Glossopharyngeal Neuralgia

Definition
Glossopharyngeal neuralgia (GPN) is a unilateral painful disorder that is characterized by brief, electric-shock-like pains, is abrupt in onset and termination, and is localized to the ear, the base of the tongue, the tonsillar fossa, or beneath the angle of the jaw. It has many of the same characteristics as trigeminal neuralgia (TN).

Epidemiology
GPN is a very rare disease, and there are very few studies on its prevalence. Its incidence in the general population has been reported as 0.2 per 100,000 people per year. It can coexist with TN.

Pathophysiology
Current opinion is that GPN is caused by compression of the glossopharyngeal nerve root close to the brainstem (dorsal root entry zone) by a tortuous blood vessel (an artery or vein), leading to mechanically twisted nerve fibers and secondary demyelination, probably mediated by microvascular ischemic damages. These changes lower the excitability threshold of affected fibers and promote cross-talk between adjacent fibers. Thus, tactile signals coming from the fast myelinated (A-beta) fibers can directly activate the slow nociceptive (A-delta) fibers, and sometimes also the C fibers, resulting in the high-frequency discharges characteristic of GPN.

Clinical Features
Location, radiation: The pain is unilateral and may be felt in any one of or all the following locations: the ear, the base of the tongue, the back of the throat (especially the tonsillar fossa), and beneath the angle of the jaw, and it can radiate down the neck.

Character: Electric-shock-like, shooting, stabbing, or sharp.

Severity: Mild to moderate.

Duration, periodicity: Each attack of pain lasts between a few seconds and two minutes, but can rapidly be followed by another attack. Spontaneous remission periods can occur, which initially
can last for months or years, but with time the remission periods get shorter, and the attacks also increase in severity.

Factors affecting it: Provoked by swallowing, talking, or coughing.

Associated features: Cardiac dysrhythmias and syncope may occur due to stimulation of the vagus.

Investigations
Computed tomography (CT) or magnetic resonance imaging (MRI) may reveal lesions, as well as neurovascular compression. They may also show an elongated styloid process, which, rarely, can cause the same pain and is termed Eagle’s syndrome.

Therapy
No trials have been conducted in patients with GPN, and so treatment is the same as for TN. First-line therapy should be carbamazepine (200–1200 mg/day) or oxcarbazepine (600–1800 mg/day).

Surgical Treatment
If medical treatment is not successful, surgical procedures can be considered. Microvascular decompression of the glossopharyngeal nerve is technically more difficult than for TN, but the results are similar. The major complications include dysphagia, hoarseness, and facial paresis.

References


Online Resources