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Acute Communicating Hydrocephalus Caused by a Lumbar Intraneural Cavernous Hemangioma in a Child: A Case Report

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Case Report

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Abstract

Hydrocephalus secondary to a spinal lesion is rare and exceptional in children. Intraneural hemangiomas of the spinal nerve roots typically present with radicular symptoms; their association with hydrocephalus has been reported almost exclusively in adults. We report a previously healthy 5-year-old boy who presented with acute communicating hydrocephalus requiring emergency external ventricular drainage. Cerebrospinal fluid protein was markedly elevated without a cellular reaction, and no intracranial cause was identified. Spinal magnetic resonance imaging revealed an intradural mass at L3–L4 within the filum terminale fibers; resection yielded complete resolution of the hydrocephalus, and histopathology confirmed an intraneural cavernous hemangioma. At 6-year follow-up the child remained well, with no recurrence. To our knowledge, this is the youngest reported case of an intraneural hemangioma of the spinal nerve roots, and the first in any age group in which this lesion presented with acute communicating hydrocephalus. Spinal pathology should be considered in unexplained pediatric communicating hydrocephalus with elevated CSF protein; definitive resection can resolve the hydrocephalus and avoid permanent shunting.

Introduction

Hydrocephalus in children most commonly arises from intracranial causes, including aqueductal stenosis, intraventricular hemorrhage, neoplasia, and infection. Hydrocephalus attributable to a spinal lesion is rare and frequently overlooked, because the clinical and radiological presentation is dominated by its intracranial consequences while the causative lesion lies in the spinal axis [4, 11]. The most widely accepted mechanism is elevation of cerebrospinal fluid (CSF) protein, which increases CSF viscosity and impairs resorption at the arachnoid granulations, producing a communicating pattern of hydrocephalus [4, 11, 14].

Intraneural hemangiomas are benign vascular lesions that may arise within the nerve roots of the cauda equina, where they usually present with radiculopathy or progressive neurological deficit [6, 8, 9, 12]. Their association with hydrocephalus or raised intracranial pressure is exceptional and has been reported almost exclusively in adults [2, 13]. We report a child in whom acute communicating hydrocephalus was the presenting feature of an intraneural cavernous hemangioma of a lumbar nerve root.

Case Report

A previously healthy 5-year-old boy presented with a short history of headache, blurred vision, and progressive drowsiness, without focal deficit, meningeal signs, trauma, fever, or preceding illness. Cranial CT showed ventricular dilatation consistent with acute communicating hydrocephalus. An emergency external ventricular drain (EVD) was placed, with prompt improvement; follow-up CT confirmed satisfactory catheter position and reduced ventricular size (Fig. 1).

CSF protein was markedly elevated at 286 mg/dL (reference, 15–45 mg/dL), with no red blood cells or cellular reaction; hematological and biochemical work-up was otherwise unremarkable, with no evidence of infection or hemorrhage. Brain MRI confirmed the ventricular dilatation seen on CT (Fig. 2e, g, i), and complete spinal MRI revealed a well-circumscribed intradural mass, 21 × 15 × 15 mm, within the filum terminale fibers at L3–L4, hypointense on T1, hyperintense on T2, and homogeneously enhancing (Fig. 2a, b).

The patient underwent single-level L3 laminectomy. The lesion was dissected free from the surrounding cauda equina roots under continuous intraoperative neuromonitoring; the parent root was stimulated and, with no motor response elicited, was coagulated before complete removal. The specimen was a lobulated, purple, capsule-like fragment, 2 × 2 × 1.5 cm, with focal hemorrhage on sectioning.

Histology showed dilated, thin-walled, focally thrombosed vascular channels within a fibrous capsule, with papillary structures showing hyalinized/fibrinous cores on organizing thrombus, consistent with severe papillary endothelial hyperplasia; endothelial cells were plump but non-atypical, without pathological mitoses. Immunohistochemistry showed positive CD34, CD31, and ERG staining, negative D2-40, continuous smooth muscle actin (SMA) staining around papillary structures, and a Ki-67 index of 1–2%; SOX-10 and S100 staining was seen in peripheral border fibers and rare capsular spindle cells. The lesion was interpreted as an intraneural cavernous hemangioma (Fig. 3).

CSF dynamics normalized after resection; the EVD was clamped and removed without recurrence, and the child was discharged with no neurological deficit. Postoperative spinal MRI confirmed complete resection with no residual or recurrent lesion (Fig. 2c, d), and follow-up brain MRI at 6 months showed normal ventricular size without hydrocephalus (Fig. 2f, h, j). At 6-year clinical follow-up, he remained well.

Discussion

Hemangiomas of the spinal nerve roots are classified as capillary or cavernous subtypes. Capillary hemangiomas are more frequently reported, typically presenting in adults with radicular pain [6, 8, 9, 12]; cavernous hemangiomas are rarer and more prone to hemorrhage, which underlies their occasional association with subarachnoid hemorrhage and raised intracranial pressure [2, 13]. An association with communicating hydrocephalus is exceptional in either subtype and has previously been described only in isolated adult cases [2,14,17].

Pediatric reports are scarce: the youngest previously reported case was a 14-year-old with a capillary hemangioma [3], and the only other clearly pediatric report did not present with hydrocephalus [1]. To our knowledge, the present 5-year-old is the youngest reported intraneural hemangioma of the spinal nerve roots, and the first intraneural cavernous hemangioma, in any age group, to present with acute communicating hydrocephalus.

The markedly elevated CSF protein with no cellular reaction in this case represents a relatively pure example of hyperproteinorrhachia, consistent with the protein-related mechanism proposed for spinal-

lesion-associated hydrocephalus (Fig. 4) [4, 11, 14]. Recurrent microhemorrhage from vascular lesions of the cauda equina may additionally provoke arachnoidal scarring and chronic communicating hydrocephalus [10, 13]; this was not evident here, as imaging and cytology showed no hemorrhagic component.

This case underscores that when a child presents with communicating hydrocephalus and elevated CSF protein without an identifiable intracranial cause, the entire spinal axis should be imaged. Temporary CSF diversion can stabilize the child while definitive resection of the causative lesion addresses the underlying pathology and may obviate permanent shunting, as illustrated by the excellent long-term outcome in this case.

Conclusion

Spinal pathology, although rare, should be considered in unexplained pediatric communicating hydrocephalus, particularly when CSF protein is markedly elevated without a cellular reaction. Prompt temporary CSF diversion followed by definitive resection of the causative lesion can achieve durable resolution and avoid permanent shunting.

Declarations

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Ethics approval: This study adhered to the principles of the 1964 Declaration of Helsinki and its later amendments, and to institutional ethical standards. [Insert name of approving institutional/ethics committee and approval or waiver reference number.]

Consent to participate: Written informed consent was obtained from the patient's parents/legal guardians.

Consent to publish: The authors affirm that the patient's parents/legal guardians provided informed consent for publication of the clinical details and accompanying images.

Author Contribution

B.S.: study conception and design, clinical data collection, surgical treatment, literature review, original illustration, and drafting of the manuscript and corresponding author. İ.T.R.: interpretation of radiological imaging. S.U.B.: histopathological and immunohistochemical examination. All authors commented on previous versions of the manuscript and read and approved the final manuscript.

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Figures

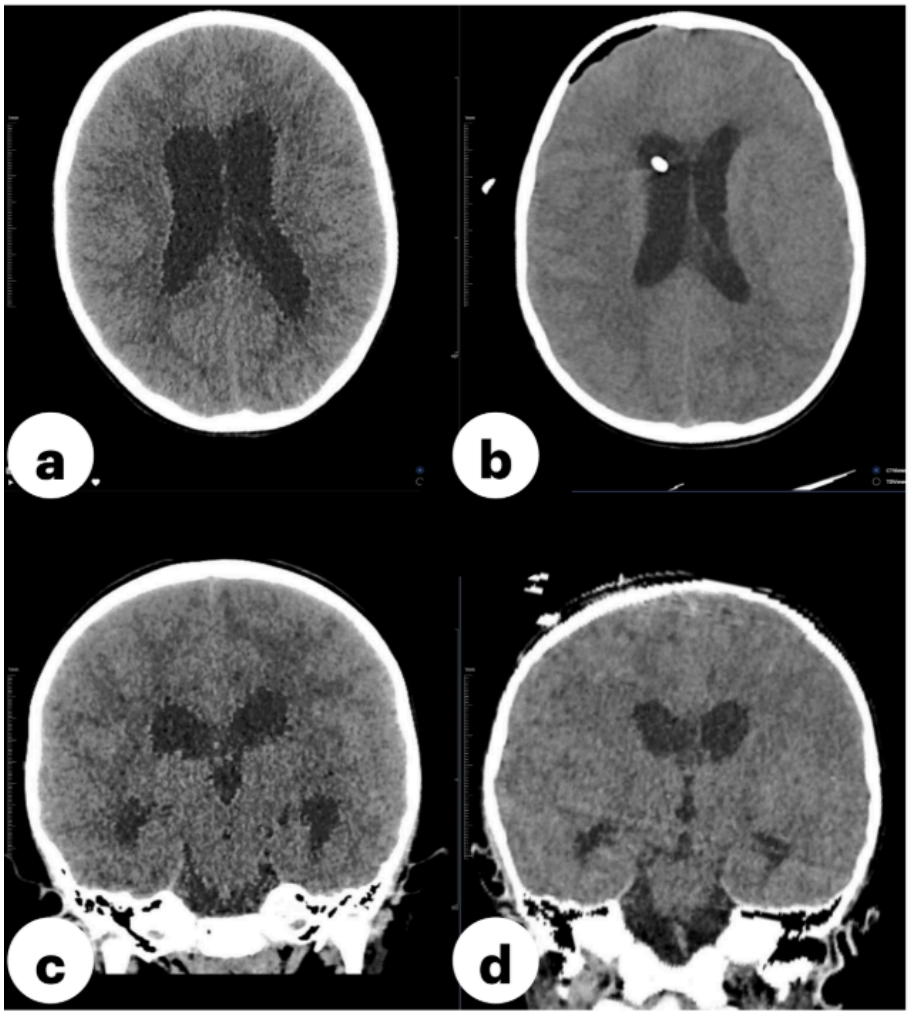


Figure 1

(a, b) Axial and (c, d) coronal cranial CT at presentation showing ventricular dilatation consistent with acute communicating hydrocephalus, before (a, c) and after (b, d) placement of the external ventricular drain, with satisfactory catheter position and reduced ventricular size.

CSF protein was markedly elevated at 286 mg/dL (reference, 15–45 mg/dL), with no red blood cells or cellular reaction; hematological and biochemical work-up was otherwise unremarkable, with no evidence of infection or hemorrhage. Brain MRI confirmed the ventricular dilatation seen on CT (Fig. 2e, g, i), and complete spinal MRI revealed a well-circumscribed intradural mass, 21 × 15 × 15 mm, within the filum terminale fibers at L3–L4, hypointense on T1, hyperintense on T2, and homogeneously enhancing (Fig. 2a, b).



Figure 2

(a, b) Preoperative lumbar spinal MRI showing the intradural tumor. (c, d) Postoperative lumbar spinal MRI showing complete tumor resection. (e, g, i) Preoperative brain MRI showing hydrocephalus. (f, h, j) Postoperative brain MRI at 6-month follow-up showing normal ventricular size, without hydrocephalus.

The patient underwent single-level L3 laminectomy. The lesion was dissected free from the surrounding cauda equina roots under continuous intraoperative neuromonitoring; the parent root was stimulated and, with no motor response elicited, was coagulated before complete removal. The specimen was a lobulated, purple, capsule-like fragment, 2 × 2 × 1.5 cm, with focal hemorrhage on sectioning.

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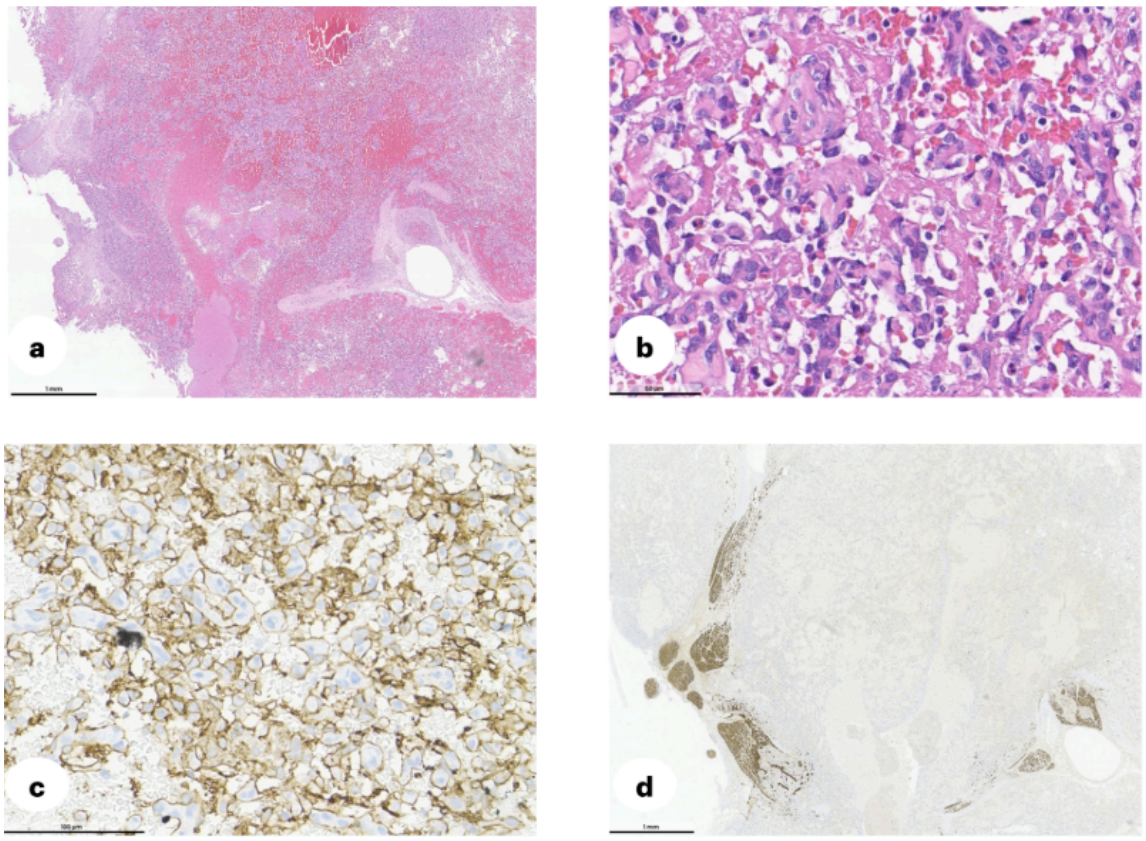


Figure 3

(a) Dilated vascular channels within a fibrous capsule, with organizing thrombus (H&E, ×20). (b) Papillary endothelial hyperplasia with plump, non-atypical endothelial cells (H&E, ×40). (c) Diffuse membranous CD31 positivity in the papillary structures (×400). (d) S100-positive peripheral nerve fascicles at the lesion margin, confirming the intraneural location (×200).

CSF dynamics normalized after resection; the EVD was clamped and removed without recurrence, and the child was discharged with no neurological deficit. Postoperative spinal MRI confirmed complete resection with no residual or recurrent lesion (Fig. 2c, d), and follow-up brain MRI at 6 months showed normal ventricular size without hydrocephalus (Fig. 2f, h, j). At 6-year clinical follow-up, he remained well.

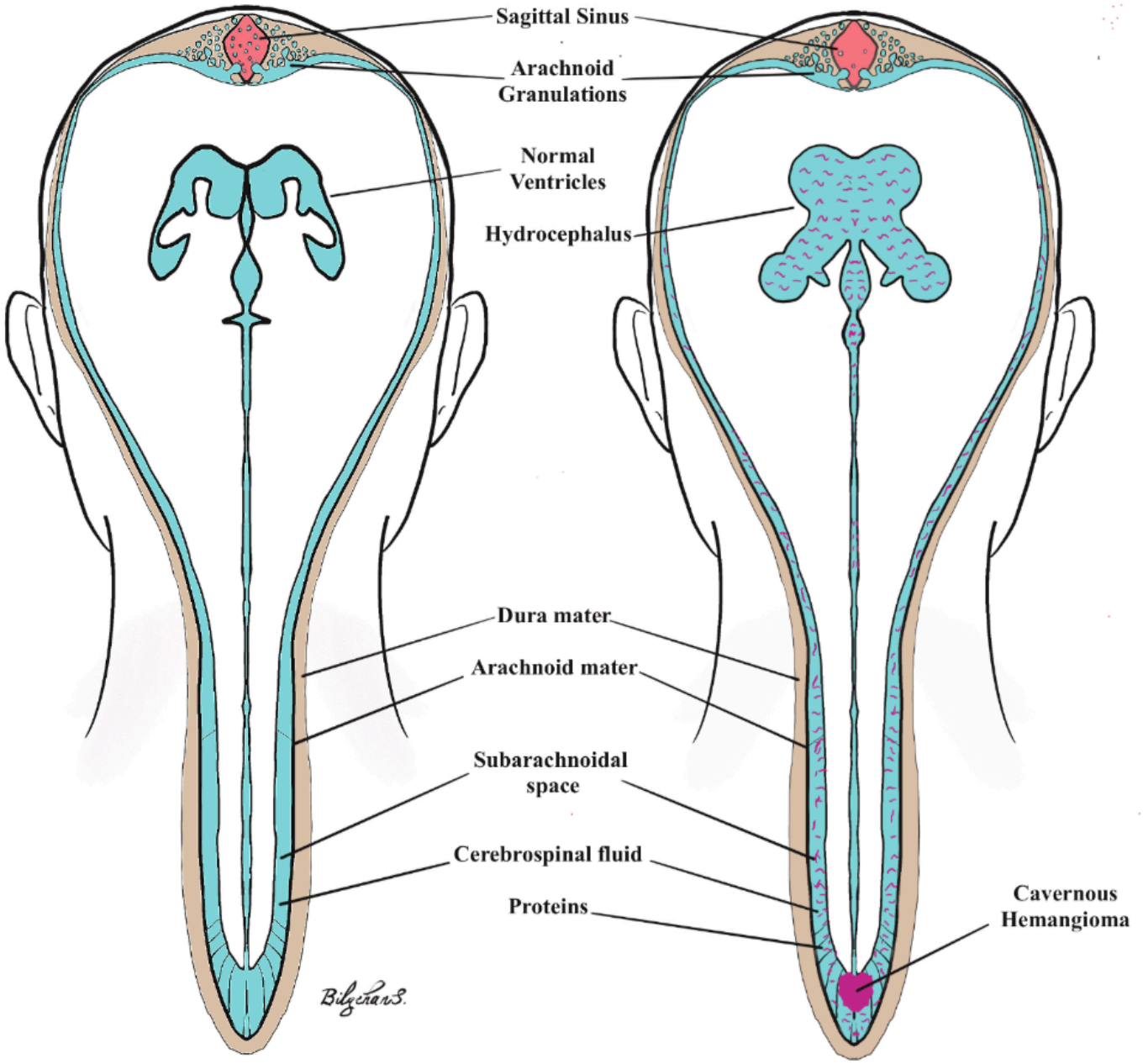


Figure 4

Schematic illustration comparing normal cerebrospinal fluid (CSF) circulation (left) with the proposed mechanism of communicating hydrocephalus in the present case (right). Under normal conditions, CSF passes from the ventricles through the subarachnoid space and is resorbed at the arachnoid granulations into the sagittal sinus. In the presence of a lumbar intraneural cavernous hemangioma, protein released into the CSF increases its viscosity and impairs resorption at the arachnoid granulations, resulting in ventricular dilatation (hydrocephalus).

This case underscores that when a child presents with communicating hydrocephalus and elevated CSF protein without an identifiable intracranial cause, the entire spinal axis should be imaged. Temporary CSF diversion can stabilize the child while definitive resection of the causative lesion addresses the

underlying pathology and may obviate permanent shunting, as illustrated by the excellent long-term outcome in this case.