Acute ataxia complicating Langherans cell histiocytosis

A Polizzi, S Coghill, M A McShane, W Squier

A case is reported of a 3 year old boy with an acute history of cerebellar impairment and x-ray evidence of apparent chest infection. At postmortem examination, his lungs but not the nervous system were found to be massively infiltrated by Langherans histiocytes. In retrospect, the acute ataxia was diagnosed as a paraneoplastic phenomenon secondary to Langherans cell histiocytosis (LCH). This represents a unique occurrence complicating LCH in childhood.

A 3 year old boy was admitted to hospital with a six week history of unsteadiness and slurred speech. There was no history of recent illness. A computed tomography brain scan, performed a few days before admission, was normal.

Perinatal history was uneventful. Motor development was normal. Family history was unremarkable.

On admission he was alert and playful. On physical examination his weight, height, and head circumference were within the 50th–75th centile range. There was no fever, anaemia, jaundice, rash, or petechiae; lymph nodes were not palpable. On auscultation the chest was clear with no added sounds. The liver was within normal limits and the spleen was palpable 4 cm below the costal margin. Neurological examination was unremarkable except for a broad based gait and mild intention tremor. Results of laboratory tests including blood cell count, biochemical tests, and urinary vanillylmandelic acid were normal. Cerebrospinal fluid was normal with no oligoclonal bands and no abnormal cells. Cerebrospinal fluid culture was sterile. Antibodies to neural antigens were negative in the serum. Magnetic resonance imaging of the brain was normal. However, on the plain film of the chest, widespread reticulonodular shadowing was present (fig 1), suggestive of either mycoplasma pneumonia or metastatic disease. Abdominal ultrasound scan revealed normal liver, spleen, kidneys, and retroperitoneum. Given the lack of respiratory signs, mycoplasma infection was suspected. Blood was taken to look for specific antibodies; the patient was discharged home for the weekend on erythromycin.

After two days he was urgently admitted to hospital for increasing malaise, vomiting, drowsiness, and dyspnoea. He died soon after arrival.

Postmortem examination revealed bilateral tension pneumothorax. Both lungs had large bronchiectatic cavities. In the

**Figure 1** Chest x-ray picture showing widespread reticulonodular shadowing.

**Figure 2** (A) Lung biopsy specimen from the child, showing a bronchiocentric nodular lesion demarcated from the surrounding lung tissue. (B) Higher magnification of the same specimen, showing Langherans cells.

**Abbreviations:** CNS, central nervous system; LCH, Langherans cell histiocytosis
central nervous system (CNS) there was only generalised con-
genestion of the meningeal vessels. Histologically the lungs were
massively infiltrated by Langherans cells (fig 2). There was
also splenic infiltration by Langherans cells and lymphocytes.
The brain showed abnormalities only in the cerebellum with
focal gliosis associated with Purkinje cell loss. There were no
changes elsewhere and there was no evidence of inflammation
or histiocyte infiltration.

DISCUSSION
Histiocytoses are rare disorders characterised by proliferation
of cells of the monocyte–macrophage system. In LCH a defini-
tive diagnosis is made by demonstration of characteristic
Birbeck granules in the abnormal cells and CD1a antigen on
the cell surface.1

LCH is extremely variable in presentation. Most frequently
there are bone lesions and seborrhoeic dermatitis; hepatic and
spleen enlargement, pancytopenia, fever, weight loss, irritabil-
ity, and generalised lymphoadenopathy are common in the
most severe forms. Isolated pulmonary infiltrates are unusual
in children.2

CNS manifestations of LCH include diabetes insipidus from
direct hypothalamic or pituitary invasion, site dependent
neurological symptoms from infiltrative lesions, and rarely
neurological dysfunction of cerebellar origin.2

In the latter group the majority of cases present in adults
with multisystem LCH have an insidious onset and a rather
slow course, with no evident infiltration within the CNS.
Degenerative changes with demyelination, Purkinje cell loss,
and reactive gliosis without typical LCH cells can be found in
the cerebellar tissue.2

Cerebellar impairment has only occasionally been reported
in children with LCH, manifesting many years after the diag-
nosis of the disease. Typically the cerebellar dysfunction has a
chronic and progressive course with no infiltration of the
CNS,3,4 supporting the hypothesis of a remote effect (paraneo-
plastic) of pathological Langherans cells by an immune med-
iated mechanism.1

To our knowledge this is the first reported case of acute cer-
ebellar dysfunction presenting as isolated manifestation of a
particularly aggressive form of LCH, supported only by x ray
evidence of an asymptomatic lung infiltration.

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