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

H. Saberi[1]*., SA. Fakhr Tabatabai*[1] *and Z. Hussainkhan*[2]

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

2) Department of Anesthesiology, 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 astroides. This opportunistic organism complicates many disorders characterized by cellular or humoral immunity disturbances.

A 36-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 imagings performed because of headaches and dysequilibrium of two weeks duration.

Craniectomy was performed and the abscess excised, Nocardia astroides grew in the pus culture . On switching the empirical therapy to a combination of trimethoprimsulfamethoxazole 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 umbrella of specific antimicrobial treatment presents the only plausible option.

Acta Medica Iranica 39 (2): 118-121; 2001

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 1888 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 Farcinia 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).

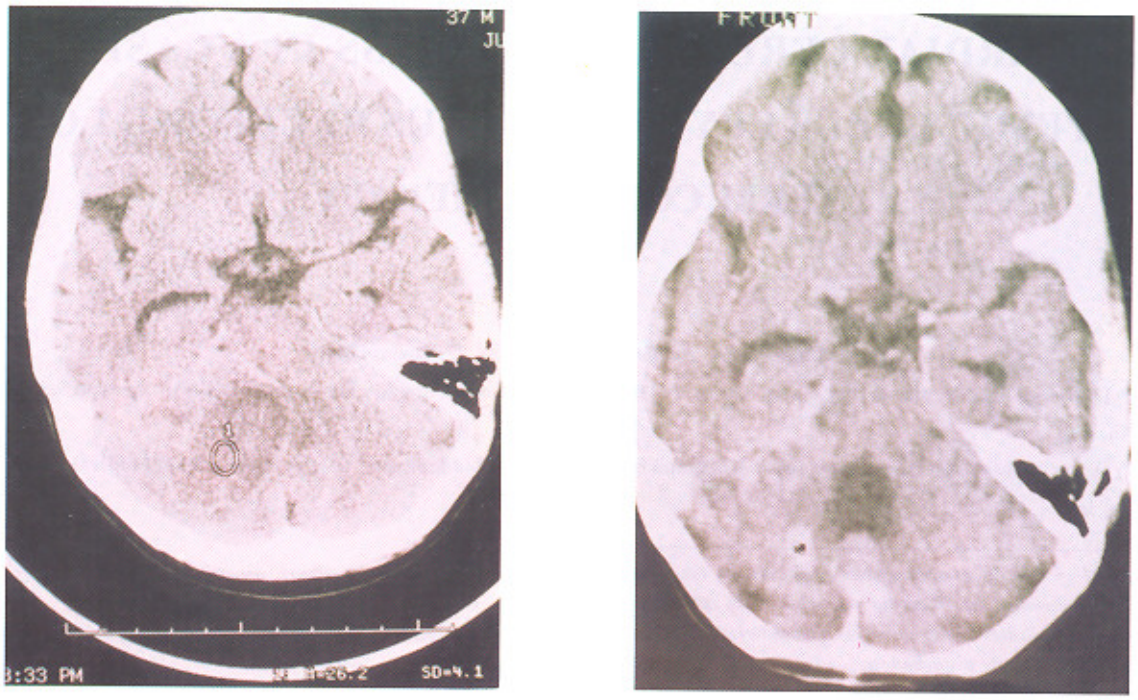
Among various predisposing factors for nocardiosis, both long term corticotherapy and systemic lupus erythematosus (SLE) have been implicated (3). Although coexistence of both factors in the same patient could have contributed to the development of the overwhelming infection, yet the exact influence 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 remediable. 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 usher 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 keypoints for a hopeful result could not be overemphasized.

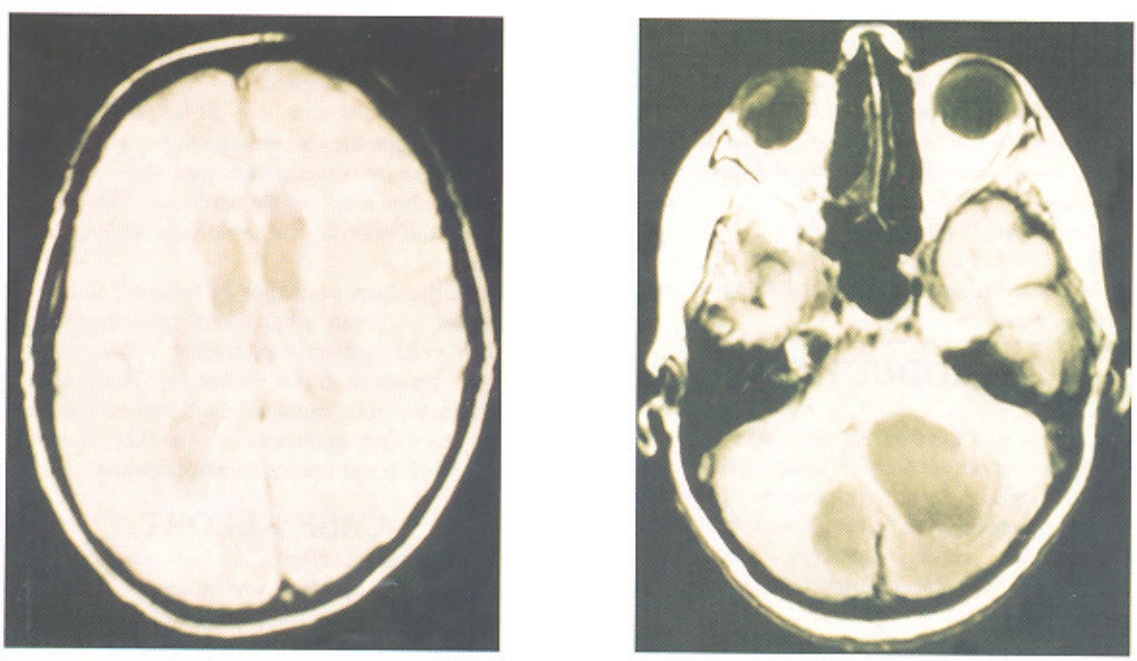
CASE REPORT

A 36-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



A B

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 scaring.



A B

Fig. 2. T2 weighted magnetic resonance imaging of the patient revealing multiple hyperintense supratentorial lesions (A) and multiloculated cerebellar abscesses compressing the forth ventricle (B).

time revealed proteinuria > 1 gr/24h, ESR = 70, 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 complements became normal, ANA titer decreased slightly and anti-ds-DNA became negative. The patient's clinical condition showed a spectacular improvement. A year later (present illnes) the patient reported with headaches, confusion, dysequilibrium 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 contrtast enhanced computed tomogram followed by magnetic resonance imaging of the brain was performed which revealed multilobed cerebellar hemispheric 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 astroides. Thus sulfamethoxazole-trimethoprim (800/160 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 microinfarcts. 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 sympoms 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 50. It's incidence is 2.8% in all SLE patients, and lung has been the most common site of involvement (81%) followed by CNS (13%); however none of the latter cases involved the cerebellum (13). Our case had both SLE and long-term corticotherapy 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 in 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 multilocularity. 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 streotactic aspiration (3).

Treatment of these lesions is also controversial. Although many authors insist on diagnostic aspiration and proper antibiotics (10,11,8,21), 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, while timely diagnostic 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

1. Lerner IP. Nocardia species. In: Mandell, Douglas, and Bennet's (eds). Principles and Practice of Infectious Diseases. Fourth edition. London: Churchill Livingstone Inc. 2273-2280; 1995.

2. Mathisen GE and Johnson JP. Brain abscess. Clin. Infec. Dis. 25(4): 763-779; 1997.

3. Pytinen J, Paakko E and Jartti P. Cerebral abscess with multiple rims of MRI. Neuroradiology. 39(2): 857-859; 1997.

4. Agular PH, Pahl FH, Ulp DE et al. Cerebellar abscess by Nocardial: a case report. Arq Neuropsiquiatr. 53(2): 307-311; 1995.

5. Attapattu M, Amarasekera N, Gunaskera WS and Hewage UC. Brain abscess caused by Nocardia astroides. Ceylon. Med. J. 41(2): 71-72; 1996.

6. Braum TI, Kerson LA and Eisenberg FP. Nocardial brain abscesses in a pregnant woman. Rev. Infec Dis. 13(4): 630-632; 1991.

7. Buxton N and Mcintosh J. Multiple nocardial brain abscesses: report of two patients. Br. J. Neurosurg. 8(4): 501-503; 1994.

8. Cheung RT, Yu YL and Chang CM. Primary cerebral abscess due to Nocardia astroides presenting as stroke. Clin. Exp. Neurol. 31: 79-84; 1999.

9. Gupta SK, Manjunath - Prasad KS, Sharma BS et al. Brain abscess in renal transplant recipients: report of three cases. Surg Neurol. 48(3): 284-287; 1997.

10. Hall WA, Artinez AJ,'Dummer JS and Lunsford LD. Nocardial brain abscess: diagnostic and therapeutic use of stereotactic aspiration. Surg. Neurol. 28(2): 114-118; 1987.

11. Krone A, Schaal KP, Brawanski A and Schukneeht B. Nocardial brain abscess cured by imipnem/amikacin and enucleation. Neurosurg. Rev. 12(4): 333-340; 1989.

12. Mamelak An, Obana WG, Flaherty JF and Resoenblum ML. Nocardial brain abscess: treatment sterategies and factors influencing outcome. Neusurg. 35(4): 622-631; 1994.

13. Mok CC, ue KY and Lau CS. Nocardiosis in systemic lupus erythematosus. Semin Arthritis Rheum. 26(4): 675-683; 1997.

14. Mollinger A, Jallo GI, Zagzag D and Kelly PJ. Nocardia abscess of the chroid plexus: clinical and pathological case report. Neurosurgery. 43(4): 949-952; 1998.

15. Oerlemans WG, Jahnsen EN, Prevo RI. and Eijsvogek MM. Primary cerebellar nocardiosis and alveolar proteinosis. Acta Neurol. Scan. 97(2): 138-141; 1998.

16. Ogg G, Lynn WA, Peters M, Curati W, McLaughlin JE and Shaunak S. Cerebral nocardia abscesses in a patient with AIDS: correlation of magnetic resonance and white cell scanning images with neuropathological findings. J. Infect. 35(3): 311-313; 1997.

17. Oktem IS, Akdemir H, Sumerkan B, Koc RK, Menku A and Tumturk F. Cerebellar abscess due to Nocardia astroides. Acta Neurochir (Wien) 14(2): 217-218; 1999.

18. Overkamp D, Waldmann B, Lins T, Lingenferser T, Petersen D and Eggstein M. Successful treatment of brain abscess caused by nocardia in an immunocompromised patient after failure of co-trimoxazole. Infection 20(6): 365-366; 1992.

19. Rowland PL. Vasculitis syndromes. In: Rowland PL (ed). Merritt's Neurology. 10th Edition. Philadelphia: Williams and Wilkins; 9029-11; 2000.

20- Sabeel A, Alrabiah F, Alfurayh O and Hassounah M. Nocardial brain abscess in a renal transplant recipient successfuly treated with tripple antimicrobials. Clin. Nephrol. 50(2): 128-130; 1998.

21. Turner E and Withby JL. Nocardial cerebral abscess with systemic involvement successfully treated by aspiration and sulphonamides. Case report. J. Neurosurg. 31(2): 227-229; 1969.

22. Tyson GW, Welsh JE, Butter AB, Jane JA and Winn HR. Primary cerebellar nocardiosis. Report of two cases. J. Neurosurg. 51(3): 408-414; 1979.