PILOCYTIC ASTROCYTOMA PRESENTING AS BILATERAL OPTIC ATROPHY IN A CHILD: CASE REPORT

Sucheta Parija[1,*], Indranil Saha[2]

[1]Assistant Professor, [2]Post Graduate Resident, Department of Ophthalmology, AIIMS, Bhubaneshwar, India

***Corresponding Author:**
E-mail: suchetaparija@yahoo.com

ABSTRACT:

Brain tumors are a very common pathology in children today. Optic atrophy is an alerting sign for ophthalmologist that may be associated with serious systemic conditions having a significant bearing on the overall morbidity of the child .We describe here a 14 years old girl who presented with visual loss and features of raised intracranial pressure. On imaging she was diagnosed to have a supratentorial tumor suggestive of central neurocytoma but on histopathology was reported to be pilocytic astrocytoma gradeI.

Keywords: *Intracranial pressure, Optic atrophy, Pilocytic astrocytoma*.

INTRODUCTION

Brain tumors comprise approximately 20% of all childhood malignancies, second in frequency only to acute lymphoblastic leukemia. [1] Infratentorial brain tumors prevail in children (60%), but in infants and toddlers most of the tumors are supratentorial, like in adults. [2] Eyes are the windows to peek into the central nervous system. There may be different signs at various levels of alteration in the optic pathway due to the location and growth of the tumors leading to visual defects and total loss of vision. Optic nerve atrophy is not a disease, but rather a sign alerting the ophthalmologist of a potentially more serious condition. A thorough investigation of a child presenting with optic atrophy is, therefore, mandatory, and knowledge of the prevailing causes aids the clinician in an appropriate work-up and subsequent intervention. [3] Timely referral and intervention can not only save the life but also the sight. We report a case of bilateral optic atrophy as presenting feature due to pilocytic astrocytoma.

CASE REPORT

A 14 years old female presented to us with complaints of headache, multiple episodes of vomiting and bilateral diminution of vision for last 2 months. She had a past history of intermittent headaches and vomiting for last 9 months with episodes of clonic convulsions which had subsided on medication.

On examination her visual acuity was 4/60 in the right eye and hand movements in the left eye. The pupil was of normal size and reacting to light in the right eye with a RAPD in the left eye. Fundus examination showed bilateral disc pallor. There were no other cranial deficits. Motor and sensory functions were intact and there were no meningeal or cerebellar signs. Computed tomography [CT] scan (Fig:1) revealed a well defined, lobulated, variably vascularized mass with moderate to strong enhancement abutting the septum pellucidum, right thalamus, third ventricle, right parietal lobe with areas of cyst and necrosis. The globe and both optic nerve sheath complexes appeared normal. MRI brain showed supratentorial lobulated right lateral ventricle based complex space occupying lesion [SOL] (T1 hypo intensity andT2 hyper intensity) with mass effect causing non- communicating hydrocephalus (Fig:2 a & b). A diagnosis of supratentorial –right ventricular cystic tumor was made.

To prevent further detoriation, she was referred to neurosurgery and operated on emergency, where a right parietal craniotomy and cerebrotomy along with cystic fluid evacuation was done. A grayish, slimy, moderately vascular tumor was found. Near total decompression was done, dura closed and bones fixed. Squash cytology showed high grade glioma. The tissue (2.5x2.5x2 cm) was sent for histopathology, which was reported as pilocyctic astrocytoma [WHO grade I] (Fig-3a &b) - a well circumscribed mass, not infiltrating the surrounding tissue, composed of glial cells which were interwoven in a fine fibrillary background, rosenthal fibers, absence of atypical mitoses and the presence of a characteristic microcystic component .

The patient presented to our clinic after one month with further deterioration of vision in the left eye. There was a left exotropia with RAPD [Fig 4]. The ocular movements were full. The visual acuity was 1/60 in the right eye and presence of projection of light in the left eye. Both the fundi showed secondary optic atrophy [Fig 5a&b].

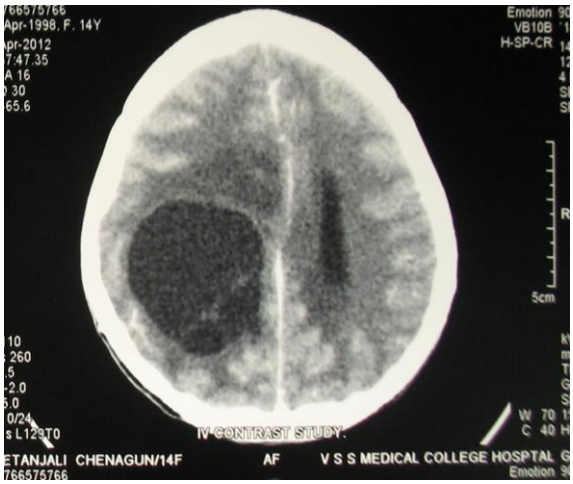
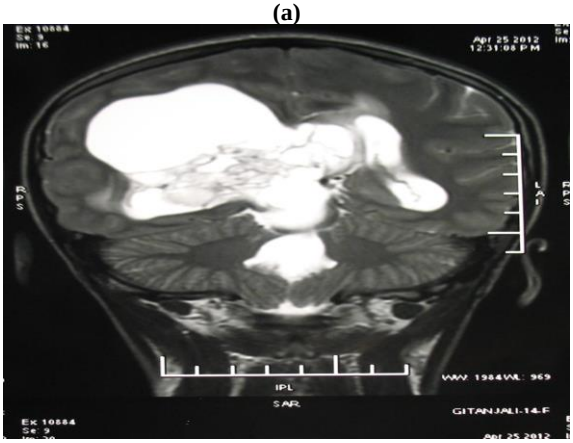


Fig. 1: CT scan of brain shows a well-defined lobulated, variably vascularized mass with moderate to strong enhancement abutting the septum pellucidum, third ventricle, right parietal lobe region with areas of cyst and necrosis.

(a)



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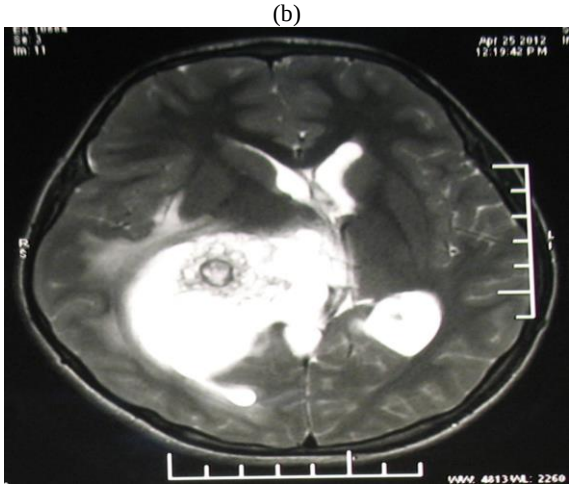


Fig. 2 (a & b): MRI shows supratentorial, lobulated solid and cystic SOL encroaching 3rd and both lateral ventricles .It also shows enhancement of some infiltrative component within its axis causing CSF outflow obstruction leading to non-communicating (obstructive) hydrocephalus

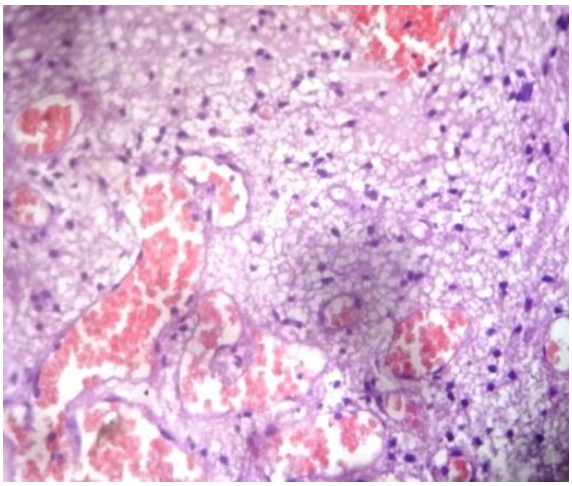


Fig. 3(a): Histopathology of tumour under (100X) showing bi-phasic appearance. Astrocytic cells are seen in a fibrillary background. In the hypercellular areas rosenthal fibres and eosiniphilic granular bodies are seen.

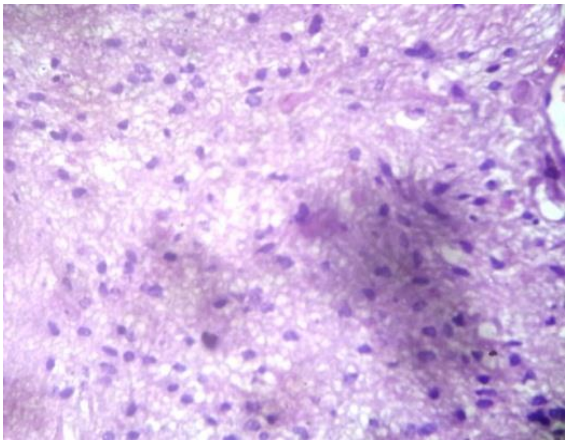


Fig. 3 b: Histopathological picture under 100 X shows microcystic spaces seen in the hypocellular area (H&E Stain).

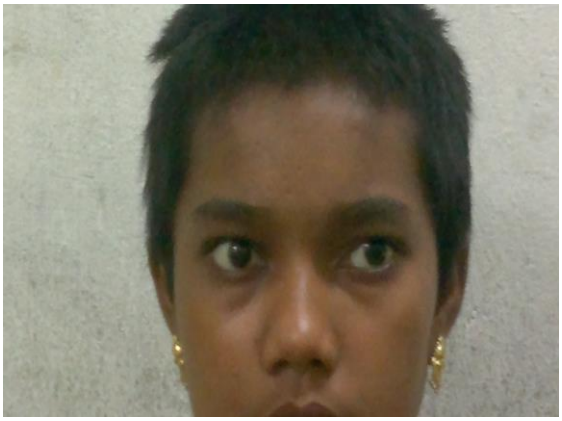
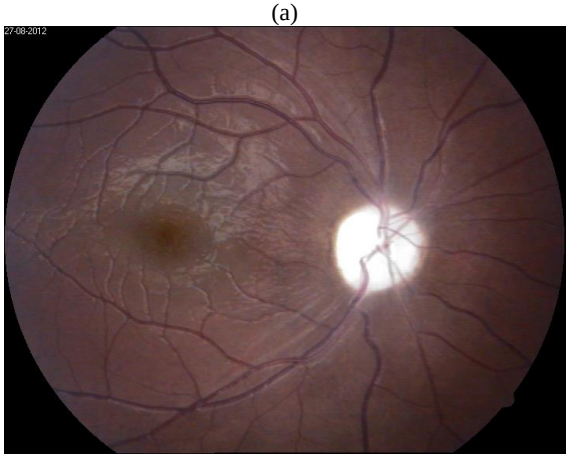


Fig. 4: Post-operative photo shows a left exotropia . (photograph is now trimmed for pts identification point of view) delete green sentence

(a)



(b)

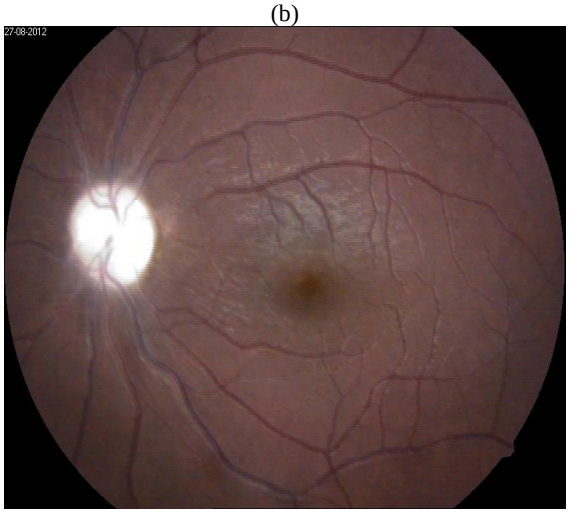


Fig. 5(a & b): Secondary optic atrophy seen in both eyes

DISCUSSION

Pilocytic astrocytoma (PA), previously known as spongioblastoma, is a histologic subtype of astrocytoma, classified as WHO Grade I, with a rather benign, slow-growing biological behavior [4]. This tumor is the most common solid neoplasms in the central nervous system in children and young adults [5]. These usually arise in the cerebellum, near the brainstem, hypothalamic region and optic chiasm and rarely in the cerebral hemisphere and spinal cord; like in our case were the right parietal lobe and surrounding structures involved. These tumors are predominantly benign with low cellularity, low proliferative and mitotic activity and rarely metastasize. Astrocytoma has neuroectodermal origins, from **astrocyte or neuroglia** and is classified in four categories depending on the grade of malignancy. Grade I astrocytoma is a benign tumor that predominantly arises in infratentorial locations such as the cerebellum and diencephalic region and rarely arises in supratentorial locations, in any hemispheric lobe, but especially in the frontal lobe. Our patient had a grade 1 astrocytoma in the supratentorial region in the right parietal lobe, which is a rarity.

As per the review of medical literature [5,7,8] ,75 % of patients present with signs due to raised intracranial pressure – headaches, vomiting and lethargy and 25 % of patients with supratentorial astrocytomas present with seizures [5,7], like in our patient. A 5 year survival rates of 95-100% are seen with surgical resection alone in these patients. Our patient too underwent a successful resection of the tumor after confirmation by neuroimaging.

Visual symptoms almost always accompany brain tumors. Many a time's tumors have been detected by fundus examination compared to imaging. Tumors lying in immediate vicinity of optic nerves and their chiasma or growing out from them are usually large and often their removal is associated with damage to the visual pathway leading to visual field defects to blindness. Papilledema is usually presented as one of the early signs of the disease, as a consequence of direct compression of the optic nerve and/or an increase of intracranial pressure, compromising ocular venous return and can be resolved after a ventricular-peritoneal derivation valve is implanted [6]. PA of the optic pathway frequently produces visual loss or visual-field deficit, with optic disc pallor and optic nerve atrophy in the involved eye secondary to axonal damage and ischemia [4], as seen in our case.

A study by Repka and Miller [9] reported 89% of cases of optic atrophy due to an intracranial space occupying lesion in contrast to the study by Chinta S et al.[3] who reported 5.6% of their cases to be of compressive aetiology. Among the compressive causes craniopharyngioma (n = 8, 44%) was the most common brain tumor. Five children were diagnosed after undergoing neuroimaging for optic atrophy and three cases presented after brain surgery.[3]

A study by Lee et al [10] described the diagnostic yield of neuroimaging and other laboratory investigations for optic atrophy. They could find a compressive cause in only 20% in their study, while 80% remained unexplained following neuroimaging. The authors felt that this warranted neuroimaging to rule out potentially serious associations in all patients of isolated optic atrophy.

CONCLUSION

Our patient was diagnosed with grade I pilocytic astrocytoma, a benign tumor, frequent in children, but in a rare location in this case. In a child, rapid rise of symptomology due to increased intracranial pressure, accompanied by rapid deterioration of the general condition and vision, should alert the clinician to investigate a possible intracranial tumor. CT scan is a non-invasive and rapid technique for the primary assessment of brain

tumor but definitive diagnosis is made through histo-pathologic examination.

A comprehensive ophthalmological examination is crucial in order to diagnose recurrence of brain tumours, where no other neurological signs are present, or no conclusions may be derived from image tests and hence our patient who underwent successful surgical resection of the tumor is on constant follow up for further care and detection of any tumor recurrence.

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