NOCARDIAL CEREBELLAR ABSCESS IN A CASE OF SYSTEMIC LUPUS ERYTHEMATOSUS RECEIVING LONG TERM CORTICOTHERAPY

H. Saberi1, S.A. Fakhr Taatabali2 and Z. Hussainkhan2

1) Department of Neurosurgery, Imam Khomeini Hospital, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran
2) Department of Anaesthesiology, Imam Khomeini Hospital, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

Abstract - Central nervous system ranks among the favorite sites to get involved by Nocardia asteroides. This opportunistic organism complicates many disorders characterized by cellular or humoral immunity disturbances.

A 38-year-old woman is being presented, known to have systemic lupus erythematosus (SLE) and having received a two year course of corticosteroid treatment. Cerebellar abscess was found on diagnostic imaging performed because of headaches and disequilibrium of two weeks duration.

Craniotomy was performed and the abscess excised. Nocardia asteroides grows in the pus culture. On switching the empirical therapy to a combination of trimethoprim-sulfamethoxazole and vancomycin, the patient exhibited dramatic improvement in symptoms later on and postoperative imaging showed an absence of the lesion. We recommend a high index of clinical suspicion for nocardia abscess in patients suffering from SLE and presenting with neurological symptoms, in whom a surgical step coupled with an antibacterial specific antimicrobial treatment presents the only plausible option.


Key Words: Nocardia, cerebellar abscess, CNS lupus, systemic lupus erythematosus

INTRODUCTION

Nocardiosis is a localized or disseminated infection caused by a soil-borne aerobic actinomycete, usually introduced through the respiratory tract. The pulmonary course may be subclinical or may provoke an acute or chronic process. Hematogenous dissemination spreads particularly to the nervous system and soft tissues (1).

In 1898 Nocard described an aerobic acid-fast organism which he isolated from cattle suffering from bovine farcy, a granulomatous disease of head and neck which occurred in Guadeloupe and the organism was named Nocardia Farciniae after him. Soon it became evident that most infections in humans were due to Nocardia asteroides, which has been described much more extensively ever since.

Many antecedent conditions are particularly associated with dysfunction of cellular immunity, but immunoglobulin and leukocyte defect may also serve as preconditions (2).

Among various predisposing factors for nocardiosis, both long term corticotherapy and systemic lupus erythematosus (SLE) have been implicated (3). Although coexistence of both factors in the single patient could have contributed to the development of the overwhelming infection, yet the exact influence of each factor in the promotion of the clinical picture remains a matter of dispute.

Cerebral involvement by SLE (cerebral lupus) commonly complicates its clinical course while nocardial brain abscess is exceedingly rare but remarkable. Therefore one may simply overlook the infectious nature of the neurological symptoms leading to poor outcome.

While the dismal outcome of systemic and cerebral nocardiosis has been emphasized, nevertheless timely diagnosis and proper treatment could render in successful results as in our patient (1). The necessity of craniotomy and total excision of the abscess followed by proper parenteral antibiotics as the keystones for a hopeful result could not be overemphasized.

CASE REPORT

A 38-year-old lady was referred to this center in 1998 for headaches, ataxia and progressive disturbance in mental state. She had been a proven case of SLE for 2 years and had been receiving corticosteroids (60 mg/day of prednisolone PO). Two years back, she reported with non-deforming polyarthritis, malar rash and peripheral edema. Investigations conducted at that
Fig. 1. Contrast enhanced computed tomogram. A: Preoperative image revealing the abscess cavity with an enhanced rim. B: Postoperative image obtained six months later depicting absence of the lesion with residual scarring.

Fig. 2. T2 weighted magnetic resonance imaging of the patient revealing multiple hyperintense supratentorial lesions (A) and multifoculated cerebellar abscesses compressing the fourth ventricle (B).
time revealed proteinuria > 1 g/24h, ESR = 76, low C3, C4, CH50 and a positive anti-nuclear and anti-DNA antibodies. Following one year of corticosteroid therapy, the ESR had reached 30, serum complement became normal, ANA titer decreased slightly and anti-dDe-DNA became negative. The patient's clinical condition showed a spectacular improvement. A year later (present illness) patient presented with headaches, confusion, dysarthria and lateralizing signs, however there was no evidence of arthritis or peripheral edema. Investigations showed an ESR of 50, CRP**, a negative VDRL test and no proteinuria. A contrast enhanced computed tomogram followed by magnetic resonance imaging of the brain was performed which revealed multifocal cerebellar hemorrhagic lesion with ring enhancement (Fig. 1A) and multiple deep supratentorial hyperintense enhancing lesions with minimal surrounding edema (Fig. 2A&B). There had been no history of otitis media or sinusitis. Heart auscultation was normal but echocardiographic examination revealed mild pericardial effusion. A chest X-ray did not reveal any parenchymal abnormality. Despite initiation of empirical therapy, her neurological status deteriorated progressively to coma and an onset of left sided hemiplegia. Emergent surgery was performed and the abscess was totally excised through a posterior fossa craniectomy. Post-operative improvement was modest and microscopic examination of the pus revealed acid fast Gram positive filaments, which on culture proved to be Nocardia asteroides. Thus, sulfamethoxazole-trimethoprim (800/160 mg q6h) and vancomycin 500 mg q12h were started. Dramatic clinical improvement was observed soon after employment of proper antimicrobial treatment confirming the diagnosis. The medications were continued intravenously for 45 days followed by oral sulfamethoxazole-trimethoprim for another 6 month period.

Follow-up contrast enhanced CT revealed resolution of the abscess (Fig 1B) along with supratentorial lesions associated with parenchymal scarring. She did well thereafter and continued her medicines for SLE.

DISCUSSION

A wide spectrum of immune disorders could contribute to the development of nocardiosis comprising of immune deficiency syndromes like acquired immune deficiency syndromes (AIDS) and autoimmune disorders, pulmonary alveolar proteinosis, renal transplant recipients, lymphoreticular neoplasms and even pregnancy. A growing array of these disorders have been mentioned elsewhere (4,5,7,9,10,14,15,17). Fortunately the condition is quite rare, however it could be easily missed.

Cerebral lupus is a medical catastrophe with poor prognosis. Neurological manifestations occur in 25% of patients with SLE. The symptoms are attributed to small vessel thrombosis and fibrinoid degeneration, petechial hemorrhage and microaneurysms. In some cases, cerebral emboli originate from the endocardium or thrombotic thrombocytopenia (19). Although very rarely infections have been speculated as the causative factor for neurological symptoms in SLE patients, however they deserve special attention because of their potential response to proper treatment.

In literature cited so far, the reported number of cases with SLE and nocardiosis are less than 30. It's incidence is 2.8% in all SLE patients, and lung has been the most common site of involvement (81%) followed by CNS (15%); however none of the latter cases involved the cerebellum (13). Our case had both SLE and long-term corticosteroid as antecedent factors, the former causing immune deficiency by consuming immune mediators and cells and the latter through generalized immunosuppression (19). Thus nocardial CNS infection can occur in patients with active SLE as well as in cases who are on remission but receiving long-term steroid therapy. It seems that the latter situation prevailed in our patient.

Contrast enhanced MRI is the best diagnostic procedure for cerebral lesions. Usually multiple discrete hyperintense lesions appear throughout neuraxis which enhance after contrast injection while vascular lesions of cerebral lupus typically do not enhance (19). Not surprisingly contrast enhanced computed tomography and neutrophil scanning have also been used by other authors (8), but their accuracy remains unsettled. Of the lesions observed, one may show enlargement, more pronounced rim and even multinodularity. Thus observation of a cerebral abscess associated with multiple deep lesions in an immune compromised host should raise the suspicion of nocardial infection in conjunction with other diagnostic possibilities as in our case and may require diagnostic sampling either by open or via stereotactic aspiration (3).

Treatment of these lesions is also controversial. Although many authors insist on diagnostic aspiration and proper antibiotics (10,11,18,21), still others report higher mortality and recurrence rate if the lesion is not excised through a formal craniotomy (6,11,12,13,20, 22). It appears that small multiple lesions without mass effect respond to medical treatment alone, while larger multiloculated abscesses require surgical excision. The cerebellar abscess in our patient was dealt with in the same way.

According to many authorities, prognosis of nocardiosis appears dismal when the central nervous system has been involved (20) and a mortality of about 90% has been associated with cerebral lesions. However
timely diagnosis and intervention followed by proper antimicrobial treatment led to long term survival in many cases as in our patient, unless the underlying disease had been life threatening during the convalescence period (2,14,15,13).

Thus in patients with SLE, the appearance of new neurological symptoms may be erroneously overlooked as cerebral lupus, whilst timely diagnosis imaging and sampling followed by employment of proper antimicrobials, if an abscess is found, could usher in good outcome for such dreadful fatal malady.

REFERENCES


