Case Report

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Craniopharyngioma in Child

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Abstract

Craniopharyngiomas are histologically benign neuroepithelial tumors of the CNS that are predominately observed in children aged 5-10 years. People may present with bitemporal inferior quadrantanopia leading to bitemporal hemianopsia, as the tumor may compress the optic chiasm These tumors arise from squamous cell embryologic rests found along the path of the primitive adenohypophysis and craniopharyngeal duct. Although histologically benign, these tumors frequently recur after treatment. In addition, because they originate near critical intracranial structures (eg, visual pathways, pituitary gland, hypothalamus), both the tumor and complications of curative therapy can cause significant morbidity. These characteristics have led to various treatment approaches, and disagreement continues regarding optimal treatment in children with this disease. Other names are Rathke pouch tumors, hypophyseal duct tumors, or adamantinomas. Evidence suggests that adult craniopharyngiomas are histologically and biologically different from paediatric craniopharyngiomas; however, only childhood craniopharyngiomas are discussed in this article.

Keywords: Craniopharyngiomas; Benign; Quadrantanopia; Bitemporal Hemianopsia; Neuroepithelial Tumors; Primitive Adenohypophysis and Craniopharyngeal Duct.

Introduction

Baby X 7 years old male child was admitted in the paediatric surgical ward on 23/02/2016 with the complaints of diminished vision from past 6 months and moderate to sever head ache on frontal area with one episode of vomiting. Thge child was apparently normal before 6 months, and his decreased in vision was reported by his school teacher that he is not able to see words in blackboard. He went for routine eye checkup and suggested for improving diet and no other treatment measures were used. The symptoms progress and the child vision worsen with which he started banging on walls and doors and thus continuous diminishing of vision for which he went for 2nd opthalmalogic check up and he was refered to PGI chandighar and advised for MRI and the child was diagnosed for brain tumor. Therefore the clint PD for the same complaints and pediatric surgeon. The child underwent craniotomy and excision on 3rd march and the tumor was removed and culture was sent for histopathological examination. The incision from frontal area starting from right ear 13 sutures was made to close the incision. The conformation of diagnosis was Craniopharyngiomas. Postoperatively the child was complaining for diarrhoea and head ache.

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Disease Condition

Book picture **Patient picture**

Definition: Craniopharyngiomas are benign neuroepithelial tumors of the CNS. These tumors arise from squamous cell embryologic rests found along the path of the primitive adenohypophysis and craniopharyngeal duct. Craniopharyngiomas are the most common childhood tumor

Incidence:

- Craniopharyngiomas are relatively rare, representing 6-10% of intracranial malignancies in children and adolescents (approximately 2-3 cases per 1,000,000 children).
- a slight male preponderance has been historically reported.
- Peak incidence occurs in individuals aged 5-14 years.
- Neonatal craniopharyngiomas are rare.
- Higher incidence rates have been observed in Asia and Africa.

Age 7 year
Sex: male
Race: Asian

Causes:
Unknown
chromosomal abnormalities
environmental or infectious causes can predispose

Unknown

Pathophysiology

Due to chromosomal abnormalities like deletions, translocations, and increased copy numbers

↓

Tumors arise from cellular remnants of the Rathke pouch, which is an embryologic structure that forms both the infundibulum and anterior lobe of the pituitary gland.

↓

These tumors have been identified extensively in suprasellar, parasellar, and ectopic locations. Typically, the tumors arise within the sella or adjacent suprasellar space.

↓

Tumor mass compresses the adjacent normal intracranial structures.

↓

Obstructing cerebral spinal fluid (CSF) pathways (ie, third ventricle, Monro foramen) and causing hydrocephalus and increased intracranial pressure that leads to headaches, nausea, and projectile vomiting.

Clinical manifestations

- **Headache**: due to increased intracranial pressure or hydrocephalus.
- **Vomiting**: Classic projectile vomiting (frequently without nausea)
- **Vision loss**
  - Children are frequently unaware of significant vision loss; nevertheless, this symptom reportedly occurs in 20-60% of pediatric patients with craniopharyngioma at presentation.
  - Anterior extension to the optic chiasm can result in a classic bitemporal hemianopsia, unilateral temporal hemianopsia, papilledema, or unilateral/bilateral decrease in visual acuity. Classically, vision loss starts with a superior temporal field cut. However, the eccentric growth of these tumors can result in varying patterns and severity of vision loss, including decreased acuity, diplopia, blurred vision, and subjective visual field deficits. Children are frequently inattentive to visual loss, and formal testing may be required.
- **Seizures** due to Temporal lobe involvemen
- **Hyperactive** children with unusual eye movements and even blindness due to extrinsic compression of the hypothalamus.
- **Endocrine deficiencies** leads to

short stature, Weight gain, Lethargy, Fatigue, Cold intolerance, Dry skin, Dry brittle hair, Slow teething, Anorexia, Large tongue, Deep voice, Myxedema, Delayed puberty.

Before surgery:
Head ache on frontal area
Vomiting
Partial Vision loss (bitemporal hemianopsia)
Unusual eye movement
After surgery:
Head ache
Diarrhoea

Diagnosis:

- History
- Physical examination
- Preoperative intellectual or psychological assessment

- **History**: The child natal history was apparently normal.
- **General appearance:** Oriented, conscious, moderate body built.

- Serum electrolytes levels
- Hormonal studies
- Skull radiography
- Head CT scanning
- Brain MRI
- Cerebral angiography
- Histological studies

- **GCS score:** Eye 4 verbal 5, and motor 6,
- **Vital signs:** stable
- **Anthropometr**y: height 154cm, weight 18kg, 1st degree malnutrition (according to Gomez classification).
- **Growth and development** seems to be normal. And child was mild hyperactive and have hurried in speech.
- **Head to foot:** after surgery suture line are present, partial visual acuity. Unusual eye movement, pupillary dilatation, partial optic atrophy. Extra ocular eye movement abnormalities. Slow teething and deep voice, weight loss.
- No other abnormal physical findings.

Investigations:

- **Haematological investigation**: Hb: 11.3gm/dl, RBC 4.56mc/cum,TLC 7500cells/cumm,DLC-N 90%,E-01%, L-05%, m-04%, platelet – 3.11 lacks/c/cumm, Hematocrit 34.8%.
- **Hormonal studies:** T3 level is elevated.
- **MRI**: suggestive of possibilitiey of Craniopharyngiomas.
- **Histopathologicla examination**: suggestive of Craniopharyngiomas.
- **Medical management**

Tab valporate 200mg OD (morning)
Tab veona CR 300 mg OD (evening)
Tab pantop -20mg OD
Tab sporlac 120mg TDS

- **Surgical management**

Craniotomy and excision was done

Treatment:

- Long-term hormone replacement is the primary medical treatment: intranasal vasopressin (desmopressin acetate [DDAVP]), corticosteroids, thyroid hormones, growth hormones, and sex hormones.
- There is no role of chemotheraphy in craniopharingioma. Immunostimulatory therapies with interferon and intracystic/intratumoral injection of chemotherapeutic agents (eg, bleomycin) are occasionally used in cases of recurrent disease. (Bleomycin (Blenoxane), Interferon alfa 2a (Roferon-A))
- Intracavitary irradiation (brachytherapy) also shows no clear outcome
- Repeated Surgical management for recurrent craniopharyngiomas after radiation therapy also shows fewer prognoses.

Surgical Care

- Radical surgery
- Conservative surgery alone
- Conservative surgery with postoperative radiotherapy

Complications:

- Vision loss
- Growth hormone deficiency, (35-95%).
- Thyroid-stimulating hormone deficiency (21-42%),
- adrenocorticotropic
- Hormone deficiency, (21-62%)
- Antidiuretic hormone deficiency,
- Luteinizing hormone or follicle-stimulating hormone deficiency (38-82%)

Diencephalic syndrome

- TSH deficiency
- Diencephalic syndrome (hyperactive with unusual eye movements)

Prognosis:

- There is 10 years of survival rates of 86-100% among patients who underwent gross total resection
- Subtotal resection or recurrence treated with surgery and radiation therapy carry 10-year overall survival rates of 57-86%.
- The perioperative mortality rate after primary surgical intervention has been estimated to be 1.7-5.4%. However, the mortality rate after re-resection for recurrent disease can be as high as 25%.

- Not evident till date
- Expected to have poor prognosis in future

- Almost all patients with craniopharyngioma ultimately suffer from chronic endocrinologic morbidities. And significant neurologic morbidities such as vision loss, ataxia, behavioral problems, cognitive disabilities, and sleep disorders

Nursing Assessment

- Identification of risk factors for exposure to radiation or chemicals that is carcinogenic.
- Identify the signs and symptoms are: headache, vomiting, and decreased vision or double vision.
- Identify any changes in client behaviour.
- Observation of hemiparese or hemiplegia.
- Changes in sensation: hyperesthesia, parasthesia.
- Observation of sensory changes: asteregnosis (not able to feel the sharp edges), agnosia (not able to recognize objects in general), apraxia (not being able to use the tool properly), agraphia (can't write).
- Observation of vital signs and level of consciousness.
- Observation circumstances fluid and electrolyte balance.
- Psychosocial: personality and behavioural changes, difficulty making decisions, anxiety and fear of hospitalization, diagnostic tests and surgical procedures, a change in the role.

Possible Nursing Diagnosis

1. Ineffective tissue perfusion related to circulatory damage caused by a tumor suppression.
2. Impaired sensory perception decrease visual acuity related to optic nerve compression
3. Pain (Acute / Chronic) related to increased intracranial pressure.

Altered comfort irritability related to increased intracranial pressure

4. Fluid and electrolyte imbalance related to vomiting
5. Impaired family coping related to poor prognosis of the disease
6. Risk for injury related to poor visual acuity.
7. Risk for recurrence related to metastatic nature of the disease
8. Potential for complications vision loss related to poor prognosis of disease
9. Potential for neurological deficit related to poor prognoses and non availability of the chemotherapy and radiation therapy.
10. Knowledge Deficit: the condition and treatment needs related to the inability to know the information.

Conclusion

Even though it is a common childhood tumor the treatment mortalities for this kind of tumor is still not clear. There is no role of chemotherapy in curing tumor. Radiation therapy and surgical interventions also shows no complete cure. Repeated surgeries needed for recurrent cases. This need consultation and team work of Pediatric neurosurgeon, Radiation oncologist, Pediatric endocrinologist and Pediatric hematologist/oncologist.

Reference

1. Suraj G "The Short Textbook Of Paediatric Nursing" 11th edition, Jaypee publications, 565-566.
2. Archar's " Textbook of Paediatric Nursing" 4th edition, universities press, 534, 541, 543.
3. Nelson, "Textbook of Paediatrics" 10th edition, volume 1, Elsevier publication, 1746, 1752.
4. Karin R, Beth B " paediatric Acute Care" Jones and Bartlett publication, 375, 637 Nanda nursing interventions.