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Does Trochlear migraine exist? A"pure" pediatric case report and nosographic considerations

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Abstract

Background: Recently Trochlear Migraine has been described as the concurrence of strictly unilateral migraine and ipsilateral trochleodynia with migraine relief after successful treatment of trochleodynia and this disorder has been interpreted similar to "Cluster-tic syndrome" or "seizure-triggered migraine".

Case: We report the case of a young boy that complained recurrent attacks of severe, pulsating headache associated to nausea, vomiting, phonophobia and photophobia with strict trochlear localization of pain. The attacks were alternating in side.

Conclusion: The previous rare Trochlear Migraine reports describe the association of two concurrence painful disorders where one influences the other. This is the first case report showing a typical migrainous phenotype strictly localized in trochlear region. To our opinion the term "Trochlear Migraine" should be reserved to a clinical migrainous syndrome strictly localized in trochlear region.

Keywords: Trochlear region, Migraine, Primary trochlear headache, Pediatric headaches, International headache classification

Abbreviations: PTH: Primary Trochlear Headache; TM: Trochlear Migraine; HIS: International Headache Classification

Introduction

Recently, some authors [1-3] have introduced two new primary headaches types: "Primary Trochlear Headache (PTH) and Trochlear Migraine (TM)". The first type describes a pain developing from trochlear region, presenting with pressure-like quality, moderate intensity, almost always continuous, exacerbating by supraduction of the symptomatic eye, relieved by local steroid inoculation, in absence of inflammatory signs and orbital or systemic disease, often coexisting with other primary headaches, mainly migraine. The second type defines a "concurrence of strictly unilateral migraine and ipsilateral trochleodynia in same side, relief of migraine is absolutely dependent on successful treatment of trochleodynia" [3]. In this report the authors interpreted the second type similar to "Cluster-tic syndrome" (co-occurence of two different disorders) or "seizure-triggered migraine" because migraine improvement follows to the treatment of trochleodynia and migrainous attacks are triggered by trochleodynia (causal relationship between two disorders) [3]. We propose instead that the definiton "pure trochlear migraine" should be reserved to a clinical migrainous syndrome strictly localized in trochlear region, similar to the "nasal migraine" used by the same group to describe migrainous attacks strictly localized in the nasal region [4].

After the first descriptions [1-3], few cases of both types have been reported [5,6]. There are not "pure trochlear migraine" description and no report of pediatric case. Here, for the first time, we describe a pediatric headache case report with typical migrainous phenotype and strict trochlear localization.

Case Report

A 12-year old male, affected also by type 1 diabetes, was admitted to the Pediatric Hospital of "ARNAS", Palermo, Italy on April 2017 for a clinical picture of severe left infraorbital pain, nausea and vomiting (five episodes in same day). He was admitted to Pediatric Diabetology Unit because he was followed for diabetes mellitus type 1 from October 2015, however the blood examination did at the admission showed only slight increase of glycemic rate (179 mg/dL) and Ketones (1,9 mmol/L). The patient underwent a hydration treatment i.v. (infusion of mixed glucose/physiological solution) and showed a gradual recovery from pain and vomiting. After 2 days, when completely asymptomatic, the child was sent to our department for a neurological consult. He reported an history of episodic headache from more than 1 year, usually at frequency of 1-2

attacks/monthly, with the following features: gradual onset, pressure-like or pulsating quality, severe intensity, duration of some hours, alternating side (prevalent to left, association to nausea, photophobia, phonophobia and more rarely episodes of vomiting (usually isolated). The attacks were attenuated by rest or sleep, and there were no report of aura, diplopia, cranial autonomic symptoms and specific triggers. Main atypical characteristic of these attacks was the pain location strictly limited to the unilateral superior –inner angle of orbit. The pain rarely had irradiation on same fronto-temporal side and had selective tenderness to pressure over infratrochlear zone without additional periorbital tenderness neither other sensitivity disorders. No swollen trochlea may be felt upon palpation. The child was not able to refer presence of pain with ocular movements because of severe pain. During the attacks the glycemic levels measured by the parents were not higher or lower than outside attacks.

Remote pathological history reported only diabetes mellitus and a positive familiarity for migraine (mother). Neurological examination was normal. The patient underwent also a complete blood examination (including also antibodies against gluten and thyroglobulin, protein C reactive etc.), an ophthalmologic consult, EEG and Magnetic Resonance Imaging with contrast medium and focused particularly on orbital cavities. All exams resulted negative. At the 6 months follow-up visit patient reported 5 attacks with similar features, attenuated by quick administration of analgesic drugs (paracetamol or ibuprofen).

Discussion

Pareja *et al.* [3] coined the term "Trochlear Migraine" referring to concurrence of unilateral trochlear pain and ipsilateral migraine attacks where the appearance of the first worsened the second pain and the relief of the first improved the migrainous attacks. Since there, only few reports have been published both on PTH and TM (where trochlear pain triggered more migrainous attacks in subjects with previous migraine syndrome) and no pediatric cases among them. Furthermore, the same and other group [3,7] have described subjects where the PTH influenced tension headache or trigeminal autonomic cephalalgias, so reducing the association specificity between PTH and Migraine. To our opinion, the term "trochlear migraine" seems to be inappropriate for this relationship, and thus it should be eventually attributed to a disorder with clinical migraine where pain is strictly localized in trochlear region without no secondary etiologies. Our patient seems to be the first "pure" trochlear migraine case report as the painful attacks raise strictly in unilateral infratrochlear region with severe soreness to pressure and associated to the typical migrainous features (recurrent episodic attacks, alternating side, severe intensity, pulsating pain, duration of attacks, vegetative associated symptoms). Furthermore the patient showed migraine familiarity and pain was relieved by rest and sleep (**Table 1**: Trochlear Migraine case summary in comparison with our patient) The Pareja's criterion [3] of pain reduction by local steroid injection cannot be satisfied as the episodic pain pattern made unnecessary this procedure; furthermore this criterion was not always met in another clinical report [6] (no effect in 26,7%) and same Spanish group recently affirmed the non-specific response of this treatment in the primary headaches [8]. Primary trochlear headache (PTH) was excluded in our case, because differently from PTH, pain side was alternating and not fixed, the temporal pattern was with recurrent episodic attacks and not continuous. Moreover, patient presented important vegetative symptoms associated to headache, very rarely described in PTH. However, our case shared some features with PTH: the side of pain and the soreness to local pressure in infrathroclear region, although the presence of painful ocular movements was not possible to investigate (however only 50% had this symptom in a recent case-collection [9]). Furthermore, other primary headaches and neuralgias, similar for size of pain or for local site, like nummular headache and supraorbital neuralgias, could easily be excluded for temporal pattern (episodic versus continuous or intermittent-remittent), alternating shift versus fixed side, quality of pain, absence of vegetative signs etc [10,11]. In our case, secondary causes (trochleitis) were ruled out by the temporal episodic and alternating pattern, the negative physical and neurological examination, blood tests, ophthalmologic consult and the neuroradiological imaging.

However, the real question is if does it exist a "Trochlear Migraine Syndrome"? In our opinion such term should instead be reserved to migrainous attacks strictly localized in trochlear region. Until now in literature, to our knowledge, outside of these studies that define *inappropriately* the TM, a such strictly unilateral infratrochlear localization has not ever been reported, neither incidentally. We initially hypothesized that such specific localization, in our case, could follow to diabetes with typical painful sensitive disorders and proneness of ocular nerves to diabetic neuropathy. Therefore, that may let the migraine predisposition of our patient (see familiarity) to emerge favoring also the regional localization of pain. However, diabetic neuropathy generally arises later in the course of the illness, since the recent diabetes onset we considered this initial hypothesis unlikely. In addition, the epidemiological evidence [12] of an inverse relationship between diabetes and migraine seems to further reduces the chance of a possible pathogenetic role of the diabetes in the headache disturbance of our patient. Finally in several series of painful oculomotor neuropathies the involvement of the IVth nerve is rare and rarely presents with pain [13]. A study [14] suggested that a large group of unilateral migraine patients (80%) perceived a referred pain from stimulation of the trochlear region by involvement of local myofascial structures during free headache interval. They speculated that a trochlear generator may participate in origin, maintenance or perpetuation of migraine. However, this feature is also present in primary headaches and doesn't explain the rarity of migraine starting from this region. It is possible that this localization is less rare than reported: if not adequately questioned, similar to other syndromes (see red ear syndrome

Table 1. *Trochlear migraine – case reports in literature.*

Clinical features	Yanguela et al, Neurology 2002					Smith et al, Eur J Neurol 2014						Raieli et al
Sex	F	F	F	F	F	F	M	F	F	F	F	M
Age at diagnosis trochlear pain	?	?	?	?	?	55	29	34	18	46	35	12
Age at onset of migraine, y	8	26	15	25	17	&	&	&	&	&	&	10
Migraine subtype	Ch	Ep	Ep	Ch	Ch	?	?	?	?	?	?	Ep
Location of migraine pain	RH	LH	LH	R>L H	L>RH	?	?	?	?	?	?	Troch.
Age at onset trochlear pain	39	50	49	72	56	?	?	?	?	?	?	10
Side trochlear pain	U	U	U	U	U	U	B	B	U	B	U	A
Quality troclear pain	Puls.	Dull	Squez.	Dull	Sand	Pre.	Puls.	Puls.	?	Pre..	Pre..	Puls./pre
Temporal pattern of active pain period	Cont.	Cont.	Cont.	Cont.	Cont.	Cont	Cont	Cont	Cont	Cont	Cont	R
Intensity of trochlear pain (not included excerbations)	4	4-5	4-5	6	3-4	6	6	?	?	4	?	7-9
Photo/phonophobia associated to trochlear pain	-	-	-	-	-	+	+	+	?	+	+	+
Nausea associated to trochlear pain	-	-	-	-	-	+	+	+	?	-	-	+
Vomiting associated to trochlear pain	-	-	-	-	-	?	?	?	?	?	?	+
Diplopia	+	-	-	-	-	-	+	+	?	-	+	-
Trigger trochlear for migraine attacks	+	+	-	-	-	+	+	+	+	+	+	NA
Response to local steroid injection	+	?	+	+	+	+	+/-	-	+	+/-	+	NA

?, not specified; & , not specified but all have prior diagnosis of migraine before start of trochlear pain or of trochlear migraine following onset of the new headache; Ch, chronic migraine , Ep, episodic migraine; RH, right hemicranias ; Lh , left hemicranias ; Troch., trochlear region; U, unilateral; B, bilateral; A, alternating; Puls, pulsating pain; Dull, dull ache, Squez, squeezing pain; Sand , sand pain; Pre, pressure-like pain; Cont, continuous; R, recurrent attacks ; NA, not applicable.

[15]) it could be less described by patients and escape to the attention of not headache trained doctors. In fact, migraine pain mostly develops within the innervation territory of the trigeminal-nerve first branch and the topographical description of migraine is often undervalued. These considerations are supported by a recent interesting report describing 43 patients with trochlear pain where two subjects had alternating pain side and nausea and photophobia were complained respectively in seven and six cases [9]. These patients could resemble to our case, unfortunately the authors did not describe other aspects regarding these possible cases of "pure" TM. No pediatric case was reported [9].

Under this respect, a relevant question could be if the IHS classification of primary headaches [16], based on syndromic criteria, should allow the excessive increasing descriptions of new primary headache syndromes based on particular criteria. For instance, new syndromes are defined in relation to atypical topography, (see PTH or idiopathic rhinalgia) [1,17]; to the direction of pain, (see epicrania fugax [7]; to the size of painful area (see nummular headache) [8]; or to associated autonomic symptoms (see red ear syndrome [15]). Alternatively, a more restrictive approach could be chosen to better reallocating headaches in relation to well defined pathophysiological model of cranial pain, based on our actual scientific knowledge where the single case confirms or disconfirm the suggested model. If we consider this second hypothesis, it should be less useful to identify a disorder in relation to localization or other clinical features.

In conclusion, we here described the first case of "pure "Trochlear Migraine in pediatric age that fits perfectly to the diagnostic term coined by Spanish authors, questioning however about the utility of descriptions of new syndromes based for some particular features into a frame of pain matrix.

Declaration of conflicting interests

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