Early Detection of Pituitary Tumor to Avoid Complications – A Case of Pre Puberty Girl.

Gurulingappa I Herakal (Research Scholar) [1] , Dr. Praveenkumar.K [1], Dr Ellen Ghazi Safadi [1] , Ms. Heba Abdollah Palzevan [1], Mr. Nithin M Raj [2], Ms. Prathima Prasad Rai [2] , Ms. Ashli Shaji [2] , Mr. Jibin Simon [2.]

[1]Institution- Department of Anesthesia Technology, College of Health Science, Gulf Medical University, AJMAN, United Arab Emirates.
[2]Department of Anesthesia & Operation Theatre Technology, KSHEMA, NITTE (Deemed to be) University, Mangalore, Karnataka, INDIA.
[3]Department of Anesthesia & Operation Theatre Technology, College of Health Professionals, Manipal University, Udupi, Karnataka, INDIA.
[4]Department of Anesthesia & Operation Theatre Technology, Allied Health Sciences, Yenepoya University, Mangalore, Karnataka, INDIA.

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***Abstract*- Background**

The pituitary tumors are one of the rare events in adolescence and childhood. They often present as invasive macro adenomas and these tumors in children are almost not malignant, it may result in significant morbidity. This case report describes the histological characteristics, laboratory data, pathology features, and hospital course followed by the pituitary surgery.

Material and Method

The study uses case study of a ten year old pre-puberty girl, who was suffering from polyuria and polydipsia, referred from an oncology department. Patient came with complain of the severe headache and blurred vision and was admitted in the emergency department. The patient was observed by one of the researchers of the study and notes were taken for further discussion.

Results & Discussion

Based on consultation of neurologist, radiologist and oncologist further course of treatment was decided. Her brain MRI showed a 6 to 7 mm enhancing lesion in the sella turcica inside the pituitary gland with extension superiorly into the stomach and cause thickening of the pituitary stalk. She denied systemic disorders, headaches or any other symptoms and she was on Desmopressin as home medications. After surgery, it was recommended to repeat pituitary MRI or CT scan paranasal sinuses without contrast with navigation protocol. The patient is doing well at the time of writing of this paper.

Conclusion

Early diagnosis of pituitary tumors in children is important to avoid the adverse events on both physiological and cognitive outcomes. Genetic counseling should be offered to the family of the child confirmed with pituitary tumor as part of a genetic syndrome and surveillance of family members. Treatment of pituitary cancer needs integrated and proactive effort to avoid complications. The findings of the case study is useful or clinicians from specialty such as oncology, neurology and radiology.

***Index Terms*-** Pituitary adenoma, VTE Prophylaxis, Brain MRI, Sella turcica, Children

I. Background

Pituitary tumors are referred as benign with slow growth and aggressive clinical outcomes. They often present as invasive macro adenomas and the cases usually well controlled longitudinally with standard interventions and later their characteristics changes dynamically. Despite the comprehensive therapies: radiation, surgery, pharmacological or conventional medical treatment; the diagnosis of a pituitary tumor should be considered in patients with a radiological invasive tumor, that expected to grow rapidly. [1]

Pituitary tumor often reveals histological features that are consistent with increased proliferation, Ki-67 indices (above 3%), increased mitotic numbers and p53 expression. However, even in the presence of these features; the likelihood of predicting future aggressive behavior of the tumor reliably and the prognostic value of these markers remain controversial. [2, 1] Tumors with invasive growth and proliferative markers exhibited a higher rate of disease progression or recurrence at follow-up. Although, the efficacy of chemotherapy was limited; partial regression or transient stabilization was prevailed among most of the cases. The life expectancy in pituitary tumor patients was also found to be reduced remarkably. [1]

Headache and decreased vision are the most common symptoms caused by insufficient pituitary hormone or the tumor expansion. The primary treatment regimens for these patients are pituitary microsurgery with transsphenoidal or transcranial approach. It helps in the rapid reduction of tumor volume as well as decompression of the optic apparatus. Tumor debulking improves visual impairments and relieves headaches. However, after transsphenoidal surgery the prevalence of tumor recurrence was 12% to 46%. [3, 4] Patients with dopamine agonists or

somatostatin analogs exhibited a limited effect on the risk of tumor recurrence. Although, the use of dopamine agonist was associated with tumor size, medical therapy is not recommended as a routine basis for postoperative nonfunctioning pituitary adenomas. Postoperative radiotherapy was extensively used to prevent tumor recurrence. However, irradiation carries the risk of hypopituitarism, visual impairments, cerebrovascular disease, and neuro cognitive dysfunction. In spite of the reduced incidence, there is a high risk of developing secondary tumors: gliomas, astrocytoma or sarcoma. [4]

The incidence of pituitary tumor among general population was estimated as 20%. [5] These benign originate from the hormone secreting epithelial cells in the adenohypophysis of the pituitary gland. According to size, the tumors are classified as micro adenomas which are usually based in sella turcica with neoplasm of < 1 cm and macro adenomas (neoplasm ≥ 1 cm). [4, 6] Transsphenoidal surgical resection is considered for hyper secreting pituitary adenomas cases. In prolactinomas cases, the surgical resection is preferred only when there is treatment failure with dopaminergic medications, adverse events, severe side effects or tumoral mass effect. Patients with non-functional pituitary adenomas undergo surgery when there are clinically manifest mass effects: visual disturbances, headaches, or hypopituitarism. [6]

The decision to undergo transcranial surgery is considered mainly when there is an extrasellar extension or invasiveness of the tumor such as fibrosis; failed transsphenoidal surgery; uncertain diagnosis and inadequate decompression. Operative mortality of transsphenoidal surgery is less than 0.5% and its complications are associated with tumor size, optic apparatus, hypothalamus, and extrasellar extension. [7] For transcranial procedure, complications are associated with frontal lobe, optic nerve damage, and hypothalamic damage. Also; hemorrhage, transient diabetes insipidus, transient vision deterioration, cerebrospinal fluid leak, and death were found during immediate postoperative period followed by pituitary surgery. [6, 8]

Despite the short term follow up and the longitudinal effects of stereotactic radio surgery on visual impairment or pituitary function; use of radio surgery (stereotactic) was found to be effective in tumor control. The benefits of stereotactic radio surgery compared with fractionated radiotherapy include lower risk of hypopituitarism, visual dysfunction and radiation induced neoplasia. However, early postoperative radiation therapy should be considered to prevent recurrence or early reoperation should be preferred for residual adenomas are a critique. [4]

This report describes a case presentation of a pre puberty girl, who was suffering from polyuria and polydipsia. Her brain MRI showed a 6 to 7 mm enhancing lesion in the sella turcica inside the pituitary gland with extension superiorly into the stomach and cause thickening of the pituitary stalk. She denied systemic disorders, headaches or any other symptoms and she was followed Desmopressin as home medications. This study was conducted to find the histological characteristics, laboratory data, pathology features, and hospital course followed by the pituitary surgery.

Case presentation:

This case report includes a 10 year old pre puberty girl, referred from an oncology department, who was suffering from polyuria and polydipsia. An investigation including brain MRI with and without contrast showed a pituitary lesion with extension into the pituitary stalk. She denies any other symptoms or headaches or systemic disorders and consulted for her recently found pituitary lesion. She was followed Desmopressin as home medications.

The diagnostic test was reviewed and VTE Prophylaxis was performed. There was no hormonal abnormality except for lack of vasopressin and D1. Brain MRI showed a 6 to 7 mm enhancing lesion in the sella turcica inside the pituitary gland with extension superiorly into the stomach and cause thickening of the pituitary stalk. It is evident that; pituitary adenoma, Rathike's cleft cyst and intrasellar caniophyarygioma are the three most common pathologies found in the same age group. There are chances of other diagnoses: histocytosis X,teratoma or germinoma also. Diagnostic confirmation was done with surgical resection and pathology, however, before the surgery; all hormones were checked and it was found as normal and only her beta hCG was slightly higher, may be suggesting a teratoma. Ophthalmologist's observation reveals that, she had no eye symptoms or visual abnormalities and the nerve scan was normal.

Table 1: Clinical profile of the case [12]

Heart rate	70
Respiratory rate	18
BP (mmHg)	123/87
Height (Cm)	140.1
Weight (Kg)	40.4
BMI (Kg/M2)	20.6
Temperature	36.2
Pulse	116
Pulse OX	100

Based on the pathology and biopsy, transcranial or transsteroidal surgeries were recommended. The risk and potential benefits of the selected endoscopic approach to the pituitary gland was explained. The risk can be infection, bleeding, (carotid damage, cavernous sinus, intracranial bleed) damage to the optic nerve or chiasm and visual problem, continued CSF leakage, hypo pituitarism, persistence or continuation or worsening of DI, unpredictable medical complications and even coma or death.

Desmopressin acetate (Minirin melt 60 Mcg Tab) 60 Mcg Tab 60 Mcg, SUBLING BID: 60 days were prescribed. Hydrocortisone IV (100 mg) before the surgery was given, then 20 mg IV hydrocortisone q6h. For IV fluids D5 + ½ NS start at 1.5 maintance (1 + ½ maintance) and continuously monitored I & O q hr with SP. Gravity qhr Urine Osm, Serum Osm, electrolyte q4h. After surgery, it was recommended to repeat pituitary MRI or CT scan paranasal sinuses without contrast with navigation protocol.

II. Discussion

Nonfunctioning pituitary adenoma (not secreting hormones) comprised of 80 % of all pituitary macro adenoma. Pituitary lesions are often detected with the help of imaging of the head. However, it is difficult to discover the incidence if case is asymptomatic to the hyper secretion of pituitary hormone. Important markers of the pituitary tumor are hormones of the pituitary gland (anterior and posterior), adrenocorticotropic hormone (ACTH), thyroid stimulating hormone (TSH), luteinizing hormone (LH), follicle-stimulating hormone (FSH), and growth hormone (GH). [9]

Table 2: Signs and symptoms of pituitary hormone deficiency (Extracted from the Pituitary Society) [9, 10]

Pituitary hormone	Target organs	Effects of deficiency
ACTH	Adrenal glands: cortisol and DHEA	Fatigue, low sodium in blood, weight loss, skin pallor
TSH	Thyroid gland: thyroid hormone	Fatigue, weight gain, dry skin, sensitivity to cold, constipation
LH and FSH in Women	Ovaries: estrogen, progesterone; ovulation	Loss of periods, loss of sex drive, infertility
LH and FSH in Men	Testes: testosterone, sperm production	Loss of sex drive, erectile dysfunction, impotence, infertility
GH in Children & Adolescents	Bone, muscle, fat	Lack of growth (height); increased body fat, failure to achieve normal peak bone mass
GH in Adults	Whole body	Poor quality of life, increased body fat, decreased muscle and bone mass
PRL	Breast	Inability to breast feed
Oxytocin	Breast, Uterus	Complete deficiency could make breast feeding difficult
Antidiuretic hormone (vasopressin)	Kidney	Frequent urination (day & night), dilute urine, excessive thirst

Source: Compiled on the basis of [Published online 2016 Apr 11. doi: 10.1186/s12998-016-0093-z**]**

Clinically relevant pituitary adenomas are less common among general population. The 5% of pituitary tumors are female in origin and it can be isolated to the pituitary as in familial isolated pituitary adenomas. [11] The pituitary tumors are one of the rare events in adolescence and childhood, with a prevalence of one per million children. Pituitary tumor accounts 2% to 6% of surgically treated tumors that occur in children. Although these tumors in children are almost not malignant and hormonal secretion is rare, it may result in significant morbidity. [12, 13]

Tumors in the pituitary fossa are craniopharyngiomas and adenomas. The craniopharyngiomas cause symptoms by compressing normal pituitary, resulting hormonal deficiencies and producing mass effects on surrounding tissues as well as the brain. Adenomas results hormonal conditions such as hyper prolactinomas, Cushing disease and acromegaly or gigantism. However, little is known about the genetic causes of sporadic lesions, which comprise the majority of pituitary tumors, but in children, more frequently than in adults, the tumors may be a manifestation of genetic conditions such as familial isolated pituitary adenoma, Carney complex, multiple endocrine neoplasia type 1, and McCune-Albright syndrome. [12]

Continuous follow up is important for the early detection of tumor recurrence. It includes MRI, assessment of systemic symptoms, blood cortisol levels, hormonal abnormalities and visual field defects. Even though, complete removal of the tumor during the first surgery is recommended, cases with residual tumors may undergo radiation or drug therapy. Due to the risk of craniotomy, recurrent pituitary tumor cases are usually treated with endoscopic transsphenoidal surgery. [14]

III. Conclusion

Early diagnosis of pituitary tumors in children is important to avoid the adverse events on both physiological and cognitive outcomes. Treatment of pediatric pituitary tumors requires critical appraisals in the process of diagnosis, treatment regimens, and proper management to facilitate early diagnosis, treatment and reduce morbidity. Genetic counseling should be offered to the family of the child confirmed with pituitary tumor as part of a genetic syndrome and surveillance of family members. Repeat pituitary MRI or CT scan was recommended to prevent the tumor recurrence and other adverse events.

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AUTHORS

First Author – Mr.Gurulingappa I Herakal (Research Scholar) Institution- Department of Anesthesia Technology,College of Health Science, Gulf Medical, University, AJMAN, United Arab Emirates.

First Author – Dr. Praveen Kumar.K, Dean of College of Health Science,Gulf Medical University,AJMAN,United Arab Emirates.

First Author – Dr Ellen Ghazi Safadi, Institution- Department of Anesthesia Technology,College of Health Science,Gulf Medical University,AJMAN,United Arab Emirates.

First Author – Ms. Heba Abdollah Palzevan, Student of Anesthesia Technology, Department of Anesthesia Technology, College of Health Science, Gulf Medical University, AJMAN, United Arab Emirates.

Second Author – Mr. Nithin M Raj, Institution- Department of Anesthesia Technology,College of Health Science,Gulf Medical University,AJMAN,United Arab Emirates.

Second Author – Ms. Prathima Prasad Rai, Department of Anesthesia and Operation Theatre Technology,KSHEMA,NITTE (Deemed to be) University,Mangalore,Karnataka,INDIA.

Second Author – Ms. Ashli Shaji, Department of Anesthesia; Operation Theatre Technology, College of Health Professionals, Manipal, University, Udupi, Karnataka, INDIA.

Second Author – Mr. Jibin Simon, Department of Anesthesia & Operation Theatre Technology, Allied Health Sciences, Yenepoya University, Mangalore, Karnataka, INDIA.