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Case Report

Characteristics of headache in relation to the manifestation of Susac syndrome

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ABSTRACT

Susac syndrome is characterized by a clinical triad of encephalopathy, branch retinal artery occlusion, and hearing loss. Due to the absence of the whole complex of the triad in the majority of cases at disease presentation, the syndrome often remains underdiagnosed and untreated. Headache is estimated to affect up to 80% of Susac syndrome patients, but the relevance of headache characteristics and profile is not yet clear. The proposed diagnostic criteria of the European Susac Consortium acknowledge headache as a possible brain manifestation if it is new, described as migrainous or oppressive, and precedes the other symptoms by not more than 6 months.

Herein, a case series of different migraine-like headache associations attributed to Susac syndrome is presented and discussed in relevance with previously published literature. Our patients experienced different presentations of migraine-like headache related with Susac syndrome: exacerbation and chronification of headache just before the manifestation of the first symptoms of Susac syndrome, the manifestation of headache during the first episode of the syndrome, and an increasing frequency of headache during the course of the disease. The diagnosis of Susac syndrome in all three cases was confirmed by typical clinical symptoms and findings in retinal fluorescein angiography, audiometry, and brain magnetic resonance imaging, based on the diagnostic criteria of the European Susac Consortium.

Based on the analysis of our presented cases, we conclude that headache attributed to Susac's syndrome is of migraine-like type but could be of different presentations in relation to the onset of the syndrome.

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1. Introduction

Susac syndrome (SuS) is a rare disorder that is thought to be caused by autoimmune-mediated occlusions of microvessels in the brain, retina, and the inner ear, and is characterized by a clinical triad consisting of encephalopathy, branch retinal artery occlusion (BRAO), and hearing loss [1]. Typical findings in patients with SuS include BRAO detected on retinal fluorescein angiography (FA), characteristic lesions (corpus callosum spokes, icicles, and snowballs on sagittal T2; black holes in central fiber seen on sagittal T1; capsule microinfarctions associated with white matter infarcts and leptomeningeal involvement) [2] on brain magnetic resonance imaging (MRI), and evidence of sensorineural hearing loss [3,4]. Due to multiple organ involvement and the absence of the classical clinical triad in the majority of cases at disease presentation, SuS often remains underdiagnosed, delaying the initiation of appropriate treatment [3,5]. The prevalence of SuS is unclear, and there were 304 cases of SuS reported worldwide [3]. Some authors estimated the prevalence rate of SuS at 0.14 cases per 100,000 population in the Central European population [6]. In Lithuania, the incidence rate of SuS is unclear because not a single case report has been published until now, and only solitary cases were diagnosed. Encephalopathy commonly is recognized as the initial manifestation of SuS and is associated with headache, psychiatric disorders (behavioral changes or cognitive impairment), and focal neurological symptoms [7]. Headache occurs in about 80% of the patients with SuS [3]. The proposed diagnostic criteria of the European Susac Consortium (EuSaC) acknowledge headache as a possible brain manifestation if it is new, described as migrainous or oppressive, and precedes the other symptoms by not more than 6 months [4]. Brain MRI, retinal FA, and audiometry are required tests for SuS diagnosis [3,8]. Due to the lack of randomized clinical trials, the treatment of SuS is symptomatic and based on clinical experience [3–5], but early immunosuppressive strategy is recommended [2]. It has been noticed that multiple sclerosis disease-modifying treatment is not effective and could deteriorate the neurological condition [9,10].

In this paper, we discuss three cases of patients who presented with different characteristics of headache associated with SuS.

2. Presentation of cases

2.1. *Case 1*

A 26-year-old woman with a history of rare migraine attacks from 15 years of age presented at the Emergency department when she experienced a sudden onset of an episode of diplopia that lasted for 30 min after 1 month of everyday migraine-like headache attacks (headache characteristics are described in Table 1; according to the ICHD-3 criteria). No specific treatment for headache was prescribed. Episodes of diplopia subsequently recurred several times during the period of a few days. Two weeks from the first episode of diplopia, the patient suddenly lost hearing in the left ear. Audiometry revealed left-sided sensorineural hearing disturbance. She was diagnosed with acute cochlear neuritis and was treated with glucocorticoids and hyperbaric oxygen therapy, which resulted in partial clinical improvement. One month later, the patient experienced an acute attack of somnolence, confusion/disorientation, vertigo, nausea, slurred speech, and hearing loss in the right ear, and was admitted to the Department of Neurology of our Hospital. Following an extensive diagnostic work-up, demyelinating disorders, vascular disorders, and chronic infections were ruled out, and the patient was diagnosed with SuS according to the diagnostic criteria of the EuSaC (all clinical presentations and diagnostic tests are shown in Table 2). The patient received treatment with high-dose glucocorticoids that was partly effective, and oral prednisolone was continued; headache attacks decreased, although sensorineural hearing loss in the right ear and slight ataxia remained. During the last 3 years of follow-up, no new bouts of SuS were suspected. Due to persistent deafness, the cochlear implantation in the right ear was performed. At this time, migraine attacks repeat 1–2 times a month, as it was before the manifestation of SuS.

2.2. *Case 2*

A 40-year-old woman with no past history of headache first presented at the Department of Neurology of our Hospital 1 year before due to an intensive headache attack that was followed by diplopia and vertigo of approximately 20 min in duration. Headache mostly affected the upper part of her head bilaterally, was of pulsating quality, and was associated with nausea and intensive tinnitus. Intensive headache lasted less than an hour, and then slowly diminished. The diagnosis of probable migraine with an aura was made. After the manifestation of the first headache episode, the patient started experiencing severe attacks of bifrontal headache 2–3 times a month lasting from 2 to 12 h and accompanied by nausea, photophobia and phonophobia, and avoidance of physical activity (the characteristics of the headache are described in Table 1 according to the ICHD-3 criteria). These headache attacks were alleviated with analgesics. Two months later, the patient experienced an episode of hemiparesis and hemihypesthesia accompanied by vertigo and balance disturbance. Due to suspected multiple sclerosis, the patient was treated with high doses of glucocorticoids, which resulted in clinical improvement; for maintenance, oral steroids were prescribed, and headache attacks decreased. Her migraine-like headache attacks repeated frequently for several months, and then gradually disappeared. A year later, such symptoms as balance impairment, hearing difficulty (mostly affecting the right ear), and diplopia appeared, and hypoesthesia and hemiparesis recurred. The patient was hospitalized in our department for further investigations for the second time. MS, vascular disorders, and chronic infections were ruled out, and the patient was diagnosed with SuS according to the diagnostic criteria of the EuSaC (Table 2). High doses of intravenous glucocorticoids were prescribed, which resulted in a significant clinical improvement, and immunosuppressive treatment was started subsequently. Currently, the patient has no complaints of headache.

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