A 30-year-old female presented with headache and gait disturbance which were gradually progressive for the past three months. On T1 weighted magnetic resonance imaging (MRI), it appeared to be a 35mm × 25mm × 20mm hyperintense mass in the left cerebellar hemisphere encroaching on the vermis and causing compression of the fourth ventricle (Figure 1). A tiny biopsy, reported as medulloblastoma elsewhere, was followed by incomplete removal of the tumor and the patient failed to turn up for a follow-up. Six months later, the symptoms reappeared and a rebiopsy was taken.

The rebiopsy specimen showed a biphasic appearance with areas of lipidized vacuolated cells and neurocytic cells with small round to oval nuclei and scant clear cytoplasm (Figures 3). Immunohistochemistry revealed positivity for neuron-specific enolase and synaptophysin in neurocytic cells and adipocyte-like cells and a low MIB-1 index of 3%.

What is your diagnosis?

See the next page
The patient was provisionally diagnosed as cerebellar liponeurocytoma, a mimic of medulloblastoma on histopathology; it is a relatively new and rare entity with a far better prognosis. To date only 31 cases have been reported.1

It has a relatively benign clinical course and recurrence may appear after a long period of time if adequately excised.2–5 The prognosis is favorable if MIB-1 index is in the range of 1% – 3% and an aggressive adjuvant therapy is not mandatory. There have been no reports of spinal drop metastasis and it is justified to avoid spinal radiation.

In conclusion, compared to medulloblastoma, with which it was earlier clubbed, this entity needs to be differentiated, as it has a longer survival, a better prognosis, a lower proliferation rate, and adjuvant radiotherapy is not necessary.

References