SHORT REPORT

Acute ataxia complicating Langerhans cell histiocytosis

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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 Langerhans histiocytes. In retrospect, the acute ataxia was diagnosed as a paraneoplastic phenomenon secondary to Langerhans cell histiocytosis (LCH). This represents a unique occurrence complicating LCH in childhood.

Figure 1 Chest x ray picture showing widespread reticulonodular shadowing.

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 vanillyl-mandelic 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

Abbreviations: CNS, central nervous system; LCH, Langerhans cell histiocytosis

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

central nervous system (CNS) there was only generalised congestion 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 definitive 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 seborrheic dermatitis; hepatic and spleen enlargement, pancytopenia, fever, weight loss, irritability, and generalised lymphadenopathy 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 diagnosis 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 (paraneoplastic) of pathological Langherans cells by an immune mediated mechanism.⁵

To our knowledge this is the first reported case of acute cerebellar 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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