;109(10):2124-31.