Sudden Onset Visual Impairment and Deafness in a Patient with “Long Standing Rheumatoid Arthritis”

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CASE

P, a 52 years housewife was seen by us for the first time on 1st October, 2001. She had polyarticular pains and swelling since 20 years, involving small and large joints symmetrically. She had been diagnosed to suffer from rheumatoid arthritis and had received non-steroidal antiinflammatory drugs (NSAIDs), steroids and ayurvedic treatment intermittently over the past 20 years but had never received disease modifying antirheumatic drugs (DMARDs) for her illness. She had progressively become disabled and walked with an aid. One month before she saw us she developed sudden onset bilateral progressive blurring of vision. There was minimal pain in the eyes but no local redness or dryness. She was seen by an ophthalmologist who diagnosed iridocyclitis and treated her with local steroid instillations along with oral steroids with improvement in 3-4 days. One week later she developed sudden onset bilateral hearing loss accompanied by vertigo. There was no history of fever, headache, vomiting, otalgia, otorrhoea, cough or dyspnoea. There was no history of diabetes mellitus, hypertension or ingestion of ototoxic drugs. Audiogram revealed bilateral sensorineural hearing loss. She was started on oral prednisolone 30 mg/d with partial improvement of hearing and decrease in vertigo. She was seen by us 15 days later while still on 30 mg/day of prednisolone.

On examination, she had Boutonierre’s deformity of fingers, wrists were fixed, shoulders had marked limitation of movements and knees had secondary osteoarthritis. There was pallor, no lymphadenopathy, pedal oedema, skin lesions or subcutaneous nodules, salivary or lacrimal glands were not enlarged. The vision was normal and there was no sicca or erythema. There was marked loss of hearing without signs of meningitis, other cranial nerve palsies or peripheral neuropathy. Examination of other systems was normal.

DISCUSSION

The patient had a history of chronic arthritis involving small and large joints symmetrically. There was joint damage resulting in deformities. This is the typical picture of long standing rheumatoid arthritis (RA) and therefore a clinical diagnosis of RA is justified. RA is characterised by Th1 overactivity. The rheumatoid synovium is infiltrated by CD4 positive Th1 lymphocytes. Though a disease with predominant articular manifestations, RA is a systemic disease.

She developed sudden onset blurring of vision. Common ocular manifestations of RA are keratoconjunctivitis sicca, episcleritis and scleritis. Loss of vision is due to corneal involvement or cataract. Corneal involvement can be secondary to episcleritis or scleritis. Other causes of visual loss include ocular infections and thromboembolism of retinal vessels due to associated antiphospholipid syndrome. None of these, except thromboembolism would produce sudden visual loss. Further, the patient had iridocyclitis. Uveitis is not a feature of RA. RA therefore does not explain her visual symptoms.

One week later, she developed sudden onset bilateral hearing loss accompanied by vertigo. There was no evidence of ear infection. Patient did not have headache, vomiting, papilloedema or involvement of other cranial nerves to indicate an intracranial pathology like a space occupying lesion or meningitis. About 38% of RA patients develop erosions and shortening of the ossicular chain which can result in a conductive hearing disturbance and not sensorineural loss as seen in this patient. Further the hearing loss would not be sudden. Sensorineural hearing loss, due to vasculitis, has been uncommonly described in RA. Vasculitis is a potentially serious complication of RA, seen in less than 1% of patients. It involves small blood vessels and is typically seen in seropositive, aggressive and long standing RA and is more common in males. The patient had not received proper therapy throughout her illness and vasculitis as a cause of her sensorineural deafness can be suspected. This still does not explain iridocyclitis, which was temporally a part of the present problems.

McCabe has described a distinct clinical entity, autoimmune inner ear disease, that is characterised by bilateral, asymmetrical and rapidly progressive hearing loss. It is usually an isolated condition and a diagnosis of exclusion. Cogan’s syndrome is another disorder that is characterised by visual blurring, hearing loss and vertigo. Cogan’s syndrome is associated with interstitial keratitis and
not iridocyclitis and is usually seen in young adults. Behcet’s disease is associated with uveitis and can cause sensorineural hearing loss. The patient did not have aphthae or genitourinary ulcers. Arthritis in Behcet’s disease is usually oligoarticular involving knees, ankles, wrists in a recurring episodic manner and is seldom destructive. Another disorder that can cause arthritis, uveitis and sensorineural hearing loss is Wegener’s granulomatosis (WG). There was no history of recurrent upper respiratory tract affection and arthritis in WG is non-destructive. Hence WG too seems unlikely. The patient did not have clinical features of any other autoimmune disease to explain her audio-visual symptoms. With a diagnosis of RA and a suspicion of vasculitis, the patient was investigated.

CASE (Contd)

Investigations showed: haemoglobin 9.6 g/dl, total white cell count 7700/mm³ with a normal differential count, ESR 35 mm/hr, normal liver and renal profiles, rheumatoid factor negative, S. calcium 9.1 mg/dl (8.5-11 mg/dl), S. phosphorus 5.4 mg/dl (2.1-6.5 mg/dl), fasting blood sugar 150 mg/dl, post-prandial blood sugar 272 mg/dl. Her chest radiograph showed bilateral hilar lymphadenopathy with reticulo-nodular shadows (Fig. 1). X-rays of the hands showed bilateral carpal destruction (Figs. 2 and 3). A repeat ophthalmological examination showed posterior synechiae. CT scan of the chest revealed multiple lobulated lymph nodes in both hilar regions. Reticular shadows were seen in both mid and lower zones.

DISCUSSION

This was an unexpected turn of events. Causes of bilateral hilar lymphadenopathy include tuberculosis, sarcoidosis, coccidiodomycosis, histoplasmosis, lymphoma, leukaemia, chronic berylliosis and hypogammaglobulinaemia. The patient did not have any evidence of infection in the form of fever, loss of appetite or weight. Despite this, TB is always a concern in India. Iridocyclitis is a feature of TB. Sensorineural hearing loss is infrequently seen in TB meningitis and is usually chronic. Coccidioidomycosis and histoplasmosis are not common in India. The peripheral blood picture did not suggest leukaemia. Lymphoma is not associated with iridocyclitis or isolated sensorineural hearing loss. The patient was a housewife and had no exposure to beryllium. There was no history of recurrent infections and hence hypogammaglobulinaemia is unlikely. Bilateral hilar lymphadenopathy is the classical radiological feature of sarcoidosis. Sarcoidosis explains both iridocyclitis and sensorineural hearing loss. In a series of 37 patients with neurosarcoidosis, cranial nerve palsies were seen in 52% patients. Two patients had hearing loss of which one had isolated hearing loss as the sole manifestation of neurosarcoidosis. Granulomatous meningitis directly infiltrates the cranial nerves and the eighth nerve is the fourth most commonly affected cranial nerve. Response to steroids is good. Hearing loss can fluctuate and may be the initial manifestation of neurosarcoidosis. With a provisional diagnosis of sarcoidosis the patient was investigated further.

CASE (Contd)

A transbronchial lung biopsy was performed. It showed multiple non-caseating epithelioid granulomas with no acid
Dyspnoea was not a very reliable symptom in our patient as she was severely incapacitated and not very ambulant. There were no skin lesions. Parotid or lacrimal glands were not enlarged. A minor salivary gland or conjunctival biopsy could have helped to make a diagnosis of sarcoidosis. These are easily accessible sites but carry a poor sensitivity especially when performed in patients without clinical evidence of tissue involvement in the form of glandular enlargement or conjunctival nodules. A transbronchial biopsy was therefore performed. It showed non-caseating granulomas without acid fast bacilli. The patient could not perform pulmonary function test.

Besides sarcoidosis, non-caseating granulomas are seen in a variety of conditions like tuberculosis, drug hypersensitivity, occupational exposure to beryllium etc. A diagnosis of sarcoidosis is made by exclusion of these conditions. TB is usually associated with caseating granulomas and acid fast bacilli. Interstitial parenchymal involvement is not a feature of TB. Isolated bilateral acute sensorineural deafness as the only form of meningitis is an uncommon feature of TB in the absence of ototoxic antituberculous drugs. One case of acute sensorineural hearing loss secondary to tuberculous meningitis has been recently reported. The authors claim this to be the only reported case of acute sensorineural hearing loss as a presentation of tuberculous meningitis. A cerebrospinal fluid examination however was not performed in our patient. The patient was on ayurvedic medications, the contents of which were not known. Bilateral hilar lymphadenopathy is not a feature of drug hypersensitivity.

One question arises automatically. Did she have chronic sarcoid arthritis all along? Sarcoidosis can present as chronic arthritis akin to RA. Joint involvement can be secondary to bone sarcoidosis or synovitis. Joint symptoms have been reported to occur in 15% to 25% of patients with sarcoidosis. Articular manifestations in sarcoidosis could be due to osseous sarcoidosis or synovitis. Bone involvement in sarcoidosis could be due to osseous sarcoidosis or synovitis. Bone involvement in sarcoidosis varies from 1% to 13% in different series. James et al have noted that patients with bone sarcoidosis usually have chronic sarcoidosis in other sites such as lungs, skin and eyes. Destructive arthritis has been described in sarcoidosis. According to Petersson joint destruction is uncommon with chronic sarcoïd synovitis, though osseous sarcoidosis can result in joint destruction. It is difficult to assess at this juncture whether joint damage is secondary to an underlying bony pathology in this patient. Most patients with chronic sarcoid arthritis have high serum ACE levels and this helps to distinguish chronic sarcoid arthritis from other chronic inflammatory polyarthritides. ACE levels were normal in this patient but this could be due to prior use of steroids. Pulmonary sarcoid can be asymptomatic and only discovered on routine investigations. One could therefore say that the patient had sarcoidosis of joints, lungs and CNS, especially as rheumatoid factor was negative and destructive.
RA is usually seropositive. Review of the patient’s old reports showed a persistent seronegativity. A synovial biopsy can be diagnostic if noncaseating granulomas can be demonstrated. However, after a period of 20 years, a synovial biopsy is more likely to show a chronic fibrotic reaction. Unfortunately old chest and joint radiographs were not available for review.

CASE (Contd)

The patient was continued on 30 mg/d of prednisolone and antidiabetic treatment with glibenclamide was started. She was shown gradual improvement in hearing over the past three months. Long term management of disease and rehabilitation is planned.

DISCUSSION

Sarcoidosis has been described in association with a variety of autoimmune diseases like SLE, rheumatoid arthritis and scleroderma. In this situation sarcoidosis, which itself is a Th1 dysfunction disorder, runs an independent course and requires separate treatment. A variety of autoantibodies like antinuclear antibodies, rheumatoid factor are seen in sarcoidosis. These autoantibodies may be a part of hypergammaglobulinaemia due to polyclonal B cell activation. Their significance is not clear. Further, sarcoidosis is a systemic disease with protean manifestations which can mimic a rheumatic disorder.

CASE (Contd)

Fifteen days ago (3.5 months after presentation), she complained of tingling numbness in both ring and little fingers. X-ray of the cervical spine showed an atlanto-axial dislocation (AAD) (Fig. 4) and symptoms improved with use of a cervical collar.

DISCUSSION

Rheumatoid arthritis is the commonest cause of non-traumatic AAD. Anterior AAD has been reported to occur in 19-71% of patients of RA. Vertebral sarcoidosis is rare and only 20 cases have been reported in literature. The commonest site of vertebral sarcoidosis is the lower thoracic and upper lumbar vertebrae and not cervical spine.

In conclusion, this is a case of RA, and sarcoidosis. The diagnosis of RA in this case is based on the presence of chronic, symmetric, erosive polyarthritis with atlanto-axial dislocation. Sarcoidosis was diagnosed on the basis of bilateral hilar lymphadenopathy, uveitis and presence of non-caseating granulomas on transbronchial lung biopsy which are not explained by RA. As reported earlier by Kucera, an association between RA and sarcoidosis is not commonly appreciated.

REFERENCES


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**Announcement**

Are you a Physician interested in Thyroid Practice and Information?
Formation of Proposed Indian Thyroid Association

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