Glossopharyngeal Neuralgia

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Risk

- Majority of cases of GPN are idiopathic.
- Increased prevalence with extracranial neoplasms, trauma/infection/inflammation to tonsils, and pharynx, arachnoiditis.
- More common in pts older than 50 y and middle-aged females.

Perioperative Risks

- Vagoglossopharyngeal neuralgia occurs in 10% of pts with GPN. Attacks of pain can trigger bradycardia/asystole, arterial hypotension, syncope, ECG changes (arrhythmias), or even cardiac arrest.
- Tonic-clonic limb jerking and facial movements that resemble seizure activity can accompany attacks of pain.

Worry About

- Bradycardia, asystole, arterial hypotension, syncope, arrhythmias, and cardiac arrest during pain attacks
- Drug interactions with anticonvulsants: Carbamazepine, phenytoin, and oxcarbazepine
- Chronic narcotic use

Overview

- Rare: Represents ~1% of facial pain cases.
- Sudden, sharp, and excruciating pain shooting to the pharynx, tonsil, base of tongue, with possible radiation to eustachian tube and inner ear structures and/or mandible angle.
- Attacks may be triggered by swallowing (most common), chewing, talking, coughing, or yawning.
- Paroxysms of pain are usually <1 min and can recur after brief periods.
- Clusters of attacks last from weeks to months.
- Trigger zones can be located when application of topical anesthetic solution relieves pain.
- Pain typically stays on same side, and left side symptoms are more common (3:2).
- Attacks can precipitate bradycardia, syncope, tachycardia, and arterial hypotension.
- Cranial nerve (IX) receives afferent input from chemoreceptor and stretch baroreceptor of carotid body and carotid sinus, which may be responsible for CV reflex symptoms.
- Differential Dx can include trigeminal neuralgia, superior laryngeal neuralgia, cluster headache, or sick sinus syndrome.

Etiology

- Usually idiopathic
- Secondary causes:
  - Vascular compression of the glossopharyngeal nerve (most common)
  - Neoplasms (cerebellopontine, skull base, pharynx, tongue, laryngeal carcinomas)
  - Infection (tonsillitis, pharyngeal abscess, arachnoiditis)
  - Trauma (skull base fractures, tonsillectomy, dental extraction, neck dissection)
  - Other (Chiari I malformation, MS, elongated styloid process [Eagle's syndrome])

Usual Treatment

- Pharmacologic treatment involves anticonvulsants: Carbamazepine, gabapentin, phenytoin, oxcarbazepine, pregabalin.
- Nerve block and possible neurolysis.
- Microvascular decompression is the best surgical treatment, with >70% success rate.
- Rhizotomy of the glossopharyngeal (IX) nerve is surgical alternative for MVD.
- Evolving care includes gamma knife surgery and stereotactic radiosurgery.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Bradycardia, tachycardia, syncope, hypotension	Syncope, palpitations	BP HR/rhythm	ECG or biotelemetry to capture pain attacks
CNS	Pain in IX/X distribution	Paroxysmal pain attacks in IX/X distribution with various triggers	Attempt to trigger pain and find distribution	MRI/MRA to ID etiology and vascular compression

Key References: Blumenfeld A, Nikosskaya G: Glossopharyngeal neuralgia, *Curr Pain Headache Rep* 17(7):343, 2013; Kandan SR, Khan S, Jeyaretna DS, Lhatoo S, Patel NK, Coakham HB: Neuralgia of the glossopharyngeal and vagal nerves: long-term outcome following surgical treatment and literature review, *Br J Neurosurg* 24(4):441–446, 2010.

Perioperative Implications

Preoperative Evaluation

- Assess triggers and subsequent pain with emphasis on Hx of bradycardia, palpitations, syncope, and seizures.
- Check medications, dosing, and efficacy, and review potential side effects along with drug interactions. Maintain preop regimen.

Monitoring

- Monitor preinduction arterial line in pts with significant CV symptoms and central venous catheter when temporary pacemaker might be indicated (vagoglossopharyngeal neuralgia).

Airway

- Direct laryngoscopy can trigger an attack.
- Topical anesthesia to oropharynx prior to laryngoscopy can blunt CV symptoms.
- Glossopharyngeal nerve block is an alternative to topical anesthesia for prophylaxis.

Maintenance

- Remain vigilant and promptly treat cardiac symptoms and labile BP.
- Watch for sudden arterial hypotension, bradycardia, and cardiac arrhythmias.

Extubation

- Look for possible IX/X nerve palsy and subsequent vocal cord paralysis following microvascular decompression surgery.

Anticipated Problems/Concerns

- Direct laryngoscopy triggering a pain attack with hypotension, bradycardia, and cardiac arrhythmias
- Periop pain attack with severe uncontrolled pain
- Chronic narcotic use and tolerance

Glucose-6-Phosphate Dehydrogenase Deficiency

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Risk

- Most common enzyme deficiency in the world.
- Due to X-linked recessive inheritance.
- Worldwide incidence: 400 million.
- Regions with highest prevalence include Africa, Southeast Asia, the Mediterranean, and the Middle East.
- In USA, G6PD deficiency is prevalent among black males and immigrant populations from the previously listed regions. Approx 10% of African-American males have G6PD deficiency.

Perioperative Risks

- Increased risk of acute hemolysis of RBCs with exposure to oxidative stressors.
- Infection and surgical stress can lead to hemolysis.
- Severe hemolysis may require transfusion and acute renal failure requiring hemodialysis.

Worry About

- General anesthesia masks early signs and symptoms of hemolytic crisis. Hypotension with hemolysis can be attributed to other causes, delaying diagnosis of acute hemolytic crisis.
- Early recognition and treatment of hemolytic anemia is required to prevent permanent neurologic damage, renal failure, or death.

Overview

- Enzyme deficiency is associated with chronic and/or acute hemolysis of RBCs.
- Most pts with G6PD deficiency are clinically asymptomatic unless exposed to triggers.
- Hemolysis occurs when pts are exposed to an oxidative stressor: infection, oxidative drug, fava beans, metabolic derangements.
- Hemolysis is usually seen 1–3 d after exposure.
- Clinical manifestations include fatigue, lumbar pain, abdominal pain, jaundice, splenomegaly, hemoglobinuria, scleral icterus, hypotension, tachycardia, dyspnea, headache, and pallor.
- Acute hemolysis is self-limited; resolution occurs within 4–7 d.
- Chronic nonspherocytic hemolytic anemia with severe deficiency may occur (<10% of normal enzyme levels).
- Associated with neonatal jaundice.

Etiology

- G6PD is an enzyme that catalyzes NADP into NADPH in the pentose phosphate pathway. NADPH then generates antioxidants that protect cells against oxidative damage.
- RBCs' only source of NADPH is through the pentose phosphate pathway. G6PD deficiency results in