POST-VACCINAL ACUTE DISSEMINATED ENCEPHALOMYELITIS: A CASE REPORT

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D. Yalçın, M.D.*/ O. Çoban, M.D.**/ M.Barlas,M.D.****/ A. Gökyiğit, M.D.***

* *Professor, Department of Neurology, Faculty o Medicine, İstanbul University, İstanbul Türkiye.*
** *Associate Professor, Department of Neurology, İstanbul Faculty of Medicine, İstanbul, Türkiye.*
*** *Specialist, Department of Neurology, Şişli Etfal Hospital, İstanbul, Türkiye.*
**** *Specialist, Department of Neurology, Faculty of Medicine, İstanbul University, İstanbul, Türkiye.*

SUMMARY

Acute disseminated encephalomyelitis (ADEM) may occur following viral exanthemata or following vaccination. The disorder represents an allergic phenomenon which appears to result from delayed hypersensitivity to myelin basic proteins. On the other hand, circulating immune complexes are postulated to cause vascular injury. In this study we report computerized tomography (CT) findings in a case of ADEM due to Semple type vaccination.

Key words : Acute disseminated encephalomyelitis, Computerized tomography, Semple type vaccine.

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an uncommon disorder of the nervous system. ADEM may occur following viral exanthemata (parainfectious encephalomyelitis) or following vaccination (postvaccinal encephalomyelitis) (1). There is general agreement that the disorder represents an allergic phenomenon which appears to result from delayed hypersensitivity to myelin basic proteins (2). ADEM has an important place under the neurologic complications of Semple rabies vaccine, which is used in a few countries because of its low cost for production.

CASE REPORT

An 8-year-old boy had been bitten by a mouse one year and two months ago and received each time intra-abdominal injections of Semple vaccine. Over a few days he developed progressive ataxia and drowsiness. Neurologic examination disclosed a drowsy boy. There was a slight right facial weakness, positive Kernig and Brudzenski signs, a gross truncal and appendicular cerebellar deficit. The tendon reflexes were brisk in all four extremities and the plantar responses were extensor. Superficial abdominal reflexes were absent. Sensory examination and strength were normal. A computed tomographic (CT) scan on the 13th day of the illness revealed contrast enhancement in front of the frontal horn (Fig. 1). Laboratory studies showed only mildly elevated sedimentation rate. Over the following days the patient developed severe truncal and appendicular ataxia, horizontal and vertical nystagmus, bilateral ophthalmoplegia, emotional incontinence, modest weakness on the right arm and left facial weakness. Electroencephalogram showed bilateral abnormalities consistent with a left dominance The cerebrospinal fluid (CSF) contained 15 mononuclear cells per cubic milimeter, a protein level of 9 mg/dl and glucose level of 65 mg/dl. The immunoglobulins in CSF were mildly elevated (Ig G: 15.3 mg/dl, Ig A: 5.1 mg/dl, Ig M: 2.8 mg/dl). Methylprednisolone (60 mg daily) was begun. Marked improvement occurred over the first week and full recovery was evident within 8 weeks. A CT scan on the 50 th day of the illness demonstrated the disappearance of contrast enhancement and nonenhancing lesions of decreased density was present in subcortical white matter (Fig. 2). The repeated CSF examination showed no cell. Parallel to the clinical improvement, the methylprednisolone treatment was tappered and ceased. The third CT scan was normal (Fig. 3).

DISCUSSION

ADEM involves neurological complications occurring after the immunization with Semple type of rabies vaccine which is in use in many developing countries including Turkey. Pathalogically, a small vessel vasculopathy involving arterioles and capillaries as well as venules in both gray and white matter is the earliest and most consistent change. Perivascular demyelinization appears to develop subsequently. The disorder represents an allergic phenomenon which appears to result from delayed hypersensitivity to myelin basic proteins. On the other hand, circulating immune complexes are postulated to cause vascular injury (1,2).

The incubation period from vaccination to onset of neurologic symptoms is usually 6-14 days but there have been reports where the incubation period was longer (3-6) The incidence of post-rabies vaccination encephalitis is said to increase with the number of injections given in one course of treatment (4).

Our patient, who was an eight year old boy, had been twice vaccinized with Semple type vaccine because of mouse bites. He developed neurological symptoms which started with drowsiness and ataxia, and progressed up to three weeks during which symptoms suggesting involvement of the central nervous system were added to the clinical picture. The first CT scan which was performed on the 13th day showed bilateral contrast enhancement in front of the frontal horn. These lesions disappeared on the 50th day when diffuse asymmetric lesions of decreased density was present in subcortical white matter. The third CT scan, which was done when neurological symptoms disappeared with the exception of minimal truncal ataxia was normal.

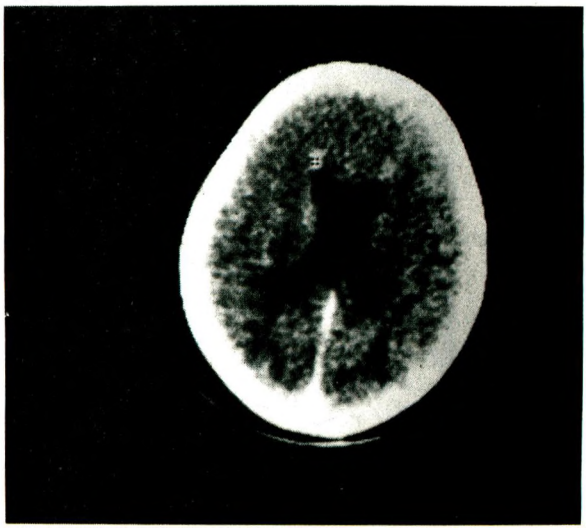

Fig. 1 CT scan on the thirteenth day of the illness revealed bilateral contrast enhancement in front of the frontal horn.

A delay between the onset of clinical signs and the appearance of lesions on CT scan is common. Usually the patients have normal scans initially but develop CT abnormalities after 5 to 14 days. It is possible that they develop further abnormalities over the following one or two weeks (1,7). There is a limited correlation between the clinical signs and the anatomical distribution of lesions seen on CT scan. The combination of extensive nonenhancing lesions of decreased density in subcortical white matter and patchy or confluent contrast-enhancing cortical lesions are unique to ADEM. CT enhancement correlates with the breakdown of blood brain barrier or vasculitis and the resolution of the contrast enhancement after steroid therapy may be related to the resolution of vasculitis component of ADEM. The persistent low density lesions may represent demyelinization and/or necrosis or scar formation which may be responsible for the sequelae of ADEM (1,7).

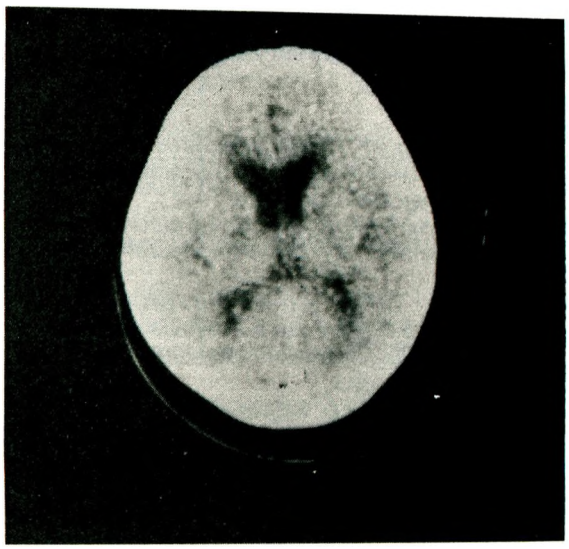

Fig. 2 CT scan on the fiftieth day of the illness demonstrated the disappearance of the contrast enhancement and non-enhancing lesions of decreased densitiy was present in subcortical white matter.

There is lack of laboratory abnormalities which may be considered specific for ADEM. CSF profiles show a normal glucose level, normal or mildly increased protein level and normal or mild lymphocytic pleocytosis. CSF Ig G might be elevated. In our case there was a mild lymphocytic pleocytosis and Ig G elevation.

In conclusion, our observations (Semple type vaccine repeated twice in two years, course of the illness, increased Ig G in the CSF, CT scan findings which are specific for ADEM) suggest that the neurological symptoms of our patient are due to Semple type vaccine.

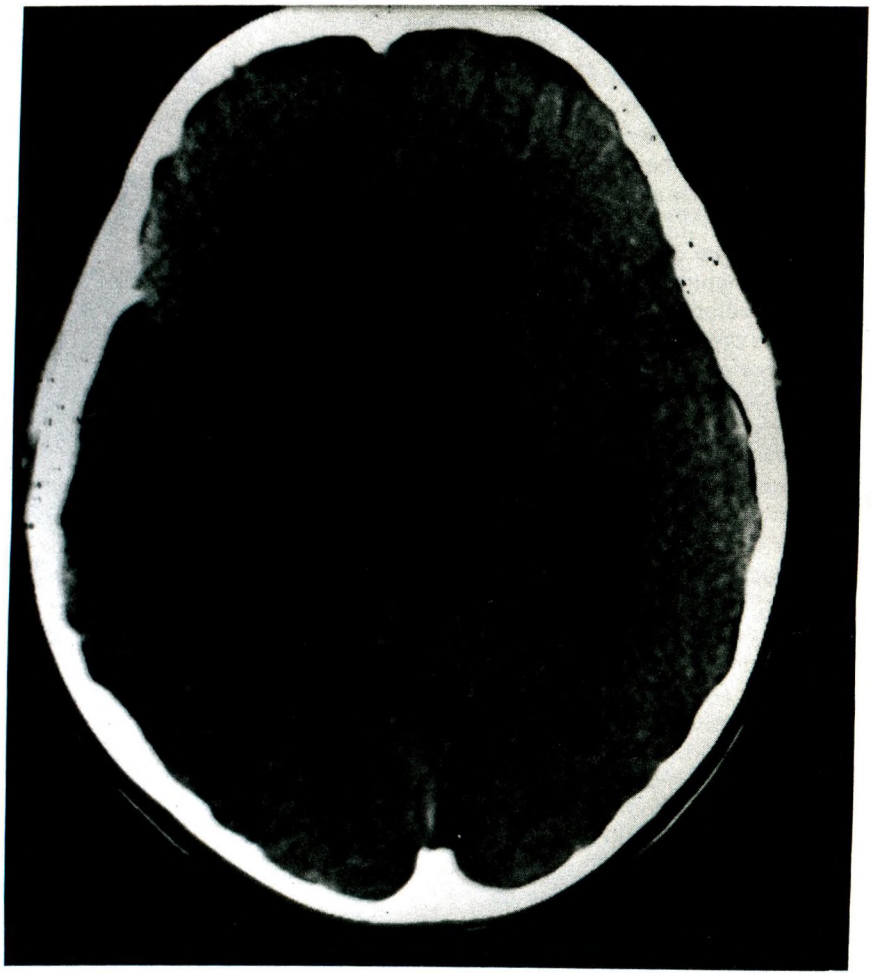

Fig. 3 The third CT scan was normal.

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