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CLINICAL IMAGE

A rare cause of chronic ataxia in childhood: ganglioneuroma

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Ganglioneuroma is an extremely rare cause of ataxia in childhood, the only descriptions of which are in the form of case reports [1, 2]. Ganglioneuroma can be asymptomatic in the abdominal and thoracic region, or else can be symptomatic with abdominal pain, scoliosis, opsomyoclonus and ataxia. It may cause dysphonia and stridor in a cervical location [1]. While chronic ataxia with associated findings such as opsoclonus and cerebellar atrophy are rare in ganglioneuromas [3–5], isolated pure chronic ataxia is exceedingly unusual [2].

A 4-year-old-boy presented with wide-base gait over the previous 2 years. He recently described hand tremor when reaching out for an object. There was no infection history prior to the symptoms. His personal medical history was uneventful, and there was no family history of ataxia or other movement disorder. Neurological examination revealed gait ataxia and dysmetria. Physical examination revealed no opsoclonus-myoclonus, nystagmus or oculomotor apraxia. Alpha fetoprotein level was borderline at 0.98 mg/dL (range 0–0.9 mg/dL). Metabolic screening, electromyography, cranial neuroimaging findings and aprataxin and senataxin gene analysis were normal. Spinal magnetic resonance imaging (MRI) revealed a mass lesion on the right adrenal gland. Abdominal MRI revealed a mass lesion 21 × 18 mm in size in the right adrenal gland with homogeneous contrast enhancement (Fig. 1). Surgery was performed following consultation with the pediatric surgery department. The right adrenal mass was excised totally. Pathological examination identified the tumor as ganglioneuroma (Fig. 2).

Peripheral neuroblastic tumors are tumors of the neural crest. They are classified into three histological types: neuroblastoma, ganglioneuroblastoma and ganglioneuroma. They derive from the sympathetic ganglia or adrenal medulla and exhibit a broad spectrum of differentiation and neuronal maturation [6]. Neuroblastoma is the most common and most highly malignant form. This exhibits a histological pattern of abundant neuroblasts and scarce stroma. Opsoclonus-myoclonus ataxia syndrome is the most common paraneoplastic manifestation, being reported in 2–4% of children with neuroblastoma [7]. Ganglioneuromas are rarer and generally benign. These are stroma-predominant tumors, with more or less mature ganglion cells. Pure ganglioneuroma does not contain immature elements, such as neuroblasts, intermediate cells, or mitotic figures [1, 8, 9]. Ganglioneuromas most commonly occur in the mediastinum, retroperitoneum, cervical region and abdominal region. Although they can cause symptoms such as abdominal pain, scoliosis, and stridor, they can also be asymptomatic. Although clinical data concerning neurological symptoms during childhood are limited, these tumors may cause opsoclonus-myoclonus ataxia as they originate from the neural crest [1]. Since there is no treatment for the majority of progressive hereditary ataxias, it is important for treatable causes not to be overlooked [10, 11].

Following complete resection of the tumor, the gait ataxia in this case resolved in a few days. Ataxia and dysmetria had resolved completely at neurological examination 1 month

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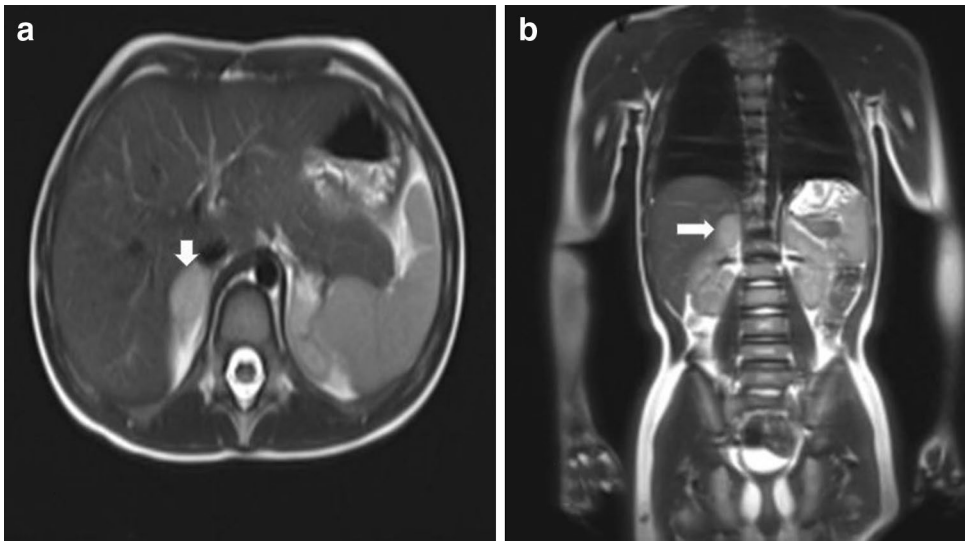

Fig. 1 A mass lesion in the right adrenal gland in the coronal (**a**) and axial (**b**) planes (white arrows)

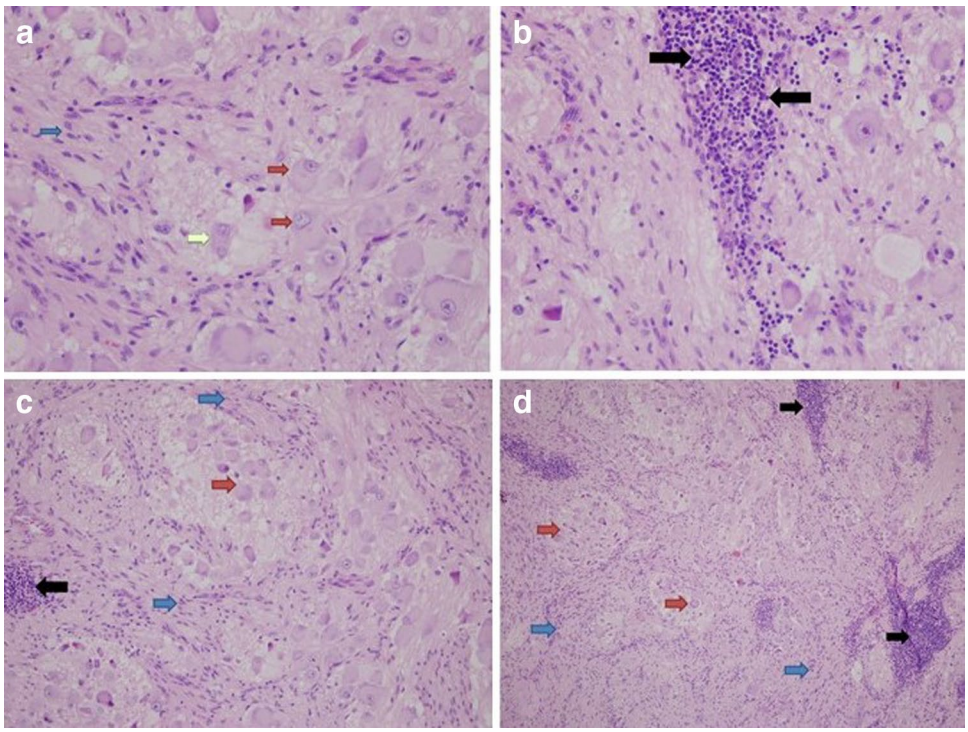

Fig. 2 Pathological examination of the tumor revealed findings of ganglioneuroma. **a** Ganglion cells (red arrows), binuclear ganglion cells (yellow arrows), Schwann cells (blue arrow) (hematoxylin and eosin staining, original magnification × 400); **b** Lymphoid aggregates that can be seen in ganglioneuromas and that may be confused with primitive neuroblastic cells (black arrows) (hematoxylin and eosin staining, original magnification × 400); **c** Ganglion cell groups (red arrow), Schwann cells (blue arrows), and lymphoid aggregates (black arrow) (hematoxylin and eosin staining, original magnification × 200); **d** Ganglion cell groups (red arrows), Schwann cells (blue arrows), and lymphoid aggregates (black arrows) (hematoxylin and eosin staining, original magnification × 100)

subsequently, and neurological examination was normal at sixth month follow-up.

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Compliance with ethical standards

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflict of interest None.

Informed consent Informed consent was obtained from the patient included in the case report.

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