Tolosa hunt syndrome: A new challenge for anaesthesia

Dear Editor,

Tolosa hunt syndrome (THS) is a rare steroid responsive painful ophthalmoplegia due to non-specific inflammation of cavernous sinus or superior orbital fissure. Granulomatous periarteritis of cavernous carotid, proliferation of fibroblasts, infiltration of cavernous sinus septa and wall, with lymphocytes and plasma cells are the primary etiologies.\(^1\) Classic causes being vascular aneurysm or fistula, neoplasms and infection. THS, also known as idiopathic pseudo tumor, commonly presents in the 5\(^{th}\) decade of life.\(^2\) The usual presentation is chronic and severe hemicranial headache, painful ophthalmoplegia, proptosis, orbital congestion, vertigo, reduced or loss of vision. The associated multiple cranial nerve palsies, intracranial extension, bony destruction, close proximity to blood vessels, endocrinopathy, raised intracranial pressure are the anesthetic challenges. We had a 56 year old male, known case of Tolosa Hunt syndrome, hypertension, uncontrolled Diabetes with complaints of right sided headache, complete ptosis of right eye with vision loss for one month. On examination neuropathy involving right sided cranial nerves II, III, IV, maxillary and mandibular branches of the trigeminal nerves were noted. MRI and CT brain with contrast showed Cavernous sinus inflammation with sphenoid sinus involvement and bony erosions, encasing the right internal carotid artery (ICA) and ophthalmic vein [Figure 1]. He was planned for nasal endoscopic biopsy under general anesthesia, after explaining the risks. Invasive arterial blood pressure monitoring was done considering the possibility of bleeding. Intra operatively he lost 700 ml blood as the lesion was close to major vessels and bleeding was controlled with nasal packing. Due to continuous bleeding from the surgical area, the patient shifted to Intensive care unextubated. Following that coiling of right ICA and embolization of inferior maxillary artery was done. After the procedure, on day 2 he had focal seizures involving right upper limb. CT brain showed multiple focal infarctions involving right anterior cerebral artery and middle cerebral artery. During the further course in the hospital, inspite of high intensive care, patient’s life could not be saved.

The ideal diagnostic work up include routine blood investigations, inflammatory markers, fasting glucose, CSF evaluation, ANA, anti-dsDNA, c-ANCA, MRI, conventional angiography, biopsy and fundoscopy.\(^3\)

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**Figure 1:** MRI image showing enhancing lesion in the right Cavernous sinus with erosions and extension into Orbital apex
The associated intracranial tumors can increase intracranial pressure which mandates neuroprotective anesthesia with cerebral perfusion pressure (CPP) of at least 60 mmHg, maintenance of normocapnia and normothermia, avoidance of stress response, rapid changes in hemodynamics, drugs that increase cerebral blood flow. Cases with peri and post operative seizures, delirium have also been reported.[4,5]

Endocrinopathy due to associated pituitary adenoma alters secretion growth hormone, prolactin, Thyroid stimulating hormone, Adrenocorticotropic hormone, Luteinizing and follicle stimulating hormone leads organ dysfunction and electrolyte imbalance requires correction pre operatively.

The presence of vascular lesions requires hypotensive anesthesia to minimize bleeding but the maintenance CPP and associated risk of stroke and Myocardial infarction makes it difficult. Patients on anticoagulants in case of primary thrombosis have to be advised regarding their stoppage and bridging therapy.

Adequate protection of eyes during anesthesia in patients with exophthalmos and pre operative evaluation and optimization vertigo are important. Although Tolosa hunt syndrome is a self limited disease, multisystem involvement, risks of bleeding and infarction with significant mortality and morbidity has to be considered during anesthesia management.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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