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RESEARCH ARTICLE

PALATAL MYOCLONUS-A CASE STUDY

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Abstract

Palatal myoclonus is a repetitive, involuntary jerking movement of the pterygopalatine arch and soft palate, frequently involving the diaphragm and vocal muscles. Palatal tremor is classified into essential palatal myoclonus (EPM) and symptomatic palatal myoclonus (SPM). In EPM, the only clinical signs are palatal tremor and ear clicks, whereas, in SPM, tremor may encompass facial, ocular, and extremities muscles. In our study, a healthy 26 years old man presented a persistent auditory clicking on the temporomandibular joint bilaterally 18 months ago. The clicking was not related to mandibular movement. The final clinical examination showed that the patient suffered from palatal myoclonus with EPM. EPM is a rare condition characterized by clicking noises and muscle contraction at the back of the throat. Due to the condition's rarity, there have yet to be any randomized controlled trials of treatment options for EPM. However, our patient showed positive results after one week of treatment using Carbendazim 200mg.

Introduction:-

A rare movement disease called palatal myoclonus causes repetitive, short motions of the soft palate. Spencer first characterized it in 1886. Initially known as "palatal myoclonus," it was renamed "palatal tremor" at the First International Congress of Movement Disorders in 1990. The mix of myoclonus and tremor characteristics in the palatal movements makes phenomenological characterization challenging. Compared to myoclonus, the movements are less "shock-like" and more continuous and persistent [1]. The tremor's frequency varies greatly amongst patients and can even change within a single person, highlighting the unusual characteristics of the tremor. Palatal tremor is classified into essential palatal myoclonus (EPM) and symptomatic palatal myoclonus (SPM). In EPM, the only clinical signs are palatal tremor and ear clicks, whereas, in SPM, tremor may encompass facial, ocular, and extremities muscles. Typically, ear clicks are absent. While the underlying reason for SPMmay frequently be identified in the dentate-olivarytractus and results in olivarypseudohypertrophy, the underlying cause of EPM is still mostly unknown. Early accounts of this hypertrophy extend back to Thomas, Marie, and Guillain in 1887 and van Bogaert and Bertrand in 1928.

Case presentation:

A healthy 26 years old man presented to the family medicine clinic in AlmadinahAlmunawarah, Saudi Arabia persistent auditory clicking on the temporomandibular joint bilaterally for 18 months ago. The clicking was not related to mandibular movement.Hearing loss, salivary discharge, and tinnitus are typical in patients with TMJ herniation[2]. However, he denied vertigo, hearing loss, and otalgia. He had no history of headaches, weakness,or dysphagia weakness in any part of his body. He visited many clinics in Sudan and was diagnosed with temporomandibular joint disorders by a dentist. Temporomandibular joint (TMJ) disorders impact the jaw joints and

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the muscles and ligaments around these joints. Trauma, an erroneous bite, arthritis, or general wear and tear are all potential causes. Jaw soreness, headaches, ear infections, and facial pain are typical symptoms of temporomandibular joint disorders[3]. However,these symptoms were not observed by the patient. He was treated by intra-articular platelet-rich plasma injection in the TMJ without any improvement.

Another dentist diagnosed lower teeth crowding and treated them by molar extraction without any improvement. Further, he presented to the ENT clinic, and an ear examination was done. During an ear examination, an ENT specialist will pump a puff of airflow into your eardrum using a pneumatic otoscope, a device with a plastic bulb on the end. Your eardrum will shift; as a result, making it easier for your pain specialist to examine for inflammation and edema behind your eardrum [4]. An ear examination was performed in ENT but without a clear diagnosis.

Then his physical examination of the oral cavity was done. The oral examination includes a full dental and medical history along with a consistent and uniform examination of the head, neck, and intraoral assessment of the soft and hard tissues by using a tongue depressor [5]. Oral examination revealed rhythmic contraction of the tongue's soft palate, uvula, and base, and the clicking sound was audible. No other abnormality was found, like cardiovascular disease, respiratory disease, CNS disease, gastrointestinal disease, etc.

He was further diagnosed with essential palatal myoclonus, and carbendazim 200 OD at night was prescript for him for one week. The first follow-up was after one week, and there was reduced in his palatal movement, and he wasfeeling better. Then carbendazim dose was increased to 400 OD at night to see further improvement.

Results And Discussions:-

A repetitive, involuntary jerking movement of the pterygopalatine arch and soft palate, known as palatal myoclonus, frequently involves the diaphragm and vocal muscles. The development of palatal myoclonus occurs months after the acute phases of stroke. Frequently, repetitive, jerky movements are also noted in the face, eyeballs, tongue, mouth, vocal cord, or extremities (particularly hands); they might not be synchronized with palatal motions. The palate's rate varies from 40 to 200 beats per minute. The Eustachian tube may be involved in the movements, producing a clicking that the patient could hear[6]. The Eustachian tube can move and produce an audible clicking sound that the patient may report hearing or the examiner may hear by placing a stethoscope on the lateral neck. Chest fluoroscopy usually makes it easy to see the pharyngeal movements, which are frequently accompanied by a flapping of the diaphragm. Surprisingly, swallowing is not much impacted by palatal myoclonus [7].

The two subtypes of PM are essential (EPM) and symptomatic (SPM). EPM describes an objective clicking secondary to rhythmic tensor velipalatini (TVP) motions that frequently happen throughout the day but not while you are sleeping [8]. According to earlier research, EPM often manifests in late childhood and does not involve any brainstem or cerebellar lesions[1], [9].The Guillain-Mollaret triangle, which consists of the dentate, red, and inferior

olivary nuclei, is the typical site of a lesion in SPM, which affects the levatorveli palatine muscles. Contrary to EPM, SPM can result in sleep-related symptoms. Hypertrophic degeneration of the inferior olive is the pathologic lesion most frequently present in these patients[10]. Injury to the central tegmental tract in pontine stroke patients, which leads to the inferior olive's hypertrophic degeneration, is thought to be a contributing factor to palatal myoclonus. Infarcts are less likely than pontine hemorrhages to cause the palatal myoclonus.Most of the time, lesions in the brain's nerve pathways cause palatal myoclonus. The most frequent cause is a stroke. The issue typically arises a year or so after the stroke. This issue can also be caused by other illnesses such as multiple sclerosis, trauma, or tumors.

Most signs and symptoms in the patients include auditory clicking, bleeding from the ear, and normal hearing causes fluttering noises in the ears. Adults are most commonly affected by the illness, which can continue indefinitely. Even 7-year-old youngsters have been reported to have it.Arteriovenous malformations and venous hums are two vascular conditions that can be the cause of the objective [11].It can be a physiological event that happens as you sleep. Up to 150 contractions can occur in a minute, a very high rate. The jerking often occurs between 1-2 Hz. The levatorveli palatine muscle may contract and change the pressure inside the eustachian tube, which has been linked to vibrations in the tympanic membrane [12]. It must be distinguished from the middle ear myoclonus when the middle ear's tensor tympani and stapedius muscles are involved, which is observed as an oscillating tympanic membrane on otoscopic evaluation and confirmed by tympanometry [13].

Our findings showed that our patient suffers from an unclear etiology of essential palatal myoclonus(EPM). Still, the auditory click may occur in the essential palatal myoclonus, as it was also observed in our patient.

Treatment for PM is challenging. Due to the condition's rarity, there have yet to be any randomized controlled trials of treatment options. First-line PM medications include phenytoin, carbamazepine, clonazepam, carbendazim , diazepam, trihexyphenidyl, and baclofen[14]. 5-HTP and presynaptic antidopaminergic medications like tetrabenazine are examples of second-line medications. PM is said to benefit from botulinum toxin injections into the tensor veli palatine muscle[15].In this patient, we tried Carbendazim 200mg OD at night for one week, which showed reduced palatal movement. Then the dose was titrated to 400mg to observe further improvements.

Conclusion:-

Palatal myoclonus with EPM is a serious complication. Palatal myoclonus is a rare disease affecting the mouth's soft palate muscles. It is characterized by clicking noises and muscle contraction at the back of the throat, but the underlying cause of EPM is still mostly unknown. Due to the condition's rarity, its treatment is challenging. However, our patient showed positive results after one week of using Carbendazim 200mg.

References:-

[1] E. M. J. Foncke, “Palatal Myoclonus,” in Encyclopedia of Movement Disorders, K. Kompoliti and L. V. Metman, Eds. Oxford: Academic Press, 2010, pp. 358–360. doi: 10.1016/B978-0-12-374105-9.00051-4.
[2] K. H. Lim, J. Y. Jung, J. Rhee, and J. Choi, “Temporomandibular joint herniation through the foramen of Huschke with clicking tinnitus,” Eur. Ann. Otorhinolaryngol. Head Neck Dis., vol. 136, no. 6, pp. 497–499, Nov. 2019, doi: 10.1016/j.anorl.2018.05.014.
[3] W. E. Contributors, “Temporomandibular Disorders (TMJ & TMD),” WebMD. https://www.webmd.com/oral-health/guide/temporomandibular-disorders-tmd (accessed Jan. 21, 2023).
[4] C. J. Hogan and P. Tadi, “Ear Examination,” in StatPearls, Treasure Island (FL): StatPearls Publishing, 2022. Accessed: Jan. 21, 2023. [Online]. Available: http://www.ncbi.nlm.nih.gov/books/NBK556014/
[5] “Oral Examination: Overview, Physical Examination, Laboratory Studies,” Jan. 2023, Accessed: Jan. 21, 2023. [Online]. Available: https://emedicine.medscape.com/article/1080850-overview
[6] J. S. Kim and L. R. Caplan, “26 - Vertebrobasilar Disease,” in Stroke (Seventh Edition), J. C. Grotta, G. W. Albers, J. P. Broderick, A. L. Day, S. E. Kasner, E. H. Lo, R. L. Sacco, and L. K. S. Wong, Eds. Philadelphia: Elsevier, 2022, pp. 368-403.e7. doi: 10.1016/B978-0-323-69424-7.00026-0.
[7] J. P. Mohr and L. R. Caplan, “26 - Vertebrobasilar Disease,” in Stroke (Fifth Edition), J. P. Mohr, P. A. Wolf, J. C. Grotta, M. A. Moskowitz, M. R. Mayberg, and R. von Kummer, Eds. Saint Louis: W.B. Saunders, 2011, pp. 446–484. doi: 10.1016/B978-1-4160-5478-8.10026-0.
[8] J. M. S. Pearce, “Palatal Myoclonus (syn. Palatal Tremor),” Eur. Neurol., vol. 60, no. 6, pp. 312–315, 2008, doi: 10.1159/000159929.

[9] R. H. Schwartz, R. S. Bahadori, and J. S. Myseros, "Loud Clicking Sounds Associated With Rapid Soft Palate Muscle Contractions," Pediatr. Emerg. Care, vol. 28, no. 2, p. 158, Feb. 2012, doi: 10.1097/PEC.0b013e3182443000.
[10] D. G, T. C, and H. M, "Symptomatic and essential palatal tremor. 2. Differences of palatal movements," Mov. Disord. Off. J. Mov. Disord. Soc., vol. 9, no. 6, Nov. 1994, doi: 10.1002/mds.870090615.
[11] A. Sismanis, "Pulsatile tinnitus. A 15-year experience," Am. J. Otol., vol. 19, no. 4, pp. 472–477, Jul. 1998.
[12] D. R. Jamieson, C. Mann, B. O'Reilly, and A. M. Thomas, "Ear clicks in palatal tremor caused by activity of the levator veli palatini," Neurology, vol. 46, no. 4, pp. 1168–1169, Apr. 1996, doi: 10.1212/wnl.46.4.1168.
[13] T. E. Zipfel, S. R. Kaza, and J. S. Greene, "Middle-ear myoclonus," J. Laryngol. Otol., vol. 114, no. 3, pp. 207–209, Mar. 2000, doi: 10.1258/0022215001905120.
[14] S.-N. Park, K. H. Park, D. H. Kim, and S. W. Yeo, "Palatal Myoclonus Associated with Orofacial Buccal Dystonia," Clin. Exp. Otorhinolaryngol., vol. 5, no. 1, pp. 44–48, Mar. 2012, doi: 10.3342/ceo.2012.5.1.44.
[15] E. Krause, A. Leunig, T. Klopstock, and R. Gürkov, "Treatment of essential palatal myoclonus in a 10-year-old girl with botulinum neurotoxin," Otol. Neurotol. Off. Publ. Am. Otol. Soc. Am. Neurotol. Soc. Eur. Acad. Otol. Neurotol., vol. 27, no. 5, pp. 672–675, Aug. 2006, doi: 10.1097/01.mao.0000224085.08344.50.