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Review

Subarachnoid hemorrhage in systemic lupus erythematosus: Systematic review and report of three cases

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ABSTRACT

Objective: Systemic lupus erythematosus (SLE) is an autoimmune disorder of multifactorial etiology with a broad range of clinical manifestations. Cerebral disorders such as psychosis, seizures and cerebrovascular accidents are commonly described in SLE patients. Subarachnoid hemorrhage (SAH) presents a higher incidence than in the general population. Saccular aneurysms are the most frequent cause, but uncommon forms of SAH had been repeatedly reported in SLE patients. Through three case examples we outline these uncommon patterns, which we hypothesized, might be responsible for the higher incidence of SAH in SLE patients.
Methods: We present three patients previously diagnosed with SLE, who suffered a SAH and we perform a systematic literature review.
Results: All three cases presented with a Fisher grade IV bleeding. One harbored a distal middle cerebral artery aneurysm, another had multiple aneurysms, and the youngest patient had no findings on the cerebral angiography. A focal vasculitic process seemed most likely responsible for the two aneurismal cases. The angiography negative case was attributed to severe SLE systemic complications and had an unexpected bad outcome. Literature review yielded 39 previously reported cases.
Conclusions: The uncommon SAH patterns make etiological diagnosis challenging. Management can also be daunting due to both local and systemic consequences of SLE. Awareness of these uncommon patterns, management challenges, and higher likelihood of bad outcomes may help physicians dealing with this disease. Further research to unveil the mechanisms behind it is granted to fully understand SAH in SLE patients.

Contents

Abbreviations: ACA, anterior cerebral artery; AICA, anterior inferior cerebellar artery; CNS, central nervous system; CT, computed tomography; EVD, external ventricular drain; GCS, Glasgow Coma Scale; ICG, indocyanine green; MRSA, methicillin-resistant staphylococcus; MCA, middle cerebral artery; mRS, modified rankin score; PCoA, posterior communicating artery; SAH, subarachnoid hemorrhage; SLE, systemic lupus erythematous.

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1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by complex-mediated lesions of blood vessels in multiple body organs, leading to different types of vascular complications. Incidence rate of SLE is quoted between 1 and 10/100,000 per year depending on the population studied (sex, ethnicity, geographical location and age). The incidence is markedly higher in non-Caucasian population [1]. Immune complexes that cause vasculitis in Lupus may contain DNA and Anti-DNA antigens, or Ro and Anti-Ro antigens and usually affect small vessels; medium and large vessels are only exceptionally affected [2]. The central nervous system (CNS) can be affected in 25–50% of SLE patients mostly due to seizures, psychosis, and cerebrovascular accidents [3]. Incidence of subarachnoid hemorrhage (SAH) is considered higher in SLE patients than in the general population, with a rate of 49.4 vs. 10.2 per 100,000 person-years [4]. However, the underlying cerebral vasculitis described in SLE does not justify the pathophysiology of all the SAH cases, and the causes leading to this increased incidence remain unclear [5,6].

In this manuscript, we review case reports of SAH in SLE patients found in the medical literature. While we do not have enough information to assess the causes of SAH in SLE patients, we hypothesized that there would be a few clinical identifiable patterns whose detailed description may help improving diagnostic and management strategies of these complex cases. Bad prognostic spontaneous SAH, multiple aneurysms and fusiform or uncommon aneurysm might reflect the characteristic behavior of SAH in SLE. We also review our own experience with SAH in SLE patients and report three additional cases that we managed in our department.

2. Methods

2.1. Literature review

We have conducted a systematic literature search in PubMed and MedLine using "systemic lupus erythematosus" and "subarachnoid hemorrhage" as key words and free text searches. We limited language to English, French and Spanish and publication period from January 1980 to December 2013. Abstracts of identified references were initially screened and full publications of potentially relevant articles were obtained for further examination. The references of each included report were also screened for additional manuscripts. The initial search strategy identified 25 manuscripts. We eliminated 9 of them that were not written in English, French or Spanish. Full text of the remaining 16 articles were obtained and 4 of them were excluded for reporting other causes of SAH (such as traumatic vertebral dissection) not corresponding with aneurysmatic or angio-negative SAH. References of the remaining 12 articles yielded 6 extra manuscripts meeting our search criteria. Out of them, one article by a Japanese author reported two new cases in English and reviewed 55 more from the Japanese literature. Detailed data for the two cases reported in English was extracted but original data for the remaining 55 cases was either incomplete or written in Japanese and therefore, was excluded. Overall, we found 18 unique articles reporting SAH in 39 SLE patients.

2.2. Cases report

Between January 1996 and December 2013, we identified 3 patients previously diagnosed with SLE out of the 641 who were admitted with non-traumatic SAH and prospectively collected in our service database. Their demographic features, clinical presentation, management strategy and outcomes were thoroughly reviewed.

Clinical outcomes were assessed using the modified Rankin Score (mRS). Good outcomes were defined as a final mRS score of 0–2, and poor outcomes were defined as a final mRS greater than 2. Improvement was defined as a decrease in mRS score (change in the mRS of less than or equal to zero), and deterioration was defined as an increase in mRS score (change in the mRS of greater than zero).

3. Results

To the best of our knowledge, 39 patients with SLE and SAH have been described in English, Spanish or French articles. Table 1 summarizes the main features, treatment strategy and outcomes of all these reported cases. Eleven cases presented an angiographically negative SAH and 5 out of those 11 had a bad outcome (4 deaths, 1 severe disability). Out of the remaining 27 reported cases, all with a confirmed aneurysmatic SAH, 6 patients presented with multiple aneurysms and 6 had a either a fusiform aneurysm, an aneurysm in an atypical location or both.

The analysis of this search revealed that three main patterns have been consistently reported as leading to SAH in SLE patients. First, distal fusiform aneurysms with aberrant morphology in uncommon locations [7,8]; second, multiple saccular aneurysms [9]; and third, angiographically negative SAH with an unexpected bad prognosis [10].

3.1. Ilustrative cases

Each of these cases illustrate one of the aforementioned 3 main patterns usually responsible for SAH in SLE patients.