Tallest Nepalese Lady with Acromegaly: A Case Report

An 18 year-old girl from Dhorna-2, Siraha presented to the department of neurosurgery, National Neurosurgical Referral Centre, Bir hospital with features of gigantism and paraparesis grade 1/5. She was seven feet and three inches tall with body weight of about 100 kilograms. Radio imagings of brain showed pituitary macroadenoma (1.9 cm in diameter) with serum Growth Hormone (GH) level 20ng/ml. She underwent total excision of tumor by transsphenoidal approach. Although she had troublesome hyperglycemic and diabetic incipidus with hyponatremia, she made a good recovery. Literature on GH secreting pituitary tumor is reviewed here.

Key words: Acromegaly, GH secreting pituitary tumour, Transsphenoidal pituitary surgery.

Case Report

An 18 year-old right handed female was admitted to the National Neurosurgical Referral Centre, Bir Hospital on 26th November 2010 with the history of abnormal height, excessive enlargement of hands and feet, hoarse voice, amenorrhea and weakness of both lower limbs. (Figure 1&2) She was a grade tenth student, unmarried girl. She was a known case of insulin dependent diabetes mellitus on regular insulin since last six months. On examination her BP was 120/80 mmHg and pulse was 68/min, regular. She had a height of seven feet and three inches with body weight of about 100 kilograms with gross acromegalic features. She was fully conscious and oriented, her cranial nerves and sensory examination were normal. Motor examination revealed paraparesis grade 1/5. There were no cerebellar sings and no signs of meningeal irritation. Her plain X-Ray of skull lateral view showed enlarged pituitary fossa. MRI Brain showed about 1.9 cm x 1.2 cm
x1.5 cm mass in sella extending to suprasellar cistern with no compression of optic chiasm (Figure No). MRI of spine revealed degenerative changes in lower dorsal and lumbar spine with mild central spinal canal stenosis with multiple level disc prolapse in the lumbar spine (Figure No). Serum cortisol and prolactin were within normal limits whereas serum growth hormone was in higher range (20 ng/ml). Hematology and biochemistry were within normal limits. Visual field examination with perimetry was normal. Preoperative nasal swab culture yielded staphylococcus aureus, sensitive to most of antibiotics.

On 10th December 2010 she underwent transsphenoidal pituitary resection. Intraoperatively, the sphenoid sinus was multiseptated. The tumor was intrasellar, soft and grayish white in color. There was total excision of tumor with no intraoperative complications. Immediate postoperatively she was fully conscious and oriented. She started having liquids from the first post operative day. She developed diabetes insipidus which was managed with fluid replacement and vasopressin injection. She had persistent hyponatremia postoperatively which was managed with oral and intravenous sodium. There was no CSF rhinorrhoea postoperatively.

She also had severe hyperglycemia from early postoperative period which was managed with intravenous and subcutaneous insulin. There was not any cardio respiratory abnormality. Postoperatively serum growth hormone level came down to 2ng/ml. There was no visual field defect. Her paraparesis didn’t require any further surgical intervention, it improved with gradual control of blood sugar and limbs physiotherapy.

**Discussion**

Most primary pituitary tumors are benign adenomas which arise from the anterior pituitary gland (adenohypophysis). Neurohypophyseal tumors are very rare. Pituitary tumors account for about 10-12% of all...
intracranial tumors. These tumors can be either microadenomas (<1 cm) or macroadenomas (>1 cm in diameter). Classically pituitary adenoma is broadly divided into functional (secreting) and non functional (non secreting). Non functional tumors account for about 40% of all pituitary tumors which are inactive (does not produce any hormone) and usually does not present until it reaches sufficient size to cause neurologic deficit by mass effect. Remaining 60 – 75% of pituitary tumors are functional which presents early with symptoms caused by physiologic effects of excess hormones they secrete. Among the functional tumors, GH secreting tumors comprise about 20-25%. These tumors when present in childhood before the closure of epiphysis of long bones leads to gigantism while causes acromegaly after closure of epiphysis.

Giants and Acromegalic persons have been described in literature and art throughout history. Pierre Marie described acromegaly as a medical syndrome in 1886. Late by 1900, Benda and others suggested that hypertrophy or hyperplasia of pituitary might be the cause of acromegaly. Growth hormone secreting pituitary tumor mostly occur during 3rd to 4th decade of life with slight female predominance. The majority of cases of acromegaly and gigantism are produced by the neoplasm of the somatotropic cells of anterior pituitary gland, however, acromegaly and gigantism can also occur as a result of hyperplasia of somatotropes either independently or in response to excessive amount of growth hormone releasing factor due to multiple endocrine neoplasia (MEN type I).

The specific clinical presentation of GH secreting pituitary tumors are acral and facial enlargement and gigantism (when prepubertal), headache, peripheral nerve entrapment syndromes, joint pain, myopathies, generalized splanchnomegaly, impaired glucose tolerance with diabetes mellitus and visual disturbances. Later in life severe degenerative osteoarthritis & spinal stenosis may occur. Surgical options include transsphenoidal and transcranial (either ptierional or transfrontal) approach. The first surgery on pituitary for acromegaly was performed in Vienna in 1908 by Hochonegg using Schloffer’s transsphenoidal approach. Transsphenoidal surgery is the first treatment of choice in otherwise healthy subjects and more than 90% of pituitary adenomas are amenable by this approach. Large tumor with suprasellar or parasellar extension may still require transcranial approach if adequate removal of tumor is to be carried out. Transsphenoidal reoperation should also be considered first if initial surgery has failed to normalize GH levels especially in small tumors confined to sella. Intraoperative C- arm, operating microscope and endoscopic systems are the standard requisites for a safe transsphenoidal pituitary surgery. Endoscopic endonasal and transnasal stereotactic surgeries have been tried but long term results has to be awaited.

The commonest postoperative complications of transsphenoidal pituitary surgery are diabetes insipidus, electrolyte imbalance, CSF fistulas and meningitis, hypopituitarism and fatal intracranial hemorrhage from vascular injuries. Thus a careful imaging to ascertain the position of the carotids in relation to the tumour with coronal MR or cavernous sinogram is therefore mandatory before surgery.

Radiation therapy is recommended after surgery in tumors invading the cavernous sinus and the skull base.
or in persistent elevated GH levels \(^9,14\). Pharmacotherapy should be used if all other treatment methods have failed or as a prerequisite to pituitary surgery in large tumours and bromocriptine or sadostatin (octreotide) are the current drugs of choice \(^4,10,17,19,25\).

The most stringent criteria for surgical cure in acromegaly-gigantism is glucose suppression GH levels < 2ng/ml or basal serum GH<2 ng/ml \(^12,14,24\). The recurrence rate is 0% if glucose-suppressed GH levels are <2ng/ml and upto 10% if basal GH levels of <5ng/ml is considered as criteria of cure \(^2,3,5,12,13,15,21,23,24\).

The present case, although a macroadenoma (1.9 cm diameter), stands a reasonable good chance of long-term remission as the postoperative basal GH level is 2ng/ml. She will however require to follow with regular endocrine and imaging studies for a long time. Cause for paraparesis couldn’t be explained but there has been few cases described in literature \(^27\) regarding the association of pituitary tumors with spinal stenosis causing paraparesis.

**References**


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**Figure 5:** MRI lumbosacral spine showing multiple level disc prolapse.

**Figure 6:** Densely granulated growth hormone cell adenoma.


