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Case report-Unusual Presentation of A Suprasellar Mass in A 4-Year-Old Child: A Bilateral Sudden Loss of Vision

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Abstract:

Paediatric suprasellar masses are considered unique in their clinical presentations and radiographic findings. Sellar and parasellar masses comprise about 10% of all paediatric brain tumour. We are attempting to describe an unusual presentation of a suprasellar mass in a 4-year-old child. Patient revealed sudden loss of vision for 7 hours with no history of pain and swelling, with past history of polydipsia and polyuria for 6 months. No relevant history of epilepsy, recent fever or raised intracranial pressure. On examination, visual acuity of both eyes was hand movement. Further, examination showed no abnormal findings with both eyes. Radiographic evaluation suggested presence of a large suprasellar mass with secondary metastasis to cranial-vault. The differential diagnosis on radiographical grounds include craniopharyngioma and hemangiopericytoma. Furthermore, there was hypopituitarism and central diabetes insipidus. This case-report aims to highlight the unusual presentation of suprasellar mass in paediatrics.

Keywords: Suprasellar mass; Loss of vision; Brain tumour; Paediatric ophthalmology.

1.INTRODUCTION:

The occurrence of brain tumour in childhood vary from those occurring in adulthood in their histological characteristics, in relation to incidence, its location, and response to therapy. With an age-standardised occurrence of 4 / 100.000, brain tumours are generally the second most familiar malignancies in children (1 , 2). Globally the survival rates of children with

brain tumour has increased as the treatment and diagnostic care have been improved (3, 4). The brain tumour survivors, on the other hand, are vulnerable to the disease's and treatment's long-term consequences (5). One of the extreme late effects of a childhood brain tumour is weakening of vision (6, 7). It has been documented that the disease's and/or treatment's negative effect on visual functioning has a significant impact on the development of general psychomotor, school attendance, and community participation later in life (7, 8). Both the efferent and afferent visual pathways may be affected by brain tumours, resulting in weakness of vision. Tumour location and elevated intracranial pressure (ICP) are important predictors of visual impairment (9). Compression of the visual pathway by the tumour might head to diminished visual acuity (VA), visual field (VF) loss and ocular motility shortfalls (6, 7).

Intracranial tumours may result in severe ocular symptoms as well as neurological problems such as increased intracranial pressure, cranial nerve dysfunction, or brain compression. As a result, early detection of intracranial tumours will lead to earlier care and the prevention of complications. The diagnosis of an intracranial tumour is usually made only in the presence of common signs or symptoms, which may lead to misreading and misdiagnosis (10). Gradual loss of vision with or without visual field weakness, deteriorate optic nerve and extraocular nerve palsies are common preliminary ocular symptoms (11). Children's brain tumours can cause vision loss, which varies depending on the location, form, and length of the tumour. Tumours of the posterior fossa make up about two-thirds of all brain tumours (12) are often connected to obstructive hydrocephalus and papilledema. The optic nerve is directly compressed by supratentorial tumours that occupy the suprasellar region (such as craniopharyngioma), but they can also cause hydrocephalus and papilledema (13).

The majority of paediatric sellar masses are craniopharyngiomas, and pituitary adenomas are extremely rare in children. A multidisciplinary approach is needed to diagnose sellar lesions, and thorough endocrinological, ophthalmological, and neurological testing is important in the evaluation of a new sellar mass (14). The most common childhood tumour affecting the hypothalamus and pituitary is craniopharyngioma. Even though endocrine abnormalities are observed in 80-90 percent of patients at presentation, only a small percentage of patients seek medical treatment for an endocrine-related issue (15).

2.CASE PRESENTATION:

A 4-year-old boy brought by his parents to the emergency department, hospital universiti sains Malaysia, with a complaint of sudden loss of vision of both eyes for 7 hours. Followed by the referral to the ophthalmology department for further investigations and management. The onset of symptoms was sudden, approximately 7 hours before their visit to the emergency. On history, the parents noticed that he is unable to see them, neither watch the television anymore, with loss of drinking and eating abilities by himself. Furthermore, parents noticed decrease in his activity and appetite followed by excessive water intake in the past 6 months. Prior to that, they claim that the vision was normal without any history of trauma to the eyes or head. Without, associated eye pain or redness, associated pain on eye movements, obvious ptosis or squinting, symptoms of raised intracranial pressure, fever or

seizures. Patient presented with the history of hospitalisation 1 month before for 4 days, due to high fever associated with gingivitis. Patient was discharged stable and given appointments to follow up regarding the dental issue.

On physical examination, the boy was conscious, alert, and cooperative yet frightened with stable vital signs. Further, on specific examination of the eyes, visual acuity of both eyes was hand movement, without relative afferent pupillary defect, nystagmus, proptosis, or ptosis. Extra ocular muscle movements of both eyes were grossly intact and no squint. Examination of the anterior and posterior segments of both eyes were normal. No signs of papilloedema and no abnormality detected in fundoscopy. Patient was referred to the department of haematology and radiology for further investigations.

2.1 Radiological and laboratory findings:

2.1.1 Non-contrast CT and Contrast-enhanced CT brain (figure 1):

a) A large suprasellar mass at the suprasellar region. The mass enhanced avidly with presence of cystic lesions within, no obvious calcification or bleed within the mass. Which was measured 2.6 x 2.8 x 1.8 cm. Normal pituitary stalk behind the mass with pituitary gland within the pituitary fossa. Perilesional oedema to the bilateral adjacent grey matter. Optic chiasm was not appreciated in this study.

b) A soft tissue enhancing mass causing erosion of the frontal bone at the level of the midline. This mass enters the intracranial cavity abutting the dural vessels. Bilateral frontal sinuses were absent. Which runs centrally and symmetrically from the hairline along the metopic suture to the superior orbital groove, erosions along these points of the bone to the anterior roof of the orbit and anterior ethmoid sinuses bilaterally.

c) Another soft tissue enhancing mass arising from the left temporal bone, which causes erosion of the left mastoid process, roof of the external auditory canal and left sphenoid bone wing. Intracranial extension of this mass directly apposes to the left temporal lobe. Inferiorly it extends to the left stylomastoid process encasing the left condyloid process of the mandible. However, there was no obvious filling defects within the cavernous sinuses and intracranial sinuses. No brain parenchymal enhancing lesions. Bilateral orbits are symmetrical and no evidence of intraconal mass or soft tissue infiltration.

Features of the CT likely to suggest aggressive suprasellar and metastatic lesions to be considered. A suggestion of tissue biopsy was raised for further investigations, yet the parents of the patient refused to undergo the craniotomy followed by the biopsy. Hence, advised to further follow up and revert.

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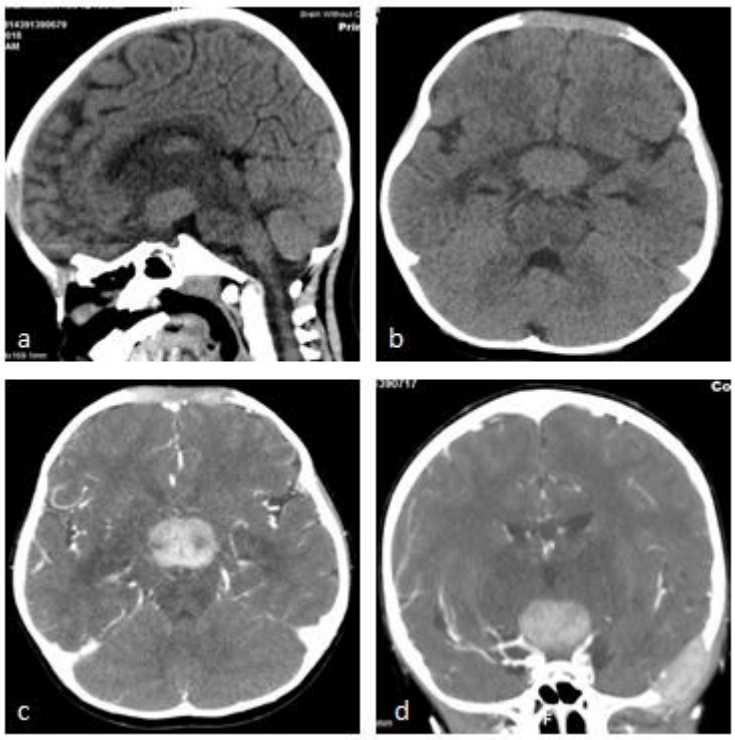

Figure 1: showing Non-contrast CT (a) sagittal, (b)axial and Contrast-enhanced CT brain (c) axial, (d)coronal views.

2.1.2 MRI Brain (figure 2):

The MRI brain of the patient revealed the following findings.

- A large suprasellar mass at the suprasellar region.
- The mass enhanced avidly with presence of cystic lesions within.
- No obvious calcification or bleed within the mass.
- It measured 2.0 x 2.2 x 2.0 cm.
- MRS shows high choline levels with a high choline-creatine ratio of 3.02 (normal <2). This indicates high cellular turnover
- Normal pituitary stalk with pituitary gland within the pituitary fossa
- Optic chiasm was not appreciated in this study.
- A soft tissue enhancing mass causing erosion of the frontal bone at the level of the midline.
- This mass appears to enhance off the frontal meninges with local bone erosions of the frontal bones. Skin layer overlying the leptomeningeal mass appears preserved.
- Soft tissue extension into the anterior ethmoidal sinuses is noted in this study.
- Another soft tissue enhancing mass arising from the left temporal bone.
- Presence of a dural tail with local erosion into the adjacent temporal bone. It causes erosion of the left mastoid process, roof of the external auditory canal and left sphenoid bone wing.
- MRS of this lesion demonstrated high lactate value, likely to suggest presence of necrotic tissue within.

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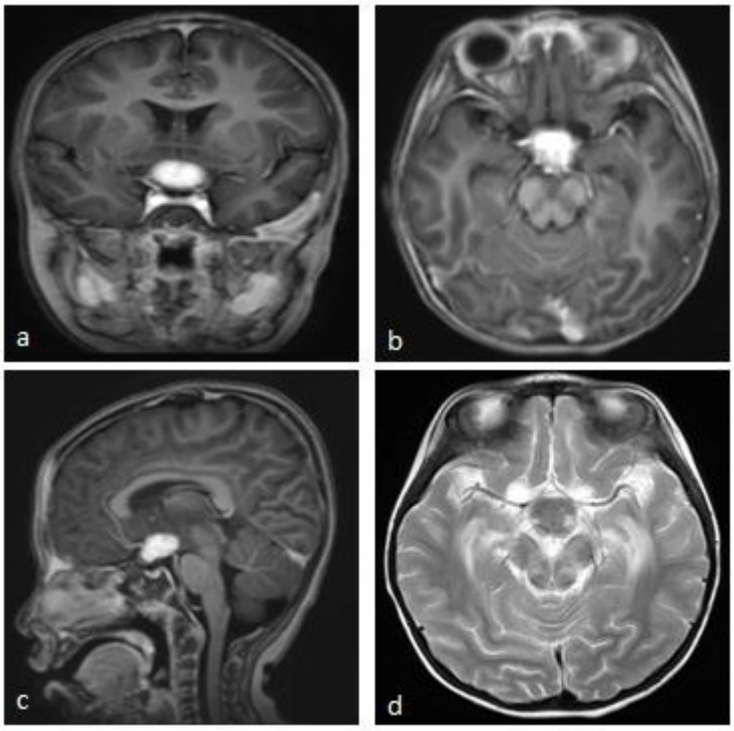

Figure 2: showing coronal, sagittal and axial views from MRI brain

2.1.3. Laboratory investigations:

The complete blood picture showed decreased levels of haemoglobin, mean cellular volume and haematocrit which is suggestive of hypochromic microcytic anaemia, further the increased levels of anti-diuretic hormone and serum osmolarity suggestive of diabetes insipidus. The hormonal investigation was suggestive of panhypopituitarism with low levels of thyroid stimulating hormone (TSH), prolactin, insulin-like growth factor-1, serum cortisol. The urine analysis was suggested diabetes insipidus with low urine osmolarity.

3. DISCUSSION:

Brain tumors as a group are meant to include all intracranial masses that arise from the brain parenchyma or other structures within the intracranial space. The 50 percentage of the brain tumor patients have the ophthalmic involvement, showing sings and or the symptoms [16]. The proper history with time and then course will give the clue to the examiner in relation to the tumors growth rate. The patients age is one of the important factor for certain type of the tumors to be likely than others, pediatric chiasmal syndrome is more commonly to be associated with craniopharyngioma in paediatric patients, whereas in pituitary adenoma is rarely seen [17]. The brain tumors shows typical ophthalmic signs and symptoms, Visual field defects, Exophthalmos, Optic disc changes (optic atrophy, papilloedema), Motility disorders (third, fourth and sixth cranial nerves palsies), Loss of somatic sensation (fifth cranial nerve palsy), Loss of colour vision (desaturation) and Loss of vision. The most common clinical presentation was decreased vision with disc pallor or swelling. Other

presentations were strabismus with disc pallor or swelling, acquired esotropia with diplopia, acquired exotropia, nystagmus, and disc swelling with headache [18].

Suprasellar tumors, by contrast, are associated with an 86% rate of visual loss in adults [19]. In a study conducted on brain tumors revealed that one-third of patients presented with ocular manifestations of brain tumors had craniopharyngiomas [20]. Literature showed rates of bilateral or unilateral loss of vision amongst patients with craniopharyngioma were 3% and 11.8% [19,21] respectively. The main finding in our case was sudden loss of vison which is agreement with the study conducted by Goldenberg-Cohen et al (2011), wherein the authors reported 40 % of the patients showed rapid loss of vision preoperative [22]. Local autoregulatory vascular changes and/or diversion of the cerebral blood flow into the ophthalmic circulation may normalize blood supply to the optic nerve in the presence of increased ICP, and therefore a rapid reduction in ICP could lead to immediate visual impairment due to the new autoregulatory vascular levels. As in the literature, we also suspect that the high susceptibility of the optic nerve to interruption in the blood supply contributed to the rapid visual loss [23,24]. Another possibility could be direct mechanical compression over the optic nerve or the optic chiasm by the suprasellar mass.

Eyes are the vital organs and plays a very important role to convey the underlying mental health status and the physical state of particular individual, hence a proper examination of eye is advisable in the patients presenting with underlying systemic conditions. The well-known fact is that the early diagnosis and treatment are essential to preserve vision in tumor-related optic neuropathy, and that recovery to normal levels is unlikely if profound loss of vision continues for weeks to months. In children, the problem is exacerbated because they often do not verbalize unilateral visual loss [25]. Further the Dorner et al (2007) revealed that half of the children included in their study were diagnosed for the other diseases and treated rather neglecting the possibility of an existing brain tumour [25].

Clinicians should be aware that in children, lack of awareness and poor cooperation during vision tests, may mask a rapid visual loss following tumor decompression. Brain tumors are well known as an important cause of optic neuropathy with visual loss, particularly suprasellar tumors such as craniopharyngioma and glioma. Paediatric suprasellar masses are considered unique in their clinical presentations and imaging features. This case-report aims to highlight the unusual presentation of suprasellar mass in paediatrics.Visual impairment is commonly associated in adult suprasellar masses, yet pediatrics can present with severe visual impairment. A multidisciplinary approach is advised in treating paediatrics with brain tumour including Ophthalmologist, Endocrinologist, Neurologist, Neurosurgeon and Radiologist. Authors conclude with a general recommendation to monitor the visual functions in all the children with Suprasellar tumors and the brain tumors in general.

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