Research Square

Preprints are preliminary reports that have not undergone peer review.
They should not be considered conclusive, used to inform clinical practice,
or referenced by the media as validated information.

Limbic encephalitis as a rare manifestation of relapsing polychondritis: A case report and literature review

Rachid Belfkih (belfkih.chu2@gmail.com)
Abdelmalek Essaadi University Faculty of Medicine and Pharmacy - Tangier: Universite Abdelmalek Essaadi Faculte de Medecine et de Pharmacie - Tanger

Omar Ghomari Khayat
Abdelmalek Essaadi University Faculty of Medicine and Pharmacy - Tangier: Universite Abdelmalek Essaadi Faculte de Medecine et de Pharmacie - Tanger

Fatima Zohra El Amrani
Abdelmalek Essaadi University Faculty of Medicine and Pharmacy - Tangier: Universite Abdelmalek Essaadi Faculte de Medecine et de Pharmacie - Tanger

Research Article

Keywords: Case report, Relapsing Polychondritis, Limbic encephalitis

Posted Date: April 8th, 2022

DOI: https://doi.org/10.21203/rs.3.rs-1521268/v1

License: This work is licensed under a Creative Commons Attribution 4.0 International License.
Read Full License

Abstract

Relapsing polychondritis is a systemic immune-mediated disease defined with recurrent inflammatory episodes of cartilaginous structures. Ear and nose chondritis are the most common clinical presentation. However, potential life-threatening complications could occur along the course of the disease. Its diagnosis is usually made based on a set of criteria determined by organs involvement. There exist several therapeutic options; they depend on the nature and severity of organs damage. We report a clinical case of a relapsing polychondritis presented with limbic encephalitis.

Introduction:

Relapsing Polychondritis (RP) is an autoimmune mediated inflammatory disease of cartilaginous and proteoglycan riche tissues. It involves primarily nose, ear, larynx and tracheobronchial tree cartilage, with possible and eventual association to cutaneous, rheumatological, cardio-vascular and neurological disorders. Its course is marked with repeated flares, leading to multiple life-threatening complications dominated by airway collapse. Herein, we report a case of a male patient, whose RP condition was unknown at first encounter, that presented a limbic encephalitis leading to the diagnosis of relapsing polychondritis.

Case Report:

A 54-year-old male patient presented to the department of neurology with a state of confusion and balance disorders, he has a history of insulin-dependent diabetes and high blood pressure. Five months earlier, the patient suffered from painless bilateral red eyes with decreased visual acuity. Over the course of the last three months, he started developping memory disorders, hearing impairment, dizziness, and experiencing moderate headaches, unexplained crying and visual hallucinations. There was no reported fever, seizures or vomiting. On physical exam, the patient was confused, but had normal vital signs values. Neurological exam revealed cerebellum ataxia with dysmetria and intention tremors, associated to bilateral hypoacusis and impaired visual acuity. Whereas, no motor or sensitive deficits were present. Cranial nerves exam, muscle tone and deep tendons reflexes were all normal. On his mini-mental status examination, he had a score of 22 objectifying a mild cognitive impairment. The rest of physical exams revealed no abnormality. Investigations included performing a brain MRI that showed bilateral hypersignal lesions, poorly bordered, in medial temporal lobe on fluid-attenuated inversion recovery (FAIR) sequence (Fig. 1). A spinal tap was done and cerebral-spinal fluid analysis was consistent with aseptic meningitis (White blood cell (WBC) count of 60/ml, glucose at 0,4g/l, and protein at 1,37g/l). An electroencephalogram (EEG) was run, it showed normal background rhythms. Fundoscopic exam revealed anterior ischemic optic neuropathy in the right eye. The audiometer on the other hand has shown severe mixed hearing loss in the right side and a presbycusis hearing loss in the left side. A full blood panel was realized; it included blood count, c-reactive protein test, renal function, liver function, thyroid function and an electrolyte panel that were all normal. Screening tests against HIV, syphilis and hepatitis B were negative. Autoantibody tests for autoimmune conditions included antinuclear antibodies (ANA),

extractable nuclear antigen (ENA) and anti-neutrophil cytoplasmic antibodies (ANCA), they came up also all negative. Screening for autoimmune anti-neuronal antibodies included testing for anti-HU, anti-RI, anti-amphiphysin, anti-Ma2, anti-CV2 and anti-YO antibodies, the results were likewise negative. Performed thoraco-abdominopelvic CT scan (TAP-CT) revealed no abnormality as well. As seen beforehand, there was no diagnosis-contributive positive test, which made establishing a diagnosis quite an ordeal. The patient had bilateral floppy ears (Fig. 2), and along with the presence of hearing loss and a history of painless red eyes, the question of a possible relapsing polychondritis underlying condition was raised. And since the Michet et al. diagnostic criteria were fulfilled (Auricular cartilage inflammation, ocular inflammation and hearing loss), a limbic encephalitis diagnosis associated to RP was established. Treatment consisted on administering high dose of intravenous methylprednisolone (1g per day) for five days, relayed by tapered doses of oral corticosteroids starting by 1mg/kg per day and an immunosuppressant agent (Azathioprine) at the dose of 2 mg/kg per day. After one month, his MMS exam score went up to 28, along with significant improvement of his memory disorders, gait and balance problems.

Discussion:

Relapsing polychondritis, usually seen between the fourth and the fifth decade of age, is characterized by a pleomorphic presentation, chondritis of auricular, nasal and Laryngotracheobronchial cartilage are the most frequent clinical features, followed by involvement of proteoglycan-rich tissues such as the eye, heart valves and blood vessels [1]. Auricular chondritis is the most common present and inaugurating manifestation of RP, it is seen in up to 90% of patient. Repeated flares could severely damage the cartilage and make the pinna loses its shape and becomes flabby [2]. Hearing loss is been observed in 46% of RP cases, it could be conductive or sensorineural [1]. The latter type is a result of vestibular structures inflammation or a vasculitis of the internal auditory artery, vestibular damage could lead to ataxia, vertigo, nausea and vomiting [2]. In the matter of ocular involvement, episcleritis is the most frequent manifestation, followed by scleritis and conjunctivitis. Less commonly, RP can be the cause of iritis, retinopathy, anterior uveitis, retinal vasculitis and ischemic optic neuritis [1]. Neurologic manifestations are rare and account for only 3% of RP patients. Clinical presentation include headache, meningitis, limbic encephalitis, cerebral infarction, confusion and dementia [3]. CNS involvement is reported to be secondary to systemic vasculitis by Stewart et al. [4]. The most recent diagnostic criteria of RP were proposed by Michet et al. in 1986 [5], they consist of confirmed inflammation in two of three cartilages among auricular, nasal and laryngotracheal OR proven inflammation in one of the aforementioned cartilages plus two other minor criteria among hearing loss, ocular inflammation, vestibular dysfunction, seronegative arthritis. Although there is still no guidelines for treatment of RP, many therapeutic options are available. They include the use of non-steroid anti-inflammatory drugs (NSAIDs) in non-severe forms of RP in order to control pain of nose, ear or joints inflammation. In NSAIDs resistant or severe forms of RP, corticosteroids are the treatment of choice. Resorting to immunosuppressive agents and biologics comes as a second line treatment in corticosteroid-intolerant patients or life-threatening complications of RP. Surgical and interventional procedures are used in cases

of bronchial stenosis, valve regurgitation or aortic aneurysms [1]. Evaluation of RP disease activity throughout its evolution is aided by the Relapsing Polychondritis Disease Activity Index that has been established in 2012 [6]. Causes of death during RP are related to organ damage complications, concomitant diseases and infections that are usually due to the immunosuppressive treatments [1].

Conclusion:

Relapsing polychondritis is a rare and potentially severe autoimmune disease with variable clinical presentation. Diagnosis is based on numerous clinical criteria, but no consensus whatsoever is been established on management protocols, however, the emergence of biologics along with a scoring system called the Relapsing Polychondritis Disease Activity Index (RPDAI) seems to have favorable impact on RP management.

Declarations:

Funding: No funding was received to work on or publish this case report.

Ethics Approval and Consent to Participate: Informed written consent was taken from the patient. Internal review board approval not needed as it is a case report.

Consent for publication: Written informed consent was obtained from all individual participants included in the study

Competing Interests: The authors declare that they have no conflict of interest.

Author’s contribution: Rachid Belfkih, Omar Ghomari Khayat and Fatima Zohra El Amrani wrote and revised critically the manuscript.

References:

1. Borgia F, Giuffrida R, Guarneri F, Cannavò SP: Relapsing Polychondritis: An Updated Review. Biomedicines. 2018 Aug 2, 6:84.
2. Sharma A, Gnanapandithan K, Sharma K, Sharma S: Relapsing polychondritis: a review. Clin Rheumatol. 2013 Nov, 32:1575-83.
3. Kashihara K, Kawada S, Takahashi Y: Autoantibodies to glutamate receptor GluRepsilon2 in a patient with limbic encephalitis associated with relapsing polychondritis. J Neurol Sci. 2009 Dec, 15:275-7.
4. Stewart SS, Ashizawa T, Dudley AW Jr, Goldberg JW, Lidsky MD: Cerebral vasculitis in relapsing polychondritis. Neurology. 1988 Jan, 38:150-2.
5. Michet CJ Jr, McKenna CH, Luthra HS, O'Fallon WM: Relapsing polychondritis. Survival and predictive role of early disease manifestations. Ann Intern Med. 1986 Jan, 104:74-8.

6. Arnaud L, Devilliers H, Peng SL, et al.: The Relapsing Polychondritis Disease Activity Index: development of a disease activity score for relapsing polychondritis. Autoimmun Rev. 2012 Dec, 12:204-9.

Figures

Figure 1

Fluid-attenuated inversion recovery (FAIR) sequence showing bilateral hypersignal lesions, poorly bordered, in medial temporal, insular and internal frontal lobes.

Figure 2

Damaged ear cartilage secondary to anterior auricular chondritis.