Auricular polychondritis presenting as non-infectious, non-paraneoplastic limbic encephalitis

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Abstract

Relapsing polychondritis (RP) is a rare, recurrent autoimmune inflammation of unknown etiology affecting cartilaginous structures, however, neurologic complications can occur. A 60 year old gentleman presented with 2 months of progressive cognitive decline and neuropsychiatric symptoms. Lumbar puncture showed a lymphocytic pleocytosis negative for infection and brain MRI revealed T2-weighted hyperintensities in bilateral mesial temporal lobes. He had no response to empiric antibiotics or antivirals for presumed limbic encephalitis. His ears were later noted to have significant edema and erythema and subsequent biopsy was consistent with auricular chondritis. Both his auricular inflammation and cognition partially responded to aggressive immunosuppression.

Our patient had auricular polychondritis presenting as non-infectious, non-paraneoplastic limbic encephalitis. With the steady increase in the prevalence of dementia worldwide, this case highlights the expanding need for increased recognition and earlier treatment of autoimmune and potentially treatable causes of limbic encephalitis. Though the patient had a partial clinical response to immunosuppression, subsequent imaging showed irreversible temporal lobe atrophy and he never returned to his prior baseline. Perhaps earlier diagnosis and therapy of the underlying autoimmune process could have improved his response and prognosis.

Keywords: Limbic encephalitis, Relapsing polychondritis, Auricular polychondritis.

ABBREVIATIONS: CSF- Cerebrospinal fluid; CT-Computed tomography; EEG- Electroencephalogram; FDG-PET- 18F-Fluorodeoxyglucose positron emission tomography; HSV PCR- Herpes simplex virus polymerase chain reaction; IV- Intravenous; IVIg- Intravenous immunoglobulin; MRI- Magnetic resonance imaging; RBC- Red blood cell; RP- Relapsing polychondritis; WBC- White blood cell.

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Relapsing polychondritis (RP) is a rare, recurrent, idiopathic autoimmune inflammation of cartilage including the nose, auricles, larynx, and trachea [1]. Less than 3% of all RP present with neurologic complications, most commonly cranial neuropathies [2,3]. Headaches, seizures, cerebellar ataxia, aneurysms, and aseptic meningitis have been described [2].

A previously healthy 60 year old man presented with 2 months of progressive headaches, cognitive decline, visual hallucinations, paranoid delusions, emotional lability and a 30 pound weight loss. He was alert, oriented to self and place, but had intermittent somnolence and agitation. He had a paucity of language with paraphasic errors. Strength and sensation were normal but reflexes were pathologically brisk. Intermittent twitching on his left face and diffuse action induced myoclonus were captured on video-EEG without underlying ictal activity.

Lumbar puncture revealed lymphocytic pleocytosis and elevated protein (WBC 140, 89% lymphocytes, RBC 285, glucose 48, protein 75). Empiric treatment for infectious encephalitis with ceftriaxone, vancomycin and acyclovir was given but workup was negative for CSF gram stain, cultures, HSV PCR, India ink and acid fast bacilli. Brain MRI revealed T2 hyperintensities in bilateral hemispheres including temporal lobes and hippocampi (Figure 1a). High dose methylprednisolone, followed by IVIg produced only mild improvement. Repeat LP showed WBC 66 (69% lymphocytes, 14% neutrophils, 11% monocytes), RBC <1,
glucose 64, protein 54. Extensive serological, CSF, CT chest/abdomen/pelvis and FDG-PET were unrevealing.

His ears were swollen, erythematous and non-painful (Figure 2b) but did not improve on antibiotics. Ear biopsy revealed eosinophilic change and predominantly lymphocytic infiltration with occasional neutrophils, consistent with auricular chondritis. Brain MRI had restricted diffusion (Figure 1b) indicative of a “prominent ear sign” previously described in RP encephalitis [4]. Rheumatology escalated immunosuppression with repeat methylprednisolone, IVlg and cyclophosphamide.

Following treatment, he became more alert, interactive and action myoclonus subsided. A week after repeat immunosuppression, he had resolution of his ear inflammation (Figure 2b). He was discharged on oral prednisone and monthly cyclophosphamide. Over the next 2 years, symptoms stabilized but recurred when immunosuppression was tapered. His neurologic exam has not improved further despite IVlg, methotrexate and infliximab. Repeat MRIs at 3 months and 1 year, revealed improvement in T2 hyperintensities but worsening of volume loss and parenchymal atrophy.

Classical clinical features of RP include bilateral auricular chondritis, ocular inflammation, nasal chondritis, dysfunction of the vestibulocochlear system, tracheobronchial chondritis, and polyarthritis [5]. The modified McAdam’s criteria for RP diagnosis includes at least 1 of the following: (a) 3 or more classical features, (b) 1 or more classical features with positive histological confirmation of chondritis, or (c) steroid-responsive chondritis at 2 or more anatomic locations [6]. Our patient fulfilled the second criteria for RP, with biopsy proven auricular chondritis.

While dapsone has also been induced response in RP encephalitis, our patient improved with steroids, cyclophosphamide and IVlg [6]. Attempts at weaning steroids to transition to steroid sparing agents have been unsuccessful, emphasizing the steroid-responsiveness of the disease. His clinical improvement was limited and repeat MRIs revealed diffuse brain atrophy; perhaps his response and prognosis would have been improved with earlier diagnosis and treatment. This case highlights the expanding need for recognition of autoimmune and potentially treatable causes of limbic encephalitis.

Review of the medical literature shows that RP with encephalitis is limited to case reports and the effect of immunosuppression on neurologic complications is not well established though there anecdotes of favorable responses [3,7]. There are no standard management protocols for RP let alone RP with nervous system

Figure 1. (a) MRI FLAIR showing T2 hyperintensities of the bilateral mesial temporal lobes (arrows). (b) Axial DWI MRI showing restricted diffusion of the right auricular cartilage (arrow).

Figure 2. (a) Swollen, edematous right ear before treatment. (b) Significant reduction in the erythema and edema after aggressive immunosuppression.
involvement. In addition, the epidemiology of RP, including racial distribution, is not well defined due to the disease rarity [1]. Our study is limited because it is a single case and the patient had an atypical neurologic presentation of an already rare disorder after two months of ongoing symptoms which further delayed diagnosis and treatment. Although previous studies have supported autoimmune mechanisms in RP, the etiology and pathogenesis of RP remain largely unknown so further studies are needed for neuropathologic examination, identification of pathogenic antibodies and to define optimal immunotherapy regimens.

References


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