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THUNDERCLAP HEADACHE AND REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY: CASE REPORT

INNOCENZO LUPO - VALENTINA SAIA - ENRICO CAMMARATA - GIUSEPPE GIGLIA - ANTONIO PALERMO - PAOLA MANGIAPANE
University of Palermo - Department of Neurology, Ophthalmology, Otorhinolaryngology and Psychiatry
(Direttore: Prof. G. Savattieri)

[Cefalea thunderclap e leucoencefalopatia posteriore reversibile: descrizione di un caso]

RIASSUNTO

Reversible posterior leukoencephalopathy syndrome (RPLS) is a clinical radiological syndrome, that is generally associated to various conditions: hypertensive encephalopathy, cronic renal failure, puerperal eclampsia, blood transfusion, immunosuppressant drugs, systemic lupus, vasculitis, acute phases of autoimmune diseases, and many other causes.

Recently it has been highlighted a clinical association between thunderclap headache and posterior reversible leukoencephalopathy syndrome.

In this study a new case of association between thunderclap headache and posterior reversible leukoencephalopathy syndrome is described and the possible etiopathogenetic mechanism is showed.

Key words: thunderclap headache, reversible posterior leukoencephalopathy syndrome, vasogenic oedema, MRI

SUMMARY

La leucoencefalopatia posteriore reversibile (RPLS) è una sindrome clinico-radiologica che può essere associata a numerose condizioni: l'encefalopatia ipertensiva, l'insufficien - za renale cronica, l'eclampsia gravidica, la trasfusione di san - gue, il trattamento con farmaci immunosoppressori, il lupus sistemico, le vasculiti, le fasi acute di malattie autoimmuni, ed altre cause ancora. Recentemente uno studio ha messo in evi - denza una associazione clinica tra la cefalea "thunderclap" e la leucoencefalopatia posteriore reversibile. In questo studio viene presentato un nuovo caso di associazione tra cefalea thunderclap e leucoencefalopatia posteriore reversibile e viene descritta la possibile connessione etiopatogenetica tra le due patologie.

Parole chiave: *cefalea thunderclap, leucoencefalopatia poste - riore reversibile, edema vasogenico, RM*

Introduzione

Thunderclap headache (TCH) refers to headache of instantaneous onset, with severe, excruciating pain.

The headache which reaches maximum intensity within 30 seconds, usually can last up several hours and episodes of TCH may occur repeatedly over a 7- to 14-day period and may recur over subsequent months to years[1]. Idiopathic TCH is associated with diffuse vasospasm and may produce fluctuating focal neurologic signs and symptoms.

Reversible posterior leucoencephalopathy sindrome (RPLS) is a clinical radiological syndrome, first described by Hinchey et al. in 1996, that can be associated with several conditions, including hypertensive encephalopathy, cronic renal insufficiency, puerperal eclampsia and many other disorders. The most common manifestations of RPLS are altered mental status, decreased allertness, seizures, cortical blindness and other visual abnormalities, and transient motor deficits. The main finding in neuroimaging is posterior white matter oedema[16], often with a strikingly symmetrical involvment of the parietal and occipital lobes.

In this study we describe an anusual association between a thunderclap headache and reversible posterior leucoencephalopathy that occured in a female patient.

Case report

The case arrived at our observation concerned a woman of 38 years old that about five months before, after a period of intense psychological and physical stress, have been affected by recurrent episodes of severe headache. The patient referred an excruciating headache of instantaneous onset, as

sudden and unespected as the *"clap of thunder"*, that reached maximum intensity within one minute and usually lasted about two hours. This headache occurred spontanously while at rest or during light activities, or was precipited by intense exerction. These episodes occurred repeatedly in a week during the last five months.

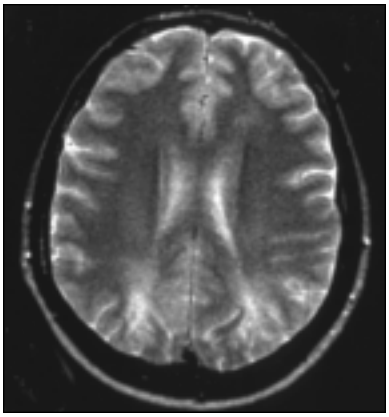

Fig. 1a

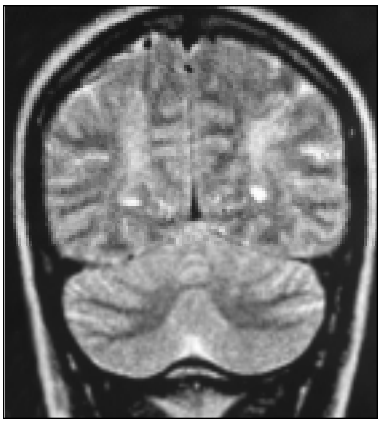

Fig. 1b

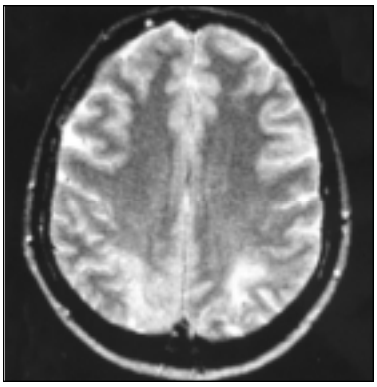

Fig. 1c

One week before coming to our observation she accused an episode, of the duration of few minutes, characterized from objective dizziness and visual loss, followed by loss of conscience.

Afterward, the patient has presented a reduction of the visus at left eye, postural brittleness, paresthesias to the left emysoma. After three days, while sleeping, muscular mioclonic contractions appeared in the left leg, motor block and block of the speech also occurred and spontaneously regressed after some minute. In the following days the patient has manifested other episodes caracterized by loss of conscience and seizures.

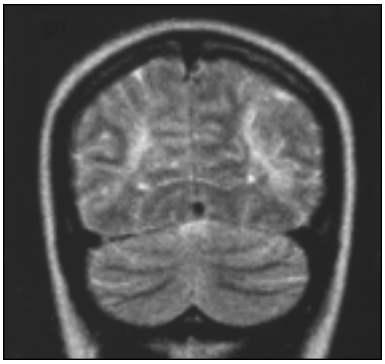

Fig. 1d:

Fig. 1a-b-c-d: Le sequenze RM T2 pesate mostrano iperintensità di segnale nelle regioni posteriori dei lobi parietali e nei lobi occipitali, a distribuzione bilaterale e simmetrica, senza enhancement dopo mezzo di contrasto.

The neurologic examination has highlighted a mild episodic confusional state. To the neuropsychological evaluation she displayed alterations of attention and memory, with amnesia and sign of amnesic aphasia, slowing of mental functions, without loss of the autobiographic memory.

An EEG was performed, resulting patologic for the presence of bouffeès of theta-delta activity on the central-anterior area of both sides; after about one month the EEG layout resulted lightly altered for the presence of rare slow generalized activity, prevalent on the central-anterior area of both sides. The MEP have highlighted an increase of the motor threshold and light increase of the time of motor conduction in the derived voltages to the brawn brief abductor of the inch of left hand; the VEP from incitement with pattern reversal with aims of 30' and 25' has highlighted a light increase of the difference intereye (OS> OD) in the values of latency of best positive component P100 in the responses to the aims of 15'.

The brain MRI has highlighted in white matter of parietal and occipital lobes two lesions with bilateral simmetric distribution with indistinct borders, iperintense in T2, with noenhancement. The examination of the cerebral liquid was normal.

It was also performed an immunological investigation that has given the followings results: IgG antibodies IgG [anti-cardiolipina] = 18 GPL, IGM [anti-cardiolipina] = 78, positive ANA 1/80. The ematologic examinations has highlighted a sideropenic anemia.

Discussion

The features of the headache showed by our patient appeared to be related to a thunderclap headache, because of instantaneous onset, duration and severity of pain and frequency of episodes.

Idiopatic TCH, which is associated with diffuse vasospasm[2], may produce fluctuating focal neurologic signs or simptoms, seizures and even stroke.

The clincal features, temporal pattern, mode of onset, and associated features of idiopatic TCH are not only unique, but quite distinct from the well-established clinical criteria of migraine and tension-type headache.

Idiopatic TCH is a diagnosis of exclusion, infact, although it is generally associated to intra or extracranial vascular pathology, such as subarachnoid hemorrage[8], cerebral venous sinus thrombosis or carotid artery dissection, it may represent a distinct primary headache syndrome.

Even in those cases where the initial CT and lumbar puncture are unremarkable, MRI (with MRA and venography) is strongly encouraged to exclude other causes of TCH such as cervicocephalic arterial dissection, venous sinus thrombosis, and pituitary apoplexy.

The study of MRI in our patient, which showed the presence of two lesions, bilateral and simmetric, in white matter of parietal and occipital lobes with a complete regression after three months, suggested the typical aspect of a reversible posterior leuchoencephalopathy.

The term *"reversible posterior leuchoen - cephalopathy"* describe a syndrome of confusion, seizures and visual disturbancies associated with transient, predominantly posterior cerebral lesions revealed by neuroimaging[5;7;15].

Descriptions of RPLS have enphasized caracteristic clinical and radiological presentation, despite the etherogeneous settings in which this transient syndrome occurs[9]. Similar clinical and radiological recovery have been described in a wide variety of clinical conditions, such as systemic lupus, vasculitis[10], hypercalcemia[6], porphyria. This syndrome is also seen in patients treated with immunosuppressive drugs[11], such as introvenous immunoglobulin, cyclosporin A, tacrolimus and interferon-alpha[14].

Hypertension is commonly associated with RPLS, but may be relatively mild and is not universally present, especially in the setting of immunosuppression[4].

Thunderclap headache has been also described in a recent report of a patient with hypertensive encephalopathy[13] and RPLS, though a number of cases which occurred in absence of severe hypertension have been reported.

Moreover a recent study showed a connection between thunderclap headache and RPLS[3], in which there was a diffused cerebral vasospasm. The mechanism of vasospasm in these patients is unclear. It has been suggested that there is a loss of autoregulation[12] resulting in dilatation of cerebral arterioles and disruption of the blood-brain barrier (BBB).

The preferential involvment of the parietal and occipital lobes is thought to be related to the relatively poor sympathetic innervation of the posterior circulation.

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Request reprints from:
Prof. INNOCENZO LUPO
Dipartimento di Neurologia, Oculistica,
Otorinolaringoiatria e Psichiatria
Università di Palermo
Via La Loggia
90100 Palermo
(Italy)