Dexmedetomidine for Conscious Sedation in Bilateral Inferior Petrosal Sinus Sampling

ABSTRACT
Cushing’s syndrome in an acromegalic patient is a very rare presentation. To differentiate a Cushing’s disease from Cushing’s syndrome due to ectopic adrenocorticotropic hormone (ACTH) secreting tumors, inferior petrosal sinus sampling (IPSS) is required. Acromegaly patients have associated airway abnormalities posing a challenge to administration of anesthesia. Traditionally, most IPSS was done under general anesthesia. But now it is being recognized that general anesthesia for this procedure has its own implications and hence conscious sedation is being used for this purpose. We describe our experience with the novel agent dexmedetomidine for conscious sedation in this procedure.

Keywords: Acromegaly, Cushing’s syndrome, Dexmedetomidine, Inferior petrosal sinus sampling.

INTRODUCTION
Cushing’s syndrome in an acromegalic patient is a very rare presentation. Inferior petrosal sinus sampling (IPSS) is a procedure done to differentiate a Cushing’s disease from Cushing’s syndrome due to ectopic adrenocorticotropic hormone (ACTH) secreting tumors. We describe a rare case of acromegaly who presented with Cushing’s syndrome posted for bilateral IPSS. This patient was managed successfully with conscious sedation on dexmedetomidine.

CASE REPORT
A 19-year-old male patient weighing 93 kg was diagnosed as a case of acromegaly presented with headache and intermittent giddiness since 2 to 3 years. He had coarsened facial features with prognathism. The patient had enlarged tongue. Mouth opening was more than 2 fingers and Mallampatti class III. He had thickened skin with thoracic kyphoscoliosis to the left. Abdominal striae were present. Blood pressure was 134/92 mm Hg. Chest X-ray and electrocardiogram was normal. A hormonal profile revealed increased growth hormone (GH), cortisol, and testosterone. Free T4 and prolactin was decreased. Blood sugar was normal. Magnetic resonance imaging of brain showed pituitary vault thickening with hyperpneumatization of mastoid air cells and frontal sinuses suggestive of acromegaly.

Clinical presentations, imaging, and hormonal study were suggestive of Cushing’s syndrome with hypothyroidism along with acromegaly. The patient was started on tablet thyronorm 75 µg twice a day. He was then posted for IPSS.

A decision was made to perform the procedure under local anesthesia with sedation. Preanesthetic preparation included difficult airway cart, long IV extension tubings due to inaccessibility, and under body warming mattress. American Society of Anesthesiologists (ASA)-standard monitors were used. The patient was premedicated with midazolam 0.03 mg/kg and fentanyl 2 µg/kg. Urinary catheterization was done. A loading dose of dexmedetomidine 0.5 µg/kg was given over 10 minutes followed by an infusion of 0.5 µg/kg/hour. Bilateral femoral puncture site was infiltrated with a mixture of 2% lignocaine and 0.5% bupivacaine. Using the Seldinger technique, catheterization of the right and left femoral veins was done. Systemic heparinization was done with 100 U/kg of heparin. Femoral catheters were advanced bilaterally up to the inferior petrosal sinuses. After confirmation of catheter location by fluoroscopy, samples for ACTH were drawn simultaneously from the right and left sinus at the same time that a peripheral level was drawn from an arm vein. Corticotropin releasing hormone was then injected to provoke ACTH release following which samples were collected again. The procedure lasted for 2 hours 10 minutes. The patient maintained stable hemodynamics throughout the procedure. Intermittent verbal communication with the patient was made to detect any neurological symptoms due to the procedure. Dexmedetomidine infusion was stopped just after completion of the procedure. The patient tolerated the procedure well and was shifted to ward.
DISCUSSION

Acromegaly involves oversecretion of GH. It has an incidence of 3 to 4 cases per million per year. Clinical features include somatic changes like coarse facial features, macroGLOSSIA, and prognathism. Airway involvement occurs and is of concern to the anesthesiologist. Pharyngeal, laryngeal and tracheal obstruction may occur due to hypertrophic changes. Laryngeal soft tissue, epiglottis, and aryepiglottic folds may be involved. Testing due to hypertrophic changes. Laryngeal soft tissue, epiglottis, and aryepiglottic folds may be involved. Testing for blood concentrations of GH along with oral glucose tolerance test is usually used for diagnosis. Magnetic resonance imaging of the pituitary is used to locate and detect the size of the tumor causing GH overproduction. Most GH-secreting tumors are macroadenomas and can compress surrounding normal pituitary tissue which alters production of other hormones. The most common cause of Cushing’s syndrome is Cushing’s disease, but in 15% of cases it may be due to ectopic production of ACTH. Double endocrine dysfunction of active pituitary adenomas leading to acromegaly and Cushing’s disease may occur simultaneously, and these lesions are very rare. They are mostly microadenomas and are usually clinically silent.

The most reliable test to differentiate Cushing’s disease from Cushing’s syndrome due to ectopic ACTH production is bilateral IPSS. The inferior petrosal sinuses (IPSs) receive drainage from the pituitary gland without mixture of blood from other sources. Therefore, if the ACTH levels in the IPS are high compared to an ACTH drawn in the periphery, the patient has Cushing’s due to pituitary adenoma. In contrast, if the ACTH in the IPS and the periphery is equivalent to its Cushing’s due to ectopic ACTH production.

Traditionally, this procedure has been performed under general anesthesia. However, the choice of anesthetic agents poses a dilemma. General anesthesia and surgery is known to affect the hypothalamic pituitary adrenal axis. Propofol, thiopentone, and etomidate can suppress ACTH secretion. They have direct antisteroidogenic effects on adrenal cells and are a weak inhibitor of adrenal steroidogenesis and, hence, can alter the results. General anesthesia in this patient poses additional problems due to difficult airway. Hence, we kept all equipment ready for difficult intubation. Another preferred technique nowadays is local anesthesia with conscious sedation. It has many advantages. The patient remains sedated but arousable and can respond to verbal stimuli. Hence, onset of neurodefects can be picked up early which may warrant abandoning the procedure.

The U.S. National Institute of Health recently published data on four patients who had neurologic complications during IPSS in a total series of 500 patients. In the two cases where the procedure continued, permanent neurologic damage ensued. All neurologic problems were resolved within 48 hours in other two cases where the procedure was stopped immediately after development of symptoms.

We used dexmedetomidine for conscious sedation. It is a highly selective α2-adrenoceptor agonist having sedative, analgesic, and anxiolytic properties with no respiratory depression. It induces sedation by decreasing activity of neurons in the locus ceruleus in the brain stem, thereby increasing activity of inhibitory gamma-aminobutyric acid neurons in the ventrolateral preoptic nucleus. Sedation by dexmedetomidine mirrors natural sleep. It has no effect on adrenal steroidogenesis and hence may not interfere in the results of the intervention.

CONCLUSION

Acromegaly with Cushing’s syndrome is a rare combination. The novel agent dexmedetomidine can be successfully used for conscious sedation for IPSS, thus avoiding the interference of other anesthetic agents and the airway challenges associated with general anesthesia in acromegalic patients.

REFERENCES