Conductive Hearing Loss in Systemic Lupus Erythematosus: A Case Report with Review of Literature

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Abstract

A 33 year- old white man with signs and symptoms of Systemic Lupus Erythematosus (SLE) confirmed by laboratory, serological and immunological tests. The patient developed typical picture of Conductive Hearing Loss (CHL) of otosclerosis 3 years after diagnosis of SLE. Previous studies reported association of Sensorineural Hearing Loss (SNHL) and SLE. The purpose of this report is to describe the first case of bilateral CHL in association with SLE. Relevant literature was reviewed and possible autoimmune involvement of middle ear was speculated.

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Keywords

Conductive hearing loss, otosclerosis, systemic lupus erythematosus.

Introduction

Systemic Lupus Erythematosus (SLE), the prototype immune complex disorder, is a multisystem disease affecting the connective tissue (collagen) of unknown etiology. It is characterized by the presence of autoantibodies, and the circulating immune complexes can produce immunologically mediated tissue injury to multiple organs including skin, kidneys, heart, lung, joints, central nervous system and serosal surfaces.[1- 3] However, otologic involvement in SLE has been reported and it was confined only to the inner ear with the end result of sensorineural hearing loss. [4- 8]

It does not appear in the literature that there are any reports on the association between Conductive Hearing Loss (CHL) and SLE.

The present report concerns a white man who had clinical and serological findings of SLE whose otological examination and hearing evaluation revealed typical picture of bilateral CHL of otosclerosis.

Case Reports

A 33 year old white Jordanian man was diagnosed to have SLE. The diagnosis was established on the basis of polyarthritis, renal disease, positive skin biopsy, hemolytic anemia with leucopenia, positive Antinuclear Antibodies (ANA), pericardial serositis, gingival bleeding and ulcers, facial rash, elevated Erythrocyte Sedimentation Rate (ESR), and present immune complexes.

Clinical Profile: The patient has been well during the past two years, complained of rash around his mouth and gums, moderate retrosternal pain when lying in bed, intermittent fever, flitting arthralgia and an episode of pericarditis.

The patient was hospitalized for loss of appetite with loss of weight of about 11 Kg in the last month, polyarthralgia, rash over the chest and face and recurrent fever. Two weeks prior to admission, he had abdominal pain, nausea and vomiting. On physical examination, he looked pale and ill-appearing. Temperature 40.4° C; gingivitis; red macular rash over trunk, face, neck and arms; large mobile lymph node in the right axilla and a smaller one in the left axilla; small node in the left neck; blood pressure 110/70 mm Hg; pulse 120 regular; and the rest of examinations were normal. No evidence of infection was found what so ever. Sternal bone marrow, right quadriceps muscle biopsy, lip biopsy excluded malignancy, polyarteritis nodosa and Sjögren's syndrome, respectively. Chest and abdominal x-rays were normal. Biopsy of lymph nodes showed reactive hyperplasia. Kidney biopsy was suggestive of tubulo-interstitial disease. Skin biopsy showed mild vasculitis.

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Laboratory Findings: Diagnostic laboratory and serological tests disclosed a White Blood Cell (WBC) count of 4000, hemoglobin 8 gm/dl ml blood, hematocrit 33%, platelets 180,000/mm^3, ESR 114 mm/hour, serum creatinine 4.1 mg/dl, blood urea nitrogen (BUN) 114 mg/dl, uric acid 9.3 mg/dl, Na^+ 131 mM, K^+ 5.5 mM, HCO_3^- 17.3 mM, Calcium 8.6 mg/dl, total serum protein 9.0 gm/dl with albumin 3.3 gm/dl. Urinalysis revealed a specific gravity of 1.010, WBC 8-10. The ANA was speckled with a titer of 1:1280, cryoglobulins strongly positive, immune complexes were present, C_3 62 mg/dl (normal 60-130), C_4 8 mg/dl (normal 12-42), the anti-DNA was negative, the LE cells was negative. Liver function tests were normal. The final diagnosis was SLE with episode of pleural effusion.

The patient was treated with prednisolone 60 mg daily with remission of symptoms and improved renal function (creatinine 1.2 mg/dl). The dose was reduced to 20 mg per day and since then he had many episodes of exacerbation of the disease.

Audiologic Findings: The patient presented to the audiology clinic complaining of gradual loss of hearing over the last 3 years after the diagnosis of SLE disease. He complained of bilateral gradual hearing loss and tinnitus in both ears. There was no history of vertigo or other otologic diseases. Routine Ear- Nose- Throat (ENT) examination was normal, Rinne tests were negative on both sides and Weber test in the center. His initial audiogram revealed a moderate conductive hearing loss of 48 and 50 dB pure tone average at 500 to 4000 Hz at the right and left ears, respectively. The second audiogram was taken six months later showed further deterioration of hearing, 52 and 54 dB conductive loss at the right and left ears, respectively (Fig. 1). The smallest air-bone gap was noted at 2000 Hz, while the gap was wider above and below. Bone conduction levels showed mild loss at 2000 and 4000 Hz and were higher than the previous levels. Speech tests were consistent with moderate conductive hearing loss bilaterally. Impedance audiometry showed reduced compliances (0.3 ml) bilaterally (Fig. 2). Both ipsi-lateral and contra-lateral acoustic reflexes were absent. Bithermal caloric tests showed normal vestibular responses at both ears. Recruitment and tone decay tests showed no abnormalities.

Brain stem which evoked response audiometry was consistent with conductive hearing loss bilaterally. Exploratory tympanotony was suggested and the patient did not show up since then.

Figure (1): Audiogram showing bilateral moderate conductive hearing loss.

Figure (2): Tympanogram showing bilateral low compliance of tympanic membranes.

Discussion

SLE is a chronic autoimmune disease characterized by B-cell hyperactivity and can affect virtually any organ system. Autoimmunity to type II collagen is found in many diseases including rheumatic arthritis, relapsing polychondritis, systemic sclerosis, polyarteritis nodosa, Cogan's syndrome and Sjögren's syndrome. Otosclerosis, which is a disorder of the human otic capsule and stapes, has been reported to be caused by autoimmune reaction to type II collagen as a major cause. [9-11] This type of disorder causes CHL. SLE has been reported to be associated with Sensorineural Hearing Loss (SNHL). [5- 8,12,13] Kastanioudakis et al [14] reported SNHL in 8 patients and unilateral CHL in 1 patient out of 38 screened SLE patient. This author did not mention the pathology lying behind the CHL in his patient. The case reported here is the first of bilateral CHL, probably due to autoimmune otosclerosis, in association with SLE in which otological examination and audiological findings were highly suggestive to otosclerotic disorder. This patient has negative family history of CHL of any type or otosclerosis.

The stimulus behind the development of atoimmune disorders remains unknown. In SLE, there is an excessive production of autoantibodies resulting in Immune Complex (IC) formation leading to inflammation and tissue damage [15,16] with local IC deposition in many cases [16] and infiltration with destruction and fibrosis. The occurrence of SLE and CHL (middle ear structure involvement) in this reported case raises the question of association or coincidence. Supportive evidence for this association comes from the results of many studies on SLE patients: deposits of immunoglobulins (IgG, IgM, and IgA) and complement C_3 were present along the resorption lacunae besides osteocytes and chondrocytes around the destructive process of the otic capsule and otosclerotic stapes, [17,18] granular deposition of IgM, IgG and fibrin in involved skin [19] significant Temporomandibular Joint (TMJ) involvement secondary to osteoarthrosis [20] and other joints deformities typical of SLE arthritis, [21, 22] nodules in tendons with fibrous material, [23] periarticular or diffuse osteoporosis, [24] and periarticular and soft tissue calcification. [24, 25]

The etiopathogenetic mechanism of otosclerosis is not fully understood and is believed to be a multifactorial disorder: hereditary and genetic causes have been widely accepted in the development of otosclerosis and account for 50 percent of cases [26] autoimmunity reaction to type II collagen has been reported as a major cause of otosclerosis [9-11] and a strong association between measles virus and otosclerosis. [27- 29]

In light of the results of previous studies [9-11,17-25] and the history and findings in this reported case; middle ear ossicular joints, muscles, tendons, and stapes footplate could be a target for SLE induced disease. Although conclusive evidence is lacking, audiologic manifestation of CHL in this SLE patients could be due to IC deposition in middle ear structures and infiltration with destruction and fibrosis in the region between bone and cartilage of the otic capsule and footplate of stapes resembling otosclerotic process, autoimmune osteoarthrosis of the middle ear ossicular joints, or middle ear soft tissue (muscles and tendons) and periarticular calcifications.

In conclusion, whatever the cause of CHL, the observation and findings in this patient suggest that the association of middle ear involvement, probably autoimmune otosclerosis, with SLE has not been previously reported. Further investigation on temporal bones, specimens of similar cases and exploratory tympanotony are needed to better delineate the pathologic characteristics of middle ear involvement in SLE. The question of whether CHL due to middle ear autoimmune involvement and SLE are somehow linked remains intriguing but unanswered.

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ضعف السمع التوصيلي في مرض اللوشب الاحمراري الجهازي
(تقرير حالة مرضية ومراجعة الأدبيات)

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ملخص:

هذا تقرير مرضي لرجل ابيض ، عمره 33 سنة يشكو من أعراض وظواهر مرض اللوشب الاحمراري الجهازي (أحد أمراض المناعة الذاتية) حيث تأكد تشخيصه بفحوصات مخبرية، مصلية ، ومناعية.

ظهر على المريض الصورة النموذجية لضعف السمع التوصيلي لمرض تصلب الأذن بعد ثلاث سنوات من تشخيص مرض اللوشب الاحمراري الجهازي.

أفادت الدراسات السابقة أن هناك علاقة بين مرض اللوشب الاحمراري الجهازي وضعف السمع الحسي العصبي.

يهدف هذا التقرير لوصف أول حالة لضعف السمع التوصيلي الثنائي الجانب في مريض اللوشب الاحمراري الجهازي.

لقد تم استعراض الأدبيات الطبية ذات العلاقة وتم توضيح احتمالية شمول الأذن الوسطى لمرض المناعة الذاتية.

مفتاح الكلمات:

ضعف سمع توصيلي، تصلب الأذن، اللوشب الاحمراري الجهازي.