Images in...

A young man with slowly progressive ataxia

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DESCRIPTION

A 22-year-old man presented with 5 years history of progressive neurological symptoms manifesting as ataxia, dysarthria and vertigo. The patient was apparently normal 5 years back when he had an episode of fever for 4–5 days that got relieved spontaneously and after few days the patient noticed swaying from side to side during work and his gait become ataxic. It was progressive with time and at present the patient is unable to walk without support. After 1 month of ataxia, the patient started having a slurred speech. For the last 3 months the patient was having mild vertigo without any associated vomiting. There

was no history of altered sensorium, motor weakness or sensory loss. There was no history of seizures, fever and headache or head injury. On neurological examination, the patient was alert, friendly, cooperative and oriented to time, place and person. His speech appeared to scan each syllable. The patient had horizontal nystagmus on both vertical and lateral gaze. There was bilateral finger-to-nose ataxia and impaired heel knee test. The motor strength and reflexes were normal. He was investigated with a CT head (figure 1) and MRI (figures 2 and 3).

The CT scan of the head in this patient revealed a large cyst in the posterior fossa (figure 1). On MRI (figures 2 and 3), T1 weighted sagittal image of the brain showed a large cerebrospinal-fluid filled cyst in the posterior fossa and a large cystic lesion occupying nearly the entire posterior fossa that was incorporating and appeared inseparable from the fourth ventricle (figure 2). Dysplasia of the cerebellar cortices was also noted (figure 3). These changes were consistent with the Dandy–Walker malformation.

The Dandy–Walker complex is a rare congenital malformation and is morphologically characterised by agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle and enlargement of the posterior fossa. It is a genetically sporadic disorder that occurs one in every 25 000 live births, mostly in females.¹ Clinical manifestation in adults is totally different from neonatal presentation. In adults, the most common symptoms are headaches, papilloedema and vomiting resulting from increased intracranial pressure. Some adults may be asymptomatic with normal intracranial pressure or they may have only mild

Figure 1 CT head of the patient.

Figure 2 Cranial MRI of the patient.

Figure 3 Cranial MRI of the patient.

unsteadiness of gait or intellectual impairment until the syndrome is activated by head trauma or a systemic infection.^{2 3} Our patient developed the symptoms at the age of 17 years after a febrile illness. Most patients with the adult Dandy–Walker syndrome are referred for lateral ventriculo-peritoneal or cysto-peritoneal shunting procedures.⁴

Learning points

- ▶ Adult slow progressive ataxia can be due to congenital malformation of cerebellum.
- ▶ Dandy Walker syndrome can manifest late in adults sometimes precipitated by infection or trauma.

Competing interests None.

Patient consent Obtained.

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