Cluster Headache (CH)

Introduction - The Trigeminal Autonomic Cephalalgias (TACs) are a group of primary headache disorders that share similar clinical features but differ in frequency, duration, triggers, and treatment. “Cluster headache” is the most common of the TACs at a prevalence of 1 in 1000.

Cluster headache is 3 times more common in men, with a typical age of onset between 20 and 40 years of age.

Cluster headache is a unilateral headache syndrome with ipsilateral cranial autonomic features *and/or* restlessness. Autonomic features in CH could be one of the following:

- Conjunctival injection *and/or* lacrimation
- Nasal congestion and/or rhinorrhea
- Eyelid edema
- Forehead and facial sweating
- Miosis *and/or* ptosis

An individual attack in CH lasts 15 to 180 minutes and occurring up to 8 times per day. The most peculiar feature of cluster headache its clocklike regularity (circadian pattern).

The two forms of cluster headache are an episodic CH (*about 85-90% of CH*), where patients have a headache-free period of more than 3 months, and a chronic CH (10-*15 % of patients*), where the headache-free period is less than 3 months

Pathophysiology- The pathophysiology of CH is complex and the hypothalamus, trigeminovascular system, and the parasympathetic system play significant roles and their involvement have been discussed in detail .Calcitonin Gene Related Peptide (CGRP) is a marker of trigeminovascular activity and involvement of CGRP in CH has been shown. CGRP is localized to regions of the nervous system that are key players in CH pathophysiology, like the hypothalamus. Importantly, the posterior hypothalamus is activated during CH attacks.

The role of cytokines and inflammation, in CH is unclear, although they appear to involved, in some setting, when the trigeminovascular system is activated.

Differential Diagnosis - Despite the well-defined criteria, a “diagnostic delay” of several years may be seen for CH. The differential diagnosis of CH includes primary and secondary headache disorder. Cluster headache can also be misdiagnosed as migraine, as typical “migrainous

features" such as photophobia, phonophobia, facial allodynia, and nausea are seen in up to 50% of patients with CH. Other forms of TAC, particularly paroxysmal hemicranias can be mistaken as CH and sometime Indomethacin challenge is necessary to differentiates these two subtypes of TAC .

Trigeminal neuralgia is another main differential diagnosis for CH .

Between secondary headache disorder, headache attributed to paranasal sinus , particularly sphenoid sinusitis , are in differential diagnosis of CH .

Work up - For the initial diagnosis of patients with CH , neuroimaging with MRI (with and without contrast) or a non-contrast had been recommended .Sometime MRA also might be needed. One main reason to do imaging in all CH patients, is to rule out secondary CH.

Secondary CH- in patients with clinical attacks that resemble CH , presence of cranial lesions could be suggestive of secondary CH , although sometime although a causal relationship is often uncertain. Lesion that reported to cause headache similar to CH includes :

- Pituitary macroadenomas
- Intracranial large artery aneurysms
- Benign posterior fossa tumor
- Meningiomas
- Sinusitis , particularly sphenoidal sinusitis
- Brain arteriovenous malformations
- Recurrent nasopharyngeal carcinoma
- Cavernous hemangioma