An 11-year-old boy presented with a history of headaches and vomiting that had been present for several months.

Physical examination showed papilledema without the presence of any other neurological deficits.

Magnetic resonance imaging (MRI) revealed a large preportine mass with dorsal displacement of the brainstem and a secondary obstructive hydrocephalus due to compression of the aqueduct.

The lesion had an inhomogeneous hypointense aspect on the T1-weighted image (T1WI) and an inhomogeneous hyperintense aspect on the T2-weighted image (T2WI).

After administration of IV gadolinium, there was some inhomogeneous enhancement (Fig.1).

Computed tomography (CT) imaging showed no bone involvement.

During the first operation, a ventriculoperitoneal shunt was inserted into the right lateral ventricle to treat the hydrocephalus.

A careful study of the MRI suggested that this infra- and supratentorially located tumour might be resected through a single approach.

A left frontotemporal transsylvian approach was performed to gain access to the tumour.

The tumour had well-defined margins and was entirely located in the intradural plane.

There were no attachments to the cranial nerves or brainstem.

A macroscopic complete resection was performed.

Postoperatively, the patient had developed a left oculomotor nerve palsy, which completely recovered within the next 4 weeks.

The postoperative MRI showed a complete removal of the tumour (Fig.2).

After careful consideration by a multidisciplinary team, we decided that there was no indication for postoperative radiation therapy.

At follow-up one and a half years later, the patient was found to have remained asymptomatic.

There were no signs of tumour recurrence on the MRI scan.

At a follow-up of more than 6 years after treatment, there were still no signs of tumour recurrence on the MRI scan.

Histological examination showed a slightly lobulated tumour consisting of a chondromyxoid matrix.

The tumour cells showed a vacuolated and pale cytoplasm.

Moderate nuclear polymorphism was observed but no obvious mitotic activity (Fig.3).

Some calcifications were seen.

The tumour cells stained positive for pan-keratin, S-100 and epithelial membrane antigen (EMA).

These findings suggest a histopathological diagnosis of chordoma.