CASE REPORT

Vestibulocochlear dysfunction in a patient with rheumatoid disease and vasculitis

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Abstract
The case is described of a patient with rheumatoid disease and evidence of vasculitis who developed acute vestibulocochlear dysfunction resulting in total deafness, which was assumed to be secondary to a vasculitic process affecting the vestibulocochlear nerve. Treatment of the underlying vasculitic process, despite effecting a general improvement of the patient's rheumatoid disease, did not significantly improve the auditory dysfunction.

Case report
A 49 year old white woman initially presented in July 1988 with a flitting polyarthropy. A provisional diagnosis of rheumatoid arthritis was made but was not treated with disease modifying agents at that time. Shortly afterwards she developed scleritis of the left eye. She remained fairly well until May 1989 when she was admitted to another hospital with systemic symptoms of weight loss and general malaise. Examination at that time showed a generalised polyarthropy, nodules on both elbows, cervical lymphadenopathy, scleritis of the left eye, and a left sided conductive deafness. Nystagmus was present, most marked on looking to the left. There was sinus tachycardia at 120 beats/minute. Investigations showed a positive rheumatoid factor (1 in 10 240), a high platelet count (788x10^9/l), erythrocyte sedimentation rate 97 mm/h, negative autoantibody screen (including antinuclear antibodies). She was treated with a reducing course of oral steroids and initially seemed to make a good symptomatic improvement. In July 1989, however, she once again became unwell with a recurrence of her previous symptoms together with a sudden onset of deafness, vomiting, and dizziness. She was admitted to our hospital for further investigation and treatment. At that time her treatment included prednisolone 30 mg/day, lofepramine 70 mg twice daily, propranolol 40 mg/day (for anxiety), aspirin 300 mg/day, ranitidine 150 mg/day, and chloramphenicol eyedrops.

Examination at that time showed a pale thin woman who looked unwell. Communication had to be carried out by written questions and instructions. There was digital vasculitis, elbow nodules, scleritis of both eyes, and evidence of active synovitis. No abnormalities were found in the chest, cardiovascular system, or the abdomen. Examination of the central nervous system showed bilateral deafness. Auroscopy disclosed a normal right drum but the left showed evidence of chronic secretory otitis media. Visual acuity was diminished, and there was cataract formation (posterior capsular) in both eyes. Fundal examination was thought to be normal through the cataracts. Tone, power, sensation, and coordination of the limbs were normal. Reflexes were symmetrical and plantar responses downgoing.

Investigations were as follows: haemoglobin 107 g/l (hypochromic), white blood cells 10.6x10^9/l, platelets 870x10^9/l, erythrocyte sedimentation rate 80 mm/h. Rheumatoid factor was positive at a titre of 1 in 2560. Albumin was low at 28 g/l, and creatinine clearance was reduced at 49.7 ml/min. Electrophonics showed an increased IgA and IgM. Biopsy of a nodule showed changes in keeping with a rheumatoid nodule. Brainstem evoked responses were absent on the right at maximum output levels of 90 dB hearing loss. The left sided responses were similar, but there was thought to be some slight correlation at 90 dB hearing loss. The conclusion was of severe hearing loss on both sides at 2-3 kHz. Although a computed tomographic scan of the brain was normal, a magnetic resonance scan was performed. T1 and T2 weighted images (8 mm) were obtained in an axial plane with 7 mm sagittal T1 weighted and 5 mm coronal T1 weighted images through the internal auditory meatuses. The last images were taken after intravenous contrast. There were non-specific changes in the white matter of both hemispheres and evidence of left inflammatory mastoid disease. The internal auditory canals appeared normal.

The following results were normal or negative: urea and electrolytes, liver function, thyroid function, vitamin B-12, folate, autoantibody screen, double stranded DNA antibodies, C3 and C4 concentrations, hepatitis B surface antigen and e antigen titres, chest radiograph, rectal biopsy, lumbar puncture, and examination of urinary sediment.

She was treated initially with a pulse of intravenous methylprednisolone (1 g) and intravenous cyclophosphamide (2-5 mg/kg). Subsequently, treatment was continued with oral prednisolone at a dose of 30 mg/day and oral cyclophosphamide at a dose of 50 mg twice daily. There was a considerable improvement in her joints (symptoms and function) and scleritis. One month after discharge she had gained 6 kg in weight and was beginning to distinguish low pitched sounds, though she was still profoundly deaf. The feelings of dizziness had disappeared. Laboratory markers of disease activity confirmed the clinical impression of an improvement. She had, however, been referred for consideration of cochlear implants.
Discussion

Vestibulolauditory dysfunction has not to our knowledge been previously described in rheumatoid disease. The sudden onset of symptoms and their relation to indicators of disease activity suggest that they were due to the disease process itself, despite the fact that there was evidence of left sided mastoid disease. Vestibulolauditory dysfunction has been previously described together with non-syphilitic keratitis in patients with an underlying vasculitis (Cogan’s syndrome).\(^1\) In a review of 53 cases of Cogan’s syndrome\(^2\) none of the patients had rheumatoid disease, though one patient has been described with a positive rheumatoid factor.\(^3\) Apart from the vestibulolauditory dysfunction, our patient had none of the other features of Cogan’s syndrome, however. Neurological effects in rheumatoid disease may develop through a variety of tissue mechanisms or through structural changes.\(^4\) A review of the neuropathology of rheumatoid disease\(^6\) suggests that brain involvement is usually characterised by formation of rheumatoid nodules or vasculitis. Damage to the brain stem most commonly results from narrowing of the bony canal, leading either to direct compression of neural tissue or to compromise of its vascular supply. Computed tomography and magnetic resonance imaging in our patient failed to show evidence of brain stem compression or infarction. There was, however, indirect evidence of cerebral vasculitis as adjudged by the white matter abnormalities on the magnetic resonance scan. The exact mechanism of the vestibulolauditory dysfunction remains unclear, though the most likely explanation for the VIIIth nerve lesion is epineural arterial vasculitis. Indirect evidence that vasculitis is the pathological process comes from observations of deafness in other systemic vasculitides—notably, polyarteritis nodosa, Wegener’s granulomatosis,\(^6\) and temporal arteritis.\(^7\) Although epineural arterial vasculitis seems a likely cause of the VIIIth nerve lesion, it has been suggested (after examination of temporal bone sections) that vasculitis of the internal auditory artery may cause deafness.\(^3\) Observations also suggest that early treatment of the vasculitic process with combined cytotoxic-immunosuppressive drugs can improve the hearing loss in Wegener’s granulomatosis.\(^9\) It would, therefore, seem appropriate to continue treating our patient in this manner, though at present there is little evidence of recovery.