

Aseptic Meningitis Revealing Isolated Kikuchi-Fujimoto Disease

Salem Bouomrani* and Nesrine Regaïeg

Department of Internal medicine, Sfax Faculty of Medicine, Tunisia

Received: 📅 November 02, 2018; Published: 📅 November 08, 2018

*Corresponding author: Salem Bouomrani, Department of Internal medicine, Military Hospital of Gabes, Gabes 6000. Tunisia

Abstract

Introduction: Kikuchi-Fujimoto's disease (KFD) or histiocytic necrotizing lymphadenitis is a rare entity that may represent a real diagnostic challenge for the clinician because of its highly polymorphous and sometimes unusual presentations. We report an original observation of KFD with aseptic meningitis as inaugural manifestation.

Case report: 30-year-old woman, without pathological medical history, was hospitalized via the emergency department for exploration of a meningeal syndrome with cervical lymphadenopathies. The lumbar puncture showed aseptic meningitis: clear, normotensive cerebrospinal fluid with leukocytes at 28/mm³ (90% lymphocytes), red blood cells at 2/mm³, proteinorrachia at 0.58 g/l, glucorrachia at 4 mmol/l for venous glycemia at 8 mmol/l, and negative direct examination and culture. Cerebromedullary MRI and cerebral angio-MR were without abnormalities. Further infectious and immunological investigations were negative. Cervical lymph node biopsy showed histological and Immunohistochemical aspects suggesting KFD. Treated with systemic corticosteroids, the evolution was favorable with no recurrence.

Conclusion: KFD-associated aseptic meningitis remains rare, and the inaugural forms are exceptional and often difficult to diagnose. A better knowledge of this association avoids unnecessary investigations, recurrence, and improves the prognosis of the disease.

Keywords: Aseptic Meningitis; Kikuchi-Fujimoto's Disease; Histiocytic Necrotizing Lymphadenitis

Introduction

Described for the first time in 1972 by Kikuchi M and Fujimoto Y [1,2], Kikuchi-Fujimoto disease (KFD) is a very rare necrotizing histiocytic lymphadenitis [3] which is classically characterized by a febrile polyadenopathy with a marked inflammatory biological syndrome in young subject, mainly Asian women. KFD may be isolated [4] or associated with several other systemic diseases of a dysimmune nature, particularly systemic lupus erythematosus [5,6]. Systemic disorders, including neurological ones, during this disease are exceptional and represent a real diagnostic challenge for clinicians, especially in the inaugural and isolated forms [1-4]. Although recognized to be the most frequent neurological event in this disease, KFD-associated lymphocytic meningitis remains rare, and the inaugural forms are exceptional and often difficult to diagnose [7-9]. We report an original observation of aseptic meningitis inaugural of an isolated KFD in a 30-year-old Tunisian woman.

Case report

A 30-year-old Tunisian woman, without pathological medical history, was hospitalized via the emergency department for exploration of a meningeal syndrome. The history of his illness dates back two days before his hospitalization, by the appearance of headaches with fever and vomiting not improved by the symptomatic treatment. The somatic examination found a patient conscious, well oriented, without focal neurological deficit, feverish at 39°C, significant stiffness of the neck, with photophobia, sonophobia, and multiple cervical adenopathies slightly sensitive. The ENT and bucco-dental examination were without abnormalities. The biology showed leukocytosis at 15 600/mm³ with 85% neutrophils, a marked biological inflammatory syndrome with a high erythrocyte sedimentation rate at 135mm/H1, a C-reactive protein at 28 mg/l and a hyperfibrinemia at 8 g/l. The other basic bioassays were without abnormalities: hemoglobin, platelets, blood glucose, serum

calcium, ionogram, liver enzymes, muscle enzymes, creatinine, serum protein electrophoresis, and urine analysis. Chest X-ray and electrocardiogram were normal. The ophthalmic examination with fundus of the eye did not show any papillary edema.

The lumbar puncture showed a clear, normotensive cerebrospinal fluid, and the biochemical and cytobacteriological analysis confirmed the diagnosis of aseptic lymphocytic meningitis: leukocytes at 28/mm³ (90% lymphocytes), red blood cells at 2/mm³, proteinorrachia at 0.58 g/l, glucorrachia at 4 mmol/l for venous glycemia at 8.6 mmol/l, and negative direct examination and culture. Specific tuberculosis tests in the blood, urine and cerebrospinal fluid were negative. Similarly, blood cultures, viral and bacterial serology for lymphocytic meningitis, anti-nuclear antibodies, and anti-soluble nuclear antigen antibodies were negative. Cerebro-medullary magnetic resonance imaging (MRI) and cerebral angio-MR showed no abnormalities. Biopsy of a posterior cervical ganglion showed follicular hyperplasia, histiocytic and monocytic infiltrate with ganglionic necrosis, and no signs of lymphoma or tuberculosis. The immuno-labeling by anti-CD68 antibodies was positive. Thus, the diagnosis of aseptic lymphocytic meningitis within the framework of an isolated KFD was retained and the patient was treated with oral corticosteroids at a dose of 1 mg/kg/day for four weeks followed by gradual decrease, in association with hydroxychloroquine at a dose of 400 mg/day. The evolution was rapidly favorable with apyrexia and disappearance of meningeal signs after two days and total regression of cervical lymphadenopathies after two weeks. The lumbar puncture performed at one month was strictly normal, and no recurrence has been noted for two years now.

Discussion

Non-infectious aseptic meningitis is a real diagnostic challenge in current medical practice because of the absence of specific clinical and biological signs and the multitude of possible etiologies [10,11]. The main causes of these meningitis are: systemic diseases (connective tissue diseases, primary vasculitis and granulomatosis), cancers and hematological malignancies (neoplastic meningitis), and drug intake (drug-induced meningitis) [10]. Among the systemic diseases, KFD remains an exceptional etiology of aseptic meningitis: indeed, only one case of aseptic meningitis was secondary to KFD in the series of 180 cases of aseptic meningitis of Jarrin I et al. [12] a frequency of 0.55%. Similarly, during a KFD, lymphocytic aseptic meningitis remains an exceptional event: only two cases in the series of 91 patients with KFD of Dumas G (2.1%) [8], and only two observations in the series of 69 cases of KFD of Nakamura I et al. [7] (2.8%).

The majority of cases are reported separately as sporadic cases and the review of the literature finds only about 30 observations of aseptic meningitis associated to KFD [7-9,13-21]. Aseptic meningitis is therefore described as unusual [13] and atypical [14]

during this disease. It can be acute [15] or chronic and recurrent [9,16], isolated or associated with other neurological signs [17-19] or other severe visceral manifestations of the disease [14,20], be seen during the evolution of already known KFD or more exceptionally be the first sign revealing it [14,17,21]. Evolution is usually favorable under systemic corticosteroids [7-9,13-21] and recurrence remains rare and the prerogative of undiagnosed forms of the disease [9,16].

Conclusion

As rare as it is, Kikuchi-Fujimoto disease as possible etiology of aseptic meningitis must be known. This knowledge will in many cases avoid unnecessary investigations, recurrence, specific or non-specific complications, and improve the prognosis of the disease. Our case is distinguished by the inaugural character of aseptic meningitis.

References

1. Kikuchi M (1972) Lymphadenitis showing focal reticulum cell hyperplasia with nuclear debris and phagocytes: a clinicopathological study. *Nippon Ketsueki Gakkai Zasshi* 35: 379-380.
2. Fujimoto Y, Kijima Y, Yamaguchi K (1972) Cervical subacute necrotizing lymphadenitis: a new clinicopathologic entity. *Naika* 20: 920-927.
3. Ruaro B, Sulli A, Alessandri E, Fraternali Orcioni G, Cutolo M (2014) Kikuchi-Fujimoto's disease associated with systemic lupus erythematosus: difficult case report and literature review. *Lupus* 23(9): 939-944.
4. Dokic M, Begovic V, Bojic I, Tasic O, Stamatovic D (2003) Kikuchi-Fujimoto disease. *Vojnosanit Pregl* 60(5): 625-630.
5. Sopena B, Rivera A, Vázquez Triñanes C, Fluiters E, González Carrero J, et al. (2012) Autoimmune manifestations of Kikuchi disease. *Semin Arthritis Rheum* 41(6): 900-906.
6. Regaïeg N, Ben Hamad M, Belgacem N, Baili H, Lassoued N, et al. (2018) Kikuchi-Fujimoto Disease Associated to Connectivitis -About Two Cases. *Open Access Journal of Internal Medicine* 1(1): 21-24.
7. Nakamura I, Imamura A, Yanagisawa N, Suganuma A, Ajisawa A (2009) Medical study of 69 cases diagnosed as Kikuchi's disease. *Kansenshogaku Zasshi* 83(4): 363-368.
8. Dumas G, Prendki V, Haroche J, Amoura Z, Cacoub P, et al. (2014) Kikuchi-Fujimoto disease: retrospective study of 91 cases and review of the literature. *Medicine (Baltimore)* 93(24): 372-382.
9. Komagamine T, Nagashima T, Kojima M, Kokubun N, Nakamura T, et al. (2012) Recurrent aseptic meningitis in association with Kikuchi-Fujimoto disease: case report and literature review. *BMC Neurol* 12: 112.
10. Bouomrani S, Trabelsi S, Nouma H, Baili H, Belgacem N, et al. (2016) Aseptic Noninfective Meningitis: Etiologic Survey. *Clin Res Infect Dis* 3(5): 1042.
11. Shukla B, Aguilera EA, Salazar L, Wootton SH, Kaewpoowat Q, et al. (2017) Aseptic meningitis in adults and children: Diagnostic and management challenges. *J Clin Virol* 94: 110-114.
12. Jarrin I, Sellier P, Lopes A, Morgand M, Makovec T, et al. (2016) Etiologies and Management of Aseptic Meningitis in Patients Admitted to an Internal Medicine Department. *Medicine (Baltimore)* 95(2): e2372.
13. Ficko C, Andriamanantena D, Dumas G, Claude V, Rapp C (2013) Kikuchi's disease: An unusual cause of lymphocytic meningitis. *Rev Neurol (Paris)* 169(11): 912-913.

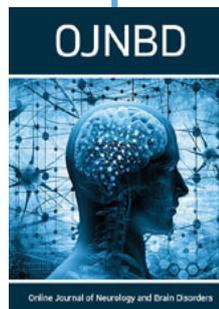
14. Méni C, Chabrol A, Wassef M, Gautheret Dejean A, Bergmann JF, et al. (2013) An atypical presentation of Kikuchi-Fujimoto disease. *Rev Med Interne* 34(6): 373-376.
15. Valle Arcos, Villarejo Galende A, Martínez González M, Martín Gil L, Calleja Castaño P, et al. (2010) Acute lymphocytic meningitis presenting as Kikuchi's disease. *Rev Neurol* 51(5): 314-315.
16. Itokawa K, Fukui M, Nakazato Y, Yamamoto T, Tamura N, et al. (2008) A case of subacute necrotizing lymphadenitis with recurrent aseptic meningitis 11 years after the first episode. *Rinsho Shinkeigaku* 48(4): 275-277.
17. Vaz M, Pereira CM, Kotha S, Oliveira J (2014) Neurological Manifestations in a Patient of Kikuchi's Disease. *J Assoc Physicians India* 62(11): 57-61.
18. Moon JS, Il Kim G, Koo YH, Kim HS, Kim WC, et al. (2009) Kinetic tremor and cerebellar ataxia as initial manifestations of Kikuchi-Fujimoto's disease. *J Neurol Sci* 277(1-2): 181-183.
19. Kido H, Kano O, Hamai A, Masuda H, Fuchinoue Y, et al. (2017) Kikuchi-Fujimoto disease (histiocytic necrotizing lymphadenitis) with atypical encephalitis and painful testitis: a case report. *BMC Neurol* 17(1): 22.
20. Jain J, Banait S, Tiewsoh I, Choudhari M (2018) Kikuchi's disease (histiocytic necrotizing lymphadenitis): A rare presentation with acute kidney injury, peripheral neuropathy, and aseptic meningitis with cutaneous involvement. *Indian J Pathol Microbiol* 61(1): 113-115.
21. Khishfe BF, Krass LM, Nordquist EK (2014) Kikuchi disease presenting with aseptic meningitis. *Am J Emerg Med* 32(10):1298(e1-e2).



This work is licensed under Creative Commons Attribution 4.0 License

To Submit Your Article Click Here: [Submit Article](#)

DOI: [10.32474/OJNBD.2018.02.000129](https://doi.org/10.32474/OJNBD.2018.02.000129)



Online Journal of Neurology and Brain Disorders

Assets of Publishing with us

- Global archiving of articles
- Immediate, unrestricted online access
- Rigorous Peer Review Process
- Authors Retain Copyrights
- Unique DOI for all articles