Nummular Headache - A case report of a rare entity

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Case report

Keywords: Nummular headache, Rare primary headaches, Coin-shaped headache, Chronic Primary headache, Epicranial headache

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Abstract

Background: Nummular Headache is a rare type of chronic primary headache wherein the pain is usually located in a well-circumscribed, round or elliptical shaped area of the scalp.

Case Report: A 26-year-old female presented to the Center for TMD/ OFP with the chief complaint of chronic persistent left parietal headache, for more than 10 years, which has been worsening over the past few days with no specific identifiable trigger. The medical history is non-contributory. The primary diagnosis is Nummular Headache.

Conclusion: A comprehensive history is the first step to achieve an accurate diagnosis. The diagnosis of Nummular Headache is challenging and confusing due to the rare occurrence. An examination must include imaging and is detrimental in eliminating other underlying pathologies. The diagnosis is made from distinct clinical features after exclusion of all other entities. **Keywords:** Nummular headache, Rare primary headaches, Coin-shaped headache, Chronic Primary headache, Epicranial headache.

Introduction

The term nummular is derived from the Latin word, nummus and/or nummular, which translates into English is as resembling a coin. Nummular headache (NH), previously referred to as coin shaped headache, was first reported in Cephalalgia in 2002 (Pareja, Caminero et al. 2002). NH is considered a rare headache, with an unknown prevalence and incidence (Grosberg, Solomon et al. 2007). This Case-Report of a 26-year-old female patient demonstrates a step by step approach for the diagnosis of NH in accordance with the International Classification of Headache Disorder (ICHD) – 3 of the International Headache Society (IHS) (2018) and its subsequent treatment.

Case Report

A 26-year-old South Asian female presented to the Center for Temporomandibular Disorders and Orofacial Pain with the chief complaint of chronic persistent pain of the left parietal region of the scalp. The pain was present for more than 10 years. It was increasing both in frequency and duration for several days prior to the first visit. No associated events or precipitating factors were identified. This was also true for the initial onset of the pain or other exacerbations of painful episodes over the past 10 years.

Upon examination, a well-circumscribed 3–4 cm wide in diameter area of pain and discomfort was localized in the left parietal region. The patient reported allodynia and dysesthesia in the same area. Symptoms occasionally switched sides to the right side. When it changed sides, the non-affected side was completely normal. The frequency is daily and continuous. The temporal pattern was chronic progressive. The background pain was of a continuous nature with paroxysmal exacerbations in intensity. The quality of continuous background pain was dull and annoying with intensity on a Numerical Pain Scale was 4 /10. The quality of the episodic exacerbations was pressure-like and sharp with the intensity of pain on a Numerical Pain Scale was 8 /10.

When severe, which was often, pain was nearly debilitating and radiated towards the left supra-orbital region. No aggravating factors were reported. There was no specific pattern to the paroxysmal exacerbations. Alleviating factors reportedly included rest and sleeping. There was no reported alleviation of pain upon taking over the counter Non-steroidal Anti-inflammatory Drugs. There were no associated features such as tearing, eye irritation or edema with the exacerbation of pain. The patient reports that the pain neither woke her up nor did she report routinely waking up with the pain. The medical history of the patient was non-contributory. The only positive finding in history was that of a migraine without aura for almost 12 years of age. The patient was able to distinctly differentiate between migraine pain and the current presenting pain symptoms.

The past investigations included only an MRI scan of the brain without contrast, ordered one year earlier. There were no reported abnormalities in the MRI and images were irretrievable. The patient was not prescribed any medications thus far, for this headache. Following the IHS, ICHD-3 criteria, the differential diagnosis included primary idiopathic stabbing headache, underlying epicranial anomalies, epicranial headaches, metastatic lesions of the cranial bones, infectious lesions of the cranial bones and nummular headache. Complete blood work including CBC with differential, clotting factors, comprehensive metabolic panel, vitamin panel including vitamin A, B3, B6, B12, C, D3, folate, SSA, SSB, ANA, RF, and thyroid panel was ordered. All these results were within normal limits. A thorough neurologic examination was also performed which failed to show any related abnormalities.

A new MRI of the brain and brainstem, with and without contrast, failed to reveal any pathology. However, there was an incidental finding of a well-circumscribed radiolucency in the left sphenoid sinus. A subsequent CT scan confirmed this finding. Upon consultation with an ENT surgeon, any association of a headache with the pathology was deemed questionable.

Nonetheless, surgical intervention in the sphenoid sinus was performed, resulting in the removal of a 'fungal ball'. There was no change in the headache pattern. Owing to the specific clinical features, the possibility of IHS, ICHD-3 A) 9.1.3 Headache attributed to intracranial fungal or other parasitic infection in the cranium and A) 9.1.3.3 Persistent Headache attributed to past intracranial fungal or other parasitic infection were ruled out. Thus, based on the patient's history, clinical findings and diagnostic imaging, a final diagnosis of 'Nummular Headache' was made.

A trial of Gabapentin titrated up to 900 mg per day but failed to achieve adequate pain relief. Patient voluntarily stopped the medication due to significant reported side-effects such as nausea, vomiting, fatigue, excessive sleepiness, inability to perform daily activity adequately, and cognitive deficits. Currently, the patient is reportedly tolerating the headache with pain intensity 2/10 on a Numerical Pain Scale with NSAIDS, and non-pharmacological approaches including cognitive behavioral therapy, yoga, and meditation.

Discussion

A very low incidence of Nummular headache has been reported in the literature (Pareja, Pareja et al. 2004). The prevalence reported in the limited literature is 1.25% of all headache patients (Guerrero, Martin-Polo et al. 2008). Some studies have reported the prevalence approaching 5% (Guerrero, Cortijo et al. 2012). Approximately only 200 cases have been reported, in the literature, through 2013 (Cortijo, Guerrero-Peral et al. 2011).

Nummular headache is usually described as a sharply contoured, fixed area in size; round or elliptical shaped; 2–6 cm in diameter, usually present in the parietal region of the scalp. In some cases, it might also be present in the occipital, frontal, vertex, temporal region or multiple sites (Schwartz, Robbins et al. 2013). The position of the foci of the pain may change and when it does the non-affected side(s) may or may not be normal (6), ((Cuadrado, Valle et al. 2009), (Chiu-Hsien, Tzu-Hui et al. 2013). The intensity of the pain is usually described as mild to moderate. On occasions, it can be severe. A continuous background pain with occasional exacerbations of severe pain has been reported (1), (4). Up to 75% of published cases have described the duration of a headache as highly variable and generally chronic. Cases have also been described with durations lasting from minutes to hours (3). The affected area commonly may show a variety of characteristics including allodynia, hyperesthesia, dysesthesia, paresthesia, and/or tenderness (3). Usually, with NH, there are no associated features like nausea, vomiting, light or sound sensitivity. Moreover, there are no autonomic features such as rhinorrhea, lacrimation and conjunctival injection. Other types of more common primary headaches, including migraines, can be a co-morbidity with NH (Moon, Ahmed et al. 2010).

The differential diagnosis should include primary idiopathic stabbing headache (1), underlying epicranial anomalies (Lopez-Ruiz, Cuadrado et al. 2014), epicranial headaches (12), metastatic lesions of the cranial bones (1), infectious lesions of the cranial bones.

The pathophysiology of NH is uncertain. Literature suggests that NH may be neuralgia of a terminal branch of the trigeminal nerve (Evans and Pareja 2005). Some studies suggest that NH might be a non-generalized disorder with sensitization restricted to the painful area (Ruscheweyh, Buchheister et al. 2010). There are some reported cases, which suggest the presence of underlying aneurysms of the superficial arteries of the scalp, which can also produce secondary NH (12). Presence of autoimmune biomarkers has been found in patients with primary NH (Chen, Chen et al. 2012). When primary headaches occur co-morbid with NH, the temporal and spatial characteristics have been reported to be independent of the NH pain (1). There are no clear guidelines or directions for treating NH patients (2). Moderately successful management using NSAIDs/ indomethacin have been reported (2), (Dach, Speciali et al. 2006). For preventive therapy, the first choice is usually Gabapentin with slow titration (2), (13). Other approaches like local nerve blocks, botulinum toxin injections, tricyclic anti-depressants, and lamotrigine have also been tried.

A comprehensive history is the first step to achieve an accurate diagnosis. Successful management of this condition is difficult. The diagnosis of Nummular Headache is challenging and confusing even for a seasoned clinician due to the rare occurrence and hence limited knowledge of this disorder. An

examination must include imaging and is quintessential in eliminating other underlying pathologies. The diagnosis is made from distinct clinical features after exclusion of all other entities.

Clinical Implications

The clinical implications of this Case Report are as follows:

1. Nummular Headache is a diagnosis by exclusion, predominantly by clinical presentation.
2. Successful therapy of Nummular Headache is difficult, and complete elimination of pain might not be achieved.
3. Along with pharmacotherapy, adjunctive analgesic medications, and other modalities should be considered.

Conclusion

A comprehensive history is the first step to achieve an accurate diagnosis. Successful management of this condition is difficult. The diagnosis of Nummular Headache is challenging and confusing even for a seasoned clinician due to the rare occurrence and hence limited knowledge of this disorder. An examination must include imaging and is quintessential in eliminating other underlying pathologies. The diagnosis is made from distinct clinical features after exclusion of all other entities.

Declarations

Informed Consent:

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of Interests:

The authors declared no potential conflicts of interest with respect to the research authorship, and/or publication of this article.

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