Lady Ridgeway Hospital for Children, Colombo.

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'myoclonic encephalopathy of Kinsbourne'

Introduction

‘Opsoclonus myoclonus syndrome’ (OMS), also known as

‘myoclonic encephalopathy of Kinsbourne’1 and ‘dancing

eyes-dancing feet syndrome’, is a very rare disorder

commonly associated with paraneoplastic features of

neuroblastoma. It has a devastating and debilitating onset

but is generally non fatal. Symptoms are steroid responsive

and recovery from acute symptoms is good. Children

suffer lifelong neurological sequelae.

Case report

A previously healthy 16 month old baby girl with normal
developmental milestones, presented to a local hospital

with a febrile illness, followed by unsteady gait for 3 days.

It was managed as viral cerebellitis with IV aciclovir and
dexamethasone. CT scan of brain done at that time was

normal. She had shown a gradual recovery and was sent

home within 10 days.

She had remained well for nearly one month when she was

readmitted with unsteady gait, dancing eye movements and
tubation. After admission, she had developed three

generalised tonic clonic seizures and was transferred to the

Lady Ridgeway Hospital. CT and MRI scans of the brain

were normal. At this juncture parents decided to seek

ayurvedic treatment and left against medical advice.

After 5 weeks, she was readmitted with fever and severe

vomiting. She was very irritable and restless, had difficulty

falling asleep, was extremely ataxic and could not be held

in one position even with support. The eye movements

were chaotic and she had myoclonic jerks. Mother was

very concerned that the child had drooling and had become

mute. There were no other focal neurological signs. There

was no hepatosplenomegaly, lymphadenopathy or other

abdominal masses. The possibility of neuroblastoma was

excluded by the normal blood picture, bone marrow,

urinary catecholamines and ultrasound scan of abdomen. A

provisional diagnosis of OMS was made and an occult

neuroblastoma was ruled out after performing CT scan of

chest and abdomen. Screening for Japanese encephalitis,

urinary catecholamines and ultrasound scan of abdomen. A

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immunosuppression. It is hypothesized that a viral

infection (EBV, Coxsackie B, enterovirus) or a tumour

initiates the cascade of immune reactions.

Discussion:

The average age of onset of OMS is 19 months with a

range of 6-36 months. The pathophysiology of OMS has

been described by autoantibodies2 directed against neural

antigens3 in cerebellar Purkinje cells, cerebral cortical

neurons and axons. The immune mechanism may be a type

11 or IV hypersensitivity. It is hypothesized that a viral

infection (EBV, Coxsackie B, enterovirus) or a tumour

initiates the cascade of immune reactions.

Tumours are found in about half of the cases of opsoclonus

myoclonus. The most common tumours in children with

OMS are neuroblastoma and ganglioneuroblastoma, which

often occur in the chest but also in abdomen and pelvis.
The body has greater success in eradicating a

neuroblastoma than any other tumour indicating a strong

immune response. Overall incidence of OMS in children

with neuroblastoma is approximately 3%. When

neuroblastoma is associated with OMS, the prognosis is

favourable with 90% showing long term survival. But 70%

of these children suffer lifelong neurological sequelae with

impaired motor, cognitive, language and behavioural

problems. In 50% of OMS a tumour is not detectable but

the possibility remains of a low grade tumour which has

regressed spontaneously before detection.

Currently there is no well accepted treatment in OMS.

Generally treatment consists of immunosuppression4 to reduce

the formation of antibodies and promote its removal. ACTH

and corticosteroids lead to rapid improvement of neurological

symptoms but most patients have frequent relapses following

tapering or withdrawal of drugs. ACTH acts as a direct

immunosuppressive and inhibits the antibody response to T--
cell dependant antigens. Corticosteroids decrease the

lymphocyte differentiation and proliferation, inhibit

phagocytosis and suppress production of interleukins. To

overcome the problem of relapses ACTH is given in high

doses over a longer period, gradually tailing of over 1 year.

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Treatment with intravenous immunoglobulin, plasmapheresis, immunoadsorption and oral immunosuppressive agents have been used to treat resistant cases. Medications to avoid are midozolam and melatonin.

References


