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CLUSTER HEADACHE ATTACKS IN A WOMAN PREVIOUSLY STRUCK BY LIGHTNING

Pathophysiology of the latent period

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ABSTRACT - *Background:* Cases of patients who developed cluster headache-like symptoms after different putative causes have been reported, indicating a direct relationship between brain lesion and this particular type of headache. Long term, delayed, neurological sequelae after lightning have also been described. *Case report:* We describe the case of a woman who, at the age of 10, was struck by lightning. Six years later she developed cluster headache-like attacks. *Conclusion:* We hypothesize that a relationship between the lightning and the cluster headache-like episodes observed in our patient. This case study may have helped throw some light into the still unknown pathophysiology of this particular type of primary headache.

KEY WORDS: cluster headache, lightning, pathophysiology, latent period.

Cefaléia em salvas em uma mulher que foi previamente atingida por raio: fisiopatologia do período latente

RESUMO - *Introdução:* Foram descritos casos de pacientes que desenvolveram cefaléia em salvas como manifestação secundária à diferentes causas, indicando uma relação direta entre uma lesão cerebral e este tipo particular de cefaléia. Seqüelas neurológicas tardias após injúria elétrica por raio também têm sido descritas. *Relato do caso:* Nós descrevemos o caso de uma mulher que aos 10 anos de idade foi atingida por um raio. Seis anos mais tarde desenvolveu sintomatologia de cefaléia em salvas. *Conclusão:* Acreditamos que pode haver relação entre a injúria elétrica causada pelo raio e o aparecimento dos episódios semelhantes à cefaléia em salvas observados na nossa paciente. A provável fisiopatologia envolvida na gênese da cefaléia em salvas de origem secundária é discutida, particularmente em relação ao intervalo latente entre a injúria precipitante inicial e a fase álgica da cefaléia.

PALAVRAS-CHAVE: cefaléia em salvas, raio, fisiopatologia, período latente.

Among natural disasters, lightning is a leading cause of morbidity and mortality throughout the world. Even so, the risk of being struck by lightning is extremely low and deaths from lightning injuries are infrequent (i.e., 0.2-0.8 per million per year)[1]. Every year in the USA, 100-150 people die and 1,000-1,500 others are injured by lightning strikes[2]. There is evidence that the cluster headache is a consequence of biochemical changes occurring in specific brain areas[3,4]. Cases of patients who developed cluster headache after different putative causes have been reported, such as dental extraction[5], wisdom tooth inflammation, vertebral artery injury[6], postmeningitic infection[7], head trauma[8], and sinusitis[9], suggesting that in a subgroup of patients (3-5%) the syndrome is secondary to a discrete central nervous system damage[10]. In this concern, the case of a 25-year-old male who developed cluster headache-like attacks in the course of an acute exacerbation of his multiple sclerosis has also been described, indicating a direct relationship between brain lesion and cluster headache[11]. Magnetic resonance imaging revealed a lesion in the area of the ipsilateral pontomedullary trigeminal nuclei[11]. Yet another major study shows that during cluster headache attacks, functional imaging with PET revealed activation of the ipsilateral posterior inferior hypothalamic gray substance, the contralateral ventroposterior thalamus, the anterior

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cingulate cortex, the ipsilateral basal ganglia, the right anterior frontal lobe, and both insulae[4]. Some of the activated areas described above are involved with the trigeminal-autonomic reflex[12], thus explaining the (a) hypofunction observed in the sympathetic system, (b) the hyperfunction of the parasympathetic system, and (c) the pain felt at the trigeminal territory.

We present a case of a woman who was struck by lightning and developed cluster headache-like symptoms with a view to discussing the pathogenesis of the disease.

CASE

A woman who, at the age of 10, took shelter beneath a tree together with her older sister and a donkey during a thunderstorm and was thrown to the ground after being struck by lightning. She remembered nothing of the accident but her sister reported that she had loss of conscience for more than 15 minutes. Her eyebrows, eyelashes, and hair were singed. She also presented burns in many parts of her body. The animal was killed by the lightning. On that occasion, no neurological deficit was detected. Six years later she developed cluster headache-like attacks. She reported a 2-months history of spontaneous intense attacks of headache in the left fronto-orbital and temporal regions, several (2-3) during the day, with 30-60 min duration. Concomitantly with the headache attacks appeared a left eye lacrimation with conjunctival injection. The CT scan of the head was normal.

DISCUSSION

Most interestingly is the fact that long term, delayed, neurological sequelae after lightning have been described. Motor neuron disease after electric injuries, including lightning, can occur 10 days to 18 years after the event[13]. Direct strikes to the head might result in petechiae or larger brain hemorrhages[14]. Demyelination and delayed myelopathy may occur as well[15,16]. Since lateralized abnormalities in the diencephalic-brainstem region had been detected in the brain of patients with cluster headache[3,4,11], we hypothesize that the lightning would have induced progressive neuronal changes that would trigger cluster headache-like attacks a few years later through a mechanism similar to the one observed with motor neurons. No doubt this is purely a speculative hypothesis. However, it is likely that this association - lightning/trauma and cluster headache - is not an incidental coincidence. No such cases have been reported until now possibly for two reasons: a) because of the rarity of cases of lightning survivors or b) due to a delay in the pain onset mechanism which could justify a lack of immediate cause-effect direct relation. The latter may explain the skepticism of the experts regarding the association between injury and delayed cluster headache attacks. Manzoni et al.[8] reported that 41 out of 180 patients with cluster headache had previous head injury, with loss of consciousness occurring in 20. Interestingly, it was noted a close correspondence between the region of the head injury and the side on which cluster headache later occurred, with a mean latency of nine years. Recently we had the opportunity to interview a 32-year-old woman with cluster headache who had been suffer a severe electrical chock 11 years ago, again suggesting a close relationship between this particular type of headache and the electrical event.

Another point to be discussed is that the cluster headache is less frequent in women and the onset of the cluster headache in our patient occurred at a young age (16-year-old). Ekbom et al.[17], studying 554 patients with cluster headache, observed that in their patients, of both sexes with the episodic form, the age of onset occurred when they were in their 20s. But, in women a second peak of onset occurred in their 50s. Farias da Silva et al.[18] reported that 81.8% of their patients with cluster headache presented the onset after the second decade. During the period of 1963 and 1997 there was a trend towards a decreasing male preponderance, but still there is a male preponderance of 3-2:1[17,18]. Recently, we reviewed our 18 cases of cluster headache with a probable secondary origin and the gender distribution was 13 men:5 women, still with a male prevalence in secondary cases of cluster headache[19].

Nevertheless, it is worth pointing out that the episodic cluster headache is known as a cyclical neurological disease with two stages: the cluster attacks period and a pain-free period. Indeed, clinical evidence has revealed that the disease may be still active in the latter period but in the absence of headaches, since ocular sympathetic impairment (e.g., ptosis and miosis) may persist during the remission phases. Micieli et al.[20] studied the pupil diameter induced by different stimuli in cluster headache patients and concluded that a dysfunction of the integrative central nervous system pathways exists intercritically, involving both autonomic regulation and pain perception mechanism. Moreover, the essential unilateral characteristic of the pain attacks observed in the majority of the patients (about 90%) with episodic cluster headache[18] is yet another argument that supports permanent biochemical brain damage asymmetrically localized in the central nervous system.

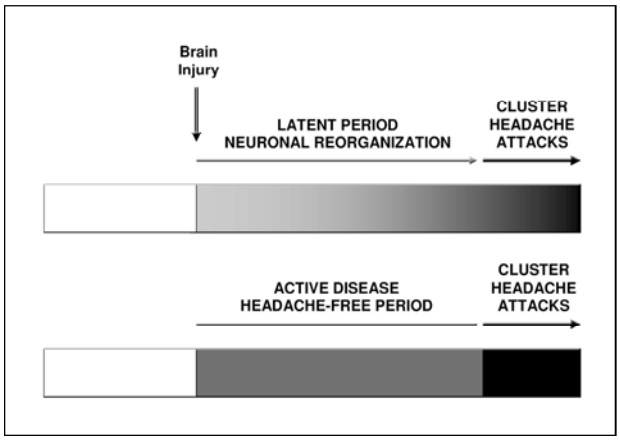

Figure. Possible pathophysiological hypothesis to explain a latent period between the initial precipitant injury and the beginning of the cluster headache attacks.

The Figure illustrates two possible physiopathogenic mechanisms in an attempt to explain a latent period between the initial precipitant injury (IPI) and the beginning of the cluster headache attacks. In the first possibility, the IPI immediately triggers the episodic cluster headache entity but in the headache-free period, taking into consideration that the disease has two distinct active phases. Or the long latent period is secondary to physicochemical changes, provoked by the IPI. And, as a result, anatomo-pathologic neuronal reorganization, or evolution to a permanent dysfunctional state of specific brain areas, occurs. Similar phenomenon is also observed in mesial temporal lobe epilepsy, in which an IPI (i.e., febrile seizure) occurs about 8-10 years before the beginning of the usual epileptic seizures[21]. Although, in the secondary cases of cluster headache is difficult to prove a cause-and-effect relationship, as was described by Dodick et al.[22].

In conclusion, based on the argumentation as well as on the evidence so far presented, we hypothesize an association between the lightning/brain trauma and the cluster headache-like episodes observed in our patient. This case report may add to previous information and help to clarify the still unknown pathophysiology of this particular type of primary headache.

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