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Case report

Relapsing polychondritis and lymphocytic meningitis with varied neurological symptoms

Policondrite recidivante e meningite linfocitária com sintomas neurológicos variados

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Introduction

Relapsing polychondritis is a rare autoimmune disease,[1,2] with a female–male ratio of 2:4,[3] with symptom's onset between 20 and 60 years of age (peak incidence around 40 y/o),[4] characterized by bilateral auricular and nasal chondritis, vestibular involvement and varied systemic symptoms due to recurrent and progressive inflammation of cartilaginous tissue and proteoglycan-rich structures in various sites of the body.[1,2] We report the case of a patient with relapsing polychondritis with several neurological manifestations.

Case report

A 69 year-old male with diabetes, hypothyroidism and dyslipidemia had a two-month history of swelling and pain of both ear lobes and edema and arthralgia of metacarpophalangeal joints and ankles, as well as generalized pain with a waxing and waning course. Twenty days prior to his admission in our hospital he started with ataxia, paraparesis, tinnitus, vertigo and confusion. He had been previously seen at another hospital, around the time of confusion onset, where he was treated for herpetic encephalitis following a lumbar puncture which disclosed elevated leukocytes, with a predominance of lymphocytes. Although at first he had an improvement of confusion, his paraparesis remained unaffected. On physical examination he had nystagmus in the downward gaze, rigidity of upper limbs, paraparesis, absent reflexes, tactile hypoesthesia, dysmetric movements, gross postural and action tremor, bradykinesia and truncal ataxia. He also had swelling and a purplish erythema of both ear lobes and arthritis in the metacarpophalangeal joints of the second and third fingers of the right hand (Fig. 1). Brain and cervical MRI disclosed a mild thickening of the dura (Fig. 1). A new lumbar puncture confirmed the presence of elevated leukocytes and laboratory exams disclosed augmented inflammatory activity and

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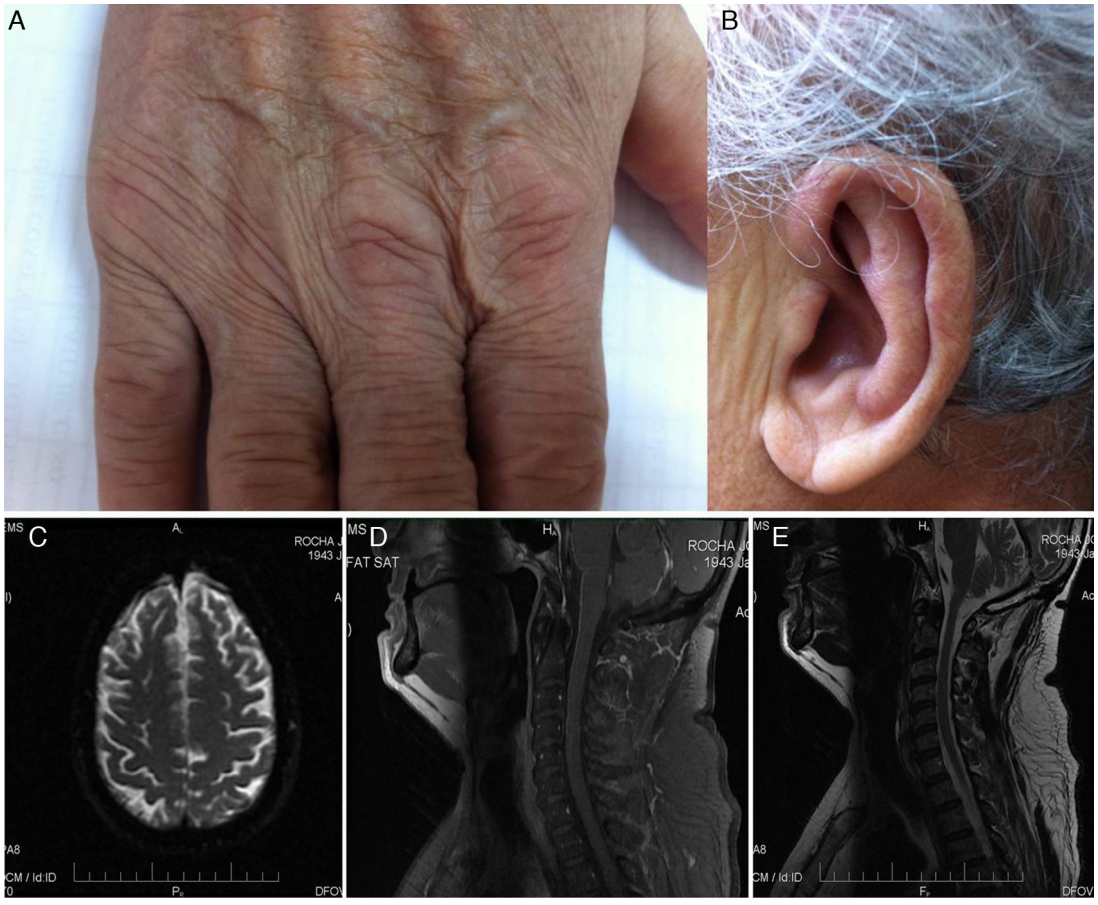

Fig. 1 – Rheumatological clinical findings (upper images): (A) edema of metacarpophalangeal joints; (B) ear lobe chondritis. Brain MRI findings (lower images): (C) axial diffusion with thickening of the dura of both frontal lobes; (D) sagittal T1 FSE FAT SAT with increased meningeal signal adjacent to the cerebellum; (E) T2 FSE showing mild cerebellar atrophy.

iron-deficient anemia (Table 1). A diagnosis of relapsing polychondritis was made based on the association of chondritis, arthritis and vestibular ataxia with predominant neurological symptoms. Following a course of Prednisone 1 mg/kg qd there was major improvement of chondritis, arthritis, ataxia and paraparesis, but the tremor remained unchanged. On a one-year follow-up visit he had developed several complications of chronic corticosteroid use, such as osteopenia, hypertension, Cushing's syndrome, worsening of obstructive apnea syndrome and one episode of bilateral cutaneous Herpes Zoster of the trunk. These complications warranted a change of immunosuppressive treatment from Prednisone to Methotrexate. In spite of this, he had no new neurological symptoms and remained with moderate ataxia.

Discussion

Relapsing polychondritis is a rare multisystemic autoimmune disease that affects cartilaginous tissue, especially hyaline cartilages at multiple sites, most often compromising the antihelix of both earlobes, with sparing of the lobule.[2,5] Seronegative polyarthritis and systemic compromise of other organs may also occur (including ocular inflammation, audiovestibular impairment, vasculitis, skin involvement, valvular insufficiency and neurological symptoms) due to compromise of proteoglycan-rich tissues.[6,7] Around 30% of the cases are associated with concurrent autoimmune disease, systemic vasculitis and myelodysplastic syndrome.[5,8] Diagnosis is made on clinical grounds, occasionally with pathology disclosing inflammatory compromise of affected cartilaginous tissue.[5,7] Currently, the diagnosis is made on the basis of demonstration of either chondritis in two of three sites (auricular, nasal, laryngotracheal); or one of these sites and two additional features, including ocular inflammation, audiovestibular damage, or sero-negative inflammatory arthritis.[1,9] There are no specific laboratory findings.[5,7] Neurological symptoms occur in a minority of cases (3%) and may range from compromise of cranial nerves to a more overt presentation with cerebellar compromise, seizures or other focal findings suggestive of cortical compromise. These are vasculitic in nature. Aseptic meningitis, with thickening of the meninges, lymphocytic meningoencephalitis, rhomboencephalitis and cerebral aneurysms can also occur.[1,10–12] In our case we made the diagnosis solely on clinical grounds, as there was evidence on physical examination of chondritis of both ear lobes, sero-negative polychondritis and neurological compromise with aseptic meningitis. About 25% of patients die in up to five years following diagnosis; laryngotracheal involvement and cardiovascular complications are the leading causes

Table 1 – Complementary exams.

Exams	Results
Bilateral X-rays of hands and knees	Normal
Angiotomography of chest, abdomen and pelvis	Normal
Upper gastrointestinal endoscopy	Normal
Colonoscopy	Small polyp
Echocardiogram	Normal
Protein electrophoresis (blood)	Normal
Calcium and inorganic phosphorus	Normal
Electroneuromyography	Normal
Anti-HIV, syphilis, hepatitis B and C	Negative
Rheumatoid factor, antinuclear factor, anticentromere factor	Negative
Erythrocyte sedimentation rate	53
Serum iron	16.3 (normal range: 59–158)
Total iron binding capacity	252.8 (normal range: 250–450)
Iron saturation ratio	6.4% (normal range: 20–50)
Ferritin	327.54 μg/l (normal range: 4.63–204)
Reticulocytes	11.8
Hemoglobin	11.8
VG	32.1
VCM	72
C3 and C4	Normal
ANCA	Negative
Spinal fluid	
Erythrocytes	0.3
Leucocytes	71 (97% lymphocytes, 3% monocytes)
Glucose	68
Protein	69.7
GRAM	Negative
BAAR	Negative
HTLV 1 and 2	Negative

of death.[1,4] Factors that have a negative impact on survival at the time of diagnosis include old age, anemia and laryngotracheal stricture.[1,2] Oral nonsteroidal anti-inflammatory drugs may be used to treat patients with arthralgias and mild arthritis. Standard immunosuppressive treatment starts with high doses of corticosteroids (Prednisone 1 mg/kg qd), which is later tapered off to a smaller dosage in patients with moderate to severe compromise. Methotrexate may be used as a second-line drug to avoid side effects of chronic corticosteroid treatment. Azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil and TNF-antagonists are other options.[5,11]

Conflicts of interest

The authors declare no conflicts of interest.

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