Asian Journal of Research and Reports in Neurology

3(2): 19-23, 2020; Article no.AJORRIN.63758

Pachymeningitis in Ulcerative Colitis: Accidental Association?

Abir Derbel[1*], Mariem Ghribi[1], Olfa Frikha[1], Leila Mnif[1], Sameh Marzouk[1], Nabil Tahri[1] and Zouhir Bahloul[1]

[1]Departement of Internal Medicine, Hédi Chaker Hospital, 3029 Sfax, Tunisia.

Authors' contributions

This work was carried out in collaboration among all authors. Author AD designed the case and wrote the first draft of the manuscript. Authors SM and MG managed the analyses of the case. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Takashi Ikeno, National Institute of Mental Health, National Center of Neurology and Psychiatry, Japan.
Reviewers:
(1) Bajko Zoltan, University of Medicine, Romania.
(2) Efrain Orlando Silva Vega, Technical University Of Manabi, Ecuador.
Complete Peer review History: http://www.sdiarticle4.com/review-history/63758

Case Study

Received 29 September 2020
Accepted 05 December 2020
Published 21 December 2020

ABSTRACT

Pachymeningitis is a rare disease defined as diffuse or localized inflammatory and fibrosing thickening of the dura mater. Its occurrence in context of Ulcerative colitis has been exceptionally described. We report a case of a woman with ulcerative colitis who developed pachymeningitis in the same time of the flare up of her disease. She was suffered from debilitating headache without any other neurological abnormalities. Brain Magnetic Resonance Imaging was revealed pachymeningitis. The patient presented a recurrent pachymeningitis associated to her ulcerative colitis; Corticosteroids improved initially her digestive and neurological symptoms therefore, a subtotal colectomy and a second round of steroids were indicated with resolution of the symptoms.

Keywords: Pachymeningitis; ulcerative colitis; neurological abnormalities.

1. INTRODUCTION

The neurological manifestations of inflammatory bowel disease (IBD) are extremely rare and diverse.

Neurological manifestations of intestinal bowel disease (IBD) are extremely rare and various.

They are reported in 3% of patients [1]. They represent a diagnostic issue and lead to

**Corresponding author: E-mail: abirderbel@gmail.com;*

important morbidity. To our knowledge, few cases of pachymeningitis associated to IBD have been described. It's a rare disease defined as diffuse or localized inflammatory and fibrosing thickening of the dura mater [2]. Severe headache and vision deterioration are the classic presenting symptoms. Many etiologies can be accused including infection, autoimmune reaction, sarcoidosis and neoplastic process.Idiopathic pachymeningitis is a diagnosis of exclusion[3]. Its occurrence in context of Ulcerative colitis (UC) has been exceptionally described [4].

We report a new case of a women followed for ulcerative colitis that developed hypertrophic pachymeningitis simultaneous with the flare up of her disease.

2. PRESENTATION OF CASE

Ms I.B., 44-years-old was diagnosed as having ulcerative colitis (UC) in 1995. She had hematologic side effects to Azathioprin. In her past medical history, we found two early abortion and three normal pregnancies and hypercholesterolemia. For her UC, she was treated with 5 ASA derivatives.

In January 2015, she developed bloody diarrhea 9 times a day associated with severe headache. She was hospitalized in gastroenterology department. Diagnosis of a flare-up of UC was made. She was treated with injectable corticosteroids relayed by oral corticosteroids (1mg /Kg/Day of prednisone equivalent) associated secondary to Azathioprin for which, she developed severe leucopenia requiring its stop cause. The symptoms improved but at tapering of Steroids (under 20mg/day), she noticed the recurrence of headaches and diarrhea. (Specialized Oto-rhino-laryngology examination, brain scan). Brain Magnetic Resonance Imaging (MRI) was practiced and revealed intensive regular and diffuse enhancement of the dura mater. Then, she was admitted in our department for exploration of this pachymeningitis. She was treated by high doses of corticosteroids, then, she was admitted in our department for exploration of this pachymeningitis.

At her admission at our department, she suffered from mouth ulcer and bloody diarrhea. She had no fever. Neurological examination was normal. On laboratory Serum tests, inflammatory markers were negatives (Erythrocyte Sedimentation Rate was at 14 mm / hour, C-reactive protein 7 mg /L). Renal and hepatic analysis was normal. Blood numeration showed normocytic anemia at 11g/dl. Infectious investigations: HCV, HBV, HIV, EBV, CMV, VZV, B19, Rickettsiosis, Brucella, Lyme, syphilis, candidiasis and tuberculosis evaluation were negative. Anti nuclear antibodies, rheumatoid factor, cryoglobulinemia and anti phospholipid anti bodies were negative. A quantitative serum immunoglobulin tests were normal. The detection of the ASCA antibodies was negative. Anti-Neutrophil Cytoplasmic Antibodies (ANCA) showed the presence of auto-antibodies targeting the serine protease proteinase-3 (PR3). Second analysis after four months was negative. Specialized Oto-rhino-laryngology examination was normal. Complex computed tomography of the cranial and facial bones was normal.

Brain MRI, done to control pachymeningitis after 4 months of corticosteroid treatment, was normal. This pachymeningitis was symptomatic by chronic headache without any other neurological manifestation which disappeared at instauration of corticosteroids and appeared again at their stopping. Corticosteroid at the doses of 10 mg of equivalent prednisone was the efficient dose. Five months after her leaving (November 2015), progress showed recurrence of bloody diarrhea and headache. A flare up of her inflammatory disease was retained and we increased the doses of corticosteroids at 20 mg of equivalent prednisone with clinical improvement. Multiple flare up of her inflammatory disease were noticed indicating a subtotal colectomy In March 2016, she had subtotal colectomy. Corticosteroids were stopped and any flare up was noticed since four years.

3. DISCUSSION

Pachymeningitis is a rare inflammatory disorder involving the cranial and spinal dura mater. Its clinical manifestations depend on location of lesions [5].

Headache and loss of cranial nerve function were the most common presenting features of HP regardless of etiology [2]. Headache predominates in cranial pachymeningitis in her idiopathic form and can be present until 100% of cases [3]. Headache profile was described by Wang and al. [6] as a chronic daily headache. After hedache, other symptoms of hypertrophic pachymeningitis can be observed: cranial nerve palsies, cerebellar dysfunction and radiculopathy

[7,8]. In our case, the main complaint was severe chronic headache.

Exact etiopathogenesis of this entity is still unknown, but auto immune mechanism, infectious or infiltrative phenomenon was largely described.

Many bacterial, viral and fungal infections were included like syphilis, Lyme, Tuberculosis HCV, HBV, HIV, EBV, CMV, VZV, B19, rickettsiosis, brucella, and candidasis [3,8].

Auto-immune and infiltrative diseases like rheumatoid arthritis, ANCA associated vasculitis, neuro sarcoidosis, Behçet vasculitis and psoriatic arthritis were largely reported as a cause of pachymeningitis [2,9,10]. Others like dural metastasis and dural changes to adjacent tumor, intra cranien hypotension may be responsible of pachymeningitis [2,3,11].

In a study guided by Lewis D. Hahn, about 22 patients [2], the most common diagnosis was idiopathic pachymeningitis in 50% of cases. There were 27% of neurosarcoidosis, 9% of Granulomatosis with polyangiitis, 5% of psoriatic arthritis, 5% of Vogt-Koyanagi-Harada disease, and 5% of inflammatory pseudo tumor.

Pachymeningitis associated to ulcerative colitis represents a rare association. It has been described at the first time by Zongqi Xia in November 2010 [4]. Neurologic disorder in IBD had been largely reported and could be caused by many mechanisms: cerebro vascular disease, cerebral vasculitis, immune mediated neuropathy or cerebral demyelinisation. Few cases reported the neurologic manifestations occurring in IBD associated to PR3 ANCA antibodies such as the case reported by Yuki Kirito in which, cranial neuropathy was diagnosed in a patient with ulcerative colitis having PR3 ANCA without granulomatous polyangeitis [12].

In our case, pachymeningitis wasn't idiopathic. In fact, in this context of inflammatory bowel disease, the proposed pathogenesis was chronic inflammation directed against antigens shared by gastrointestinal mucosa and meninges. This hypothesis was supported in our patient due to the significant improvement observed by corticosteroids.

Another finding in our patient was the positivity of anti PR3 antibodies.

The positivity of anti PR3 antibodies in ANCA associated vasculitis was described as a cause of cerebral vasculitis [13]. They are also responsible of pachymeningitis in this disease [9]. Their positivity in ulcerative colitis patients was largely described as a marker of IBD [14] but their pathogenic role in pachymeningitis in patient with ulcerative colitis was unknown.

Pachymeningitis represent a challenge in diagnosis and treatment. Its diagnosis was based at MRI features which showed dural thickening and intensive enhancement. In a short study about 12 patients [15] meningeal thickening was localized and asymmetrical (n = 6) or diffuse (n = 4), intensely enhanced after contrast injection. Associated intraparenchymatous lesions were found in five cases. MRI findings cannot be used to identify etiology. Idiopathic pachymeningitis generally presented diffuse and regular enhancement involving the posterior fossa. Thickening of the falx and tentorium has previously been reported to be the most common finding of idiopathic hypertrophic pachmeningitis [2]. Cavernous sinus involvement is also common. Regular and nodular patterns have been seen in prior series. The gold standard to diagnose idiopathic pachymeningitis relies on biopsy [2,8].

There is no optimal treatment for pachymeningitis but if untreated, it progress frequently. There is no therapeutic consensus. The most common approach is to initiate corticosteroid therapy. Benefit may be partial and temporary. Then, when recurrences were noticed, adjunction of immunosuppressant was recommended [1,3,5]. Our patient had a favorable response to steroid therapy but she suffered from recurrence of headache with steroid taper. We try to add an immunosuppressant Azathioprin but she developed severe leukopenia leading to their stop. In trying to stop inflammatory process by surgical treatment in our patient was efficient. That's why, stopping inflammatory process in digestive system lead to arrest inflammatory reaction against meninges' antigens.

Neurological interventional surgery was sometimes required in advanced cases with intracranial hypertension or nerve impairment.

4. CONCLUSION

In summary, our case report about pachymeningitis which occurred in female with

ulcerative colitis was the second described in the literature. The dura mater can be involved in many inflammatory diseases. Corticosteroid therapy with or without immunosuppressant was effective.

In this research on pachymeningitis in which the case of a woman with ulcerative colitis is presented, it is determined that the dura mater may be involved in many inflammatory bowel diseases. Given the above, it is concluded that treatment with corticosteroids with or without immunosuppressants is very effective.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

ACKNOWLEDGEMENTS

We would like to thank all those who provided assistance, preparation and funding for the manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Ferro JM, Oliveira SN, Correia L. Neurologic manifestations of inflammatory bowel diseases. Handb Clin Neurol. 2014;120:595-605.
DOI: 10.1016/B978-0-7020-4087-0.00040-1
PMID: 24365340
2. Hahn LD, Fulbright R, Baehring JM. Hypertrophic pachymeningitis. J Neurol Sci. 2016;15(367):278-83.
DOI: 10.1016/j.jns.2016.06.024
Epub 2016 Jun 11
PMID: 27423604
3. Fain O, Seror O, Wirth JF, Heron C, Mathieu E, Chamouard JM and Al. Les pachyméningites crâniennes [Cranial pachymeningitis]. Revue de Med Interne French. 1999;20(3):234-46.
DOI: 10.1016/s0248-8663(99)83051-3
PMID: 10216880
4. Xia Z, Chen-Plotkin A, Schmahmann JD. Hypertrophic pachymeningitis and cerebral venous sinus thrombosis in inflammatory bowel disease. J Clin Neurosci. 2010;17(11):1454-6.
DOI: 10.1016/j.jocn.2010.04.019
Epub 2010 Aug 19
PMID: 20727771
PMCID: PMC2946484
5. Karthik SN, Bhanu K, Velayutham S, Jawahar M. Hypertrophic pachymeningitis. Ann Indian Acad Neurol. 2011;14(3):203-4.
DOI: 10.4103/0972-2327.85896
PMID: 22028536
PMCID: PMC3200046
6. Wang YJ, Fuh JL, Lirng JF, Lu SR, Wang SJ. Headache profile in patients with idiopathic hypertrophic cranial pachymeningitis. Headache. 2004;44(9): 916-23.
DOI: 10.1111/j.1526-4610.2004.04175.x
PMID: 15447702
7. Prabhakar S, Bhatia R, Lal V, Singh P. Hypertrophic pachymeningitis: Varied manifestations of a single disease entity. Neurol India. 2002;50(1):45-52.
PMID: 11960151
8. Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. Neurology. 2004;9;62(5): 686-94.
DOI:10.1212/01.wnl.0000113748.53023.b7
PMID: 15007115
9. Sakairi T, Sakurai N, Nakasatomi M, Ikeuchi H, Kaneko Y, Maeshima A, Nojima Y, Hiromura K. Hypertrophic pachymeningitis associated with antineutrophil cytoplasmic antibody-associated vasculitis: A case series of 15 patients. Scandinavian Journal of Rheumatology. 2019;48(3):218-224.
DOI: 10.1080/03009742.2018.1498916
10. Gelfand JM, Jeffrey M. Neurosarcoidosis, Pachymeningitis, Behcet's Disease. The American Academy of Neurology Institute. 2017;6.
11. Lakshmi Nayak, Lauren E. Abrey, Fabio M. Iwamoto. Intracranial Dural Metastases, American Cancer Journal. 2009;115(9): 1947-53.
DOI: 10.1002/cncr.24203
12. Kirito Y, Yamamoto D, Uchiyama T. Proteinase 3-antineutrophil cytoplasmic antibody-positive ulcerative colitis presenting with abducens neuropathy. BMJ Case Rep. 2017;9:bcr2016218353

13. Unnikrishnan A, Azodi S, Ansari N, Brown M, Kamnetz J, Uchiyama RC. PR3ANCA related cerebral vasculitis in ulcerative colitis presenting with orbital involvement: A case report with review of literature. Case Rep Rheumatol. 2014;2014:582094.
DOI: 10.1155/2014/582094
Epub 2014 Jul 2.
PMID: 25097791
PMCID: PMC4101947

14. Radice A, Bianchi L, Maggiore U, Vaglio A, Sinico RA. Comparison of PR3-ANCA specific assay performance for the diagnosis of granulomatosis with polyangiitis (Wegener's). Clin Chem Lab Med. 2013;51(11):2141-9.
DOI: 10.1515/cclm-2013-0308
PMID: 23798618.

15. Ech-Cherif El Kettani N, Regragui W, Arkha Y, Ait Benhaddou E, Fikri M, Benomar A. Les pachyméningites intracrâniennes: Aspects TDM et IRM, Feuillets de Radiologie. 2012;52(5):263-272. ISSN. 0181-9801. French.
DOI:https://doi.org/10.1016/j.frad.2012.04.009

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/63758