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Letter to the Editor

Diagnosing early upward cerebellar herniation by computed tomography: A diagnostic boom, a savior

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Sir,

Preoperative diversion of cerebrospinal fluid (CSF) is a common procedure for posterior fossa lesions. Although rare in incidence, upward cerebellar herniation may occur after such procedures, and is associated with a high mortality. Most of these patients present with sudden apnea and neurological deterioration with imaging showing dense intratumoral hemorrhage.[5] We bring to light a similar such case where early detection by computed tomography (CT) prompted urgent definitive surgery, resulting in a favorable outcome.

A 10-year-old male presented with the history of headache and gait imbalance for 3 weeks. Examination revealed gait ataxia, dysmetria, and papilloedema. CT scan suggested a midline cerebellar lesion. Magnetic resonance imaging (MRI) revealed a vermian lesion that appeared hypointense on T1-weighted (T1W) image, hyperintense on T2W image with focal hypointensity, and slight enhancement on Gadolinium. Associated mass effect on the brainstem with hydrocephalus was remarkable and so was the focal calcification [Figure 1]. Patient underwent right ventriculoperitoneal (VP) shunt, and the symptoms of raised intracranial pressure resolved thereafter. CT performed the next day depicted decompressed ventricles but with new onset intratumoral hemorrhage at the right lateral aspect of the tumor as well as blood within the third ventricle. The flattening of the quadrigeminal cistern was also evident. Clinically, the patient remained alert with pupils bilaterally symmetrical and reacting. With a working diagnosis of early upward cerebellar herniation, the child was taken up for urgent midline suboccipital craniotomy. Intraoperatively, a firm, vascular tumor was seen with clots at anterior aspect, suggestive of intratumoral hemorrhage, that extended through the aqueduct into the third ventricle. Gross total excision was achieved and the patient had an uneventful recovery.

Spontaneous hemorrhage in intracranial tumors is a known phenomena, the mechanisms of which are varied. It accounts for up to 6–10% of all intracranial hemorrhage.[4] Large sized tumors in the posterior fossa have a tendency to prolapse into supratentorial cavity following the CSF diversion. The sudden decompression of ventricles precipitates hemorrhage within the tumor that extends through the capsule into fourth ventricle and then to supratentorial ventricles.[2] The clinical presentation includes sudden neurological deterioration, abnormal fixed pupils, and respiratory irregularity. On radiology, one may find intratumoral hemorrhage and flattening of the quadrigeminal cistern.[1,3] The etiology of such hemorrhage is varied and includes hypertensive hemorrhage, arteriovenous malformation, medulloblastoma, acoustic schwannoma, and choroid plexus tumor.[3] CSF diversion is a commonly performed procedure in posterior fossa lesions prior to definitive surgery. It helps by improving symptoms of raised intracranial pressure and provides a better operative field.

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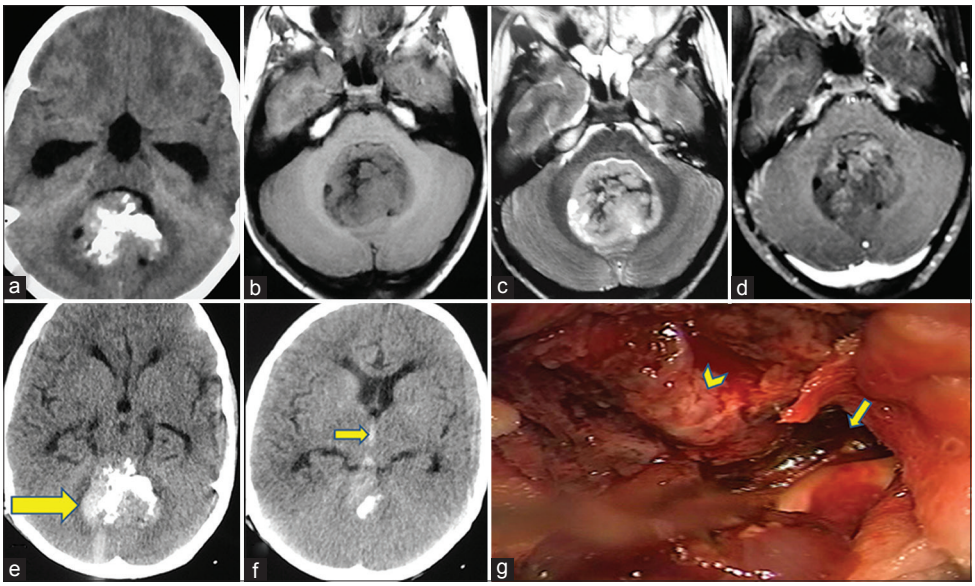

Figure 1: (a) Computed tomography (CT) axial image showing midline cerebellar lesion with hydrocephalus. (b-d) T1-weighted plain, T2-weighted (T2W), and T1-weighted (T1W) contrast axial magnetic resonance images showing vermian lesion hypointense on T1W (b), hyperintense on T2W (c), and faint enhancement (d). (e-f) Post shunt surgery noncontrast CT axial images showing intratumoral hemorrhage (arrow) (e) and intraventricular hemorrhage (arrow) (f). (g) Intraoperative photograph confirming tumor at the roof of fourth ventricle (arrowhead) and clot at anterior aspect near brainstem and aqueduct (arrow)

Although the incidence of upward cerebellar herniation is reported to be 3%, sudden clinical deterioration after shunt surgery is observed more often.[3] The high associated mortality rate reflects the importance of early diagnosis and prompt management. Hemorrhagic infarction of the brainstem may occur in later stage and can lead to dismal outcome.[3] Other than CSF diversion, factors aggravating such herniation are location of lesion near vermis and large opening in the incisura.[1] Lesions located adjacent to vermis, fourth ventricle, pons, and cerebellopontine angle have been associated with upward cerebellar herniation. Interruption of shunt and mechanical ventilation may halt the herniation process,[3] however, we proceeded directly for definitive surgery as the child was clinically stable.

The CSF diversion in such cases can be done by VP shunt, external ventricular drain (EVD), and endoscopic third ventriculostomy (ETV). Although, theoretically, VP shunt is better than EVD by providing guarded decompression of ventricles, EVD has its own merits as the CSF output can be carefully monitored and controlled in the inpatient setting. In addition, the CSF diversion with an EVD can be turned off more rapidly than with a VP shunt in the event of symptomatic upward herniation. Furthermore, an EVD can potentially be removed after definitive treatment avoiding a permanent indwelling system. Similarly, ETV is helpful by avoiding the need of a permanent shunt, with the disadvantage being the loss of control of CSF diversion. We believe that any evidence of new onset bleeding within the tumor extending rostrally to supratentorial ventricles after CSF diversion must be considered a sign of upward herniation, despite no neurological worsening. An easily available investigation in the form of CT scan can occasionally provide early diagnosis. We emphasize on early suspicion in case of any new onset intratumoral or intraventricular hemorrhage because prompt action in the form of urgent definitive surgery can help in averting a tragic outcome.

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Conflicts of interest
There are no conflicts of interest.

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