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IntracranialRapidlyInvoluting Congenital Hemangioma (RICH) in a Neonate: Early Diagnosis and Favorable OutcomeafterVentriculoperitonealShunting — A Case Report

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IntracranialRapidlyInvoluting Congenital Hemangioma (RICH) in a Neonate: Early Diagnosis and Favorable OutcomeafterVentriculoperitonealShunting — A Case Report

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Running title:Intracranial RICH Presenting as NeonatalHydrocephalus

Abstract

Background: Congenital hemangiomas are benignvasculartumorsthat are fullydeveloped at birth and mostcommonlylocated in the skin or subcutaneous tissue. They are characterized by complete maturation in utero and a variable pattern of postnatal involution. Intracraniallocalizationisexceptionally rare.
Case presentation:We report the case of a femaleneonateborn at 38 weeks' gestation with Apgar scores of 9, 10, and 10 at 1, 5, and 10 minutes, respectively, whopresentedwithmacrocephaly and signs of increasedintracranial pressure. Brain computedtomography (CT) revealedtriventricularhydrocephalusassociatedwith a hemorrhagiccerebellar mass, initially suggestive of a hematoma. An emergency ventriculoperitoneal shunt wasperformed.
Subsequentbrainmagneticresonanceimaging (MRI) confirmed the diagnosis of an intracranialcongenitalhemangioma. The lesionwasultimatelyidentified as a rapidlyinvolutingcongenitalhemangioma (RICH). The clinical and radiological course was favorable. At 2 years of follow-up, the patient exhibited normal age-appropriatepsychomotordevelopment, as assessedduring serial pediatricneurological and developmentalexaminations.

Conclusion:Although RICH lesions are typicallycutaneous or subcutaneous, thisexceptionally rare intracranialpresentationshouldbeconsidered in the

differentialdiagnosis of neonatalhemorrhagic central nervous system lesions, particularlywhenimagingfindings are atypical for a simple hematoma.

Keywords

Congenital Hemangioma; Infant, Newborn;Hydrocephalus;CerebellarNeoplasms; Magnetic ResonanceImaging;Ventriculoperitoneal Shunt

Introduction

Vascular anomalies are broadlyclassifiedintotwocategories:vasculartumors and vascular malformations [1]. Congenital hemangiomas are benignvasculartumorsthat are fullyformed at birth, havingreachedtheir maximum size in utero, and are characteristicallydevoid of a postnatal proliferative phase [1,2]. They differfrom infantile hemangiomas, whichappearafterbirth and follow a typicalproliferative phase followed by involution [1,2].

Based on theirnaturalhistory, congenitalhemangiomas are subdividedintothreeclinicalentities:rapidlyinvolutingcongenitalhemangioma (RICH), non-involutingcongenitalhemangioma (NICH), and partiallyinvolutingcongenitalhemangioma (PICH) [2–4]. Intracraniallocalization of congenitalhemangiomasisextremely rare and represents a major diagnostic challenge due to itsatypicalpresentation and resemblance to otherneonatalhemorrhagiclesions [5–7]. To date, only a verylimitednumber of intracranial cases have been reported in the literature, mostly as isolated case reports or smallseries.

We report a rare case of intracranial RICH diagnosed in the neonatalperiod and discuss diagnostic pitfalls, the differentialdiagnosis of neonatalposteriorfossahemorrhagic masses, and the therapeutic implications of thisexceptionalpresentation.

Case Presentation

We report the case of a femaleneonateborn at 38 weeks' gestation withmacrosomia (birthweight 4140 g;length 53 cm;headcircumference 40.5 cm [>97th percentile]) and Apgar scores of 9, 10, and 10 at 1, 5, and 10 minutes, respectively. She wasbornafter an inadequatelymonitoredpregnancy and delivered by cesarean section because of severepre-eclampsia. The neonatewasadmitted to the neonatal intensive care unit for respiratorydistress and vomiting.

Clinicalexaminationrevealedmacrocephaly (headcircumference 40.5 cm, >97th percentile), a bulging and widenedanterior fontanelle communicatingwith the posterior fontanelle, splayedcranial sutures, axial hypotonia, and systemicarterial hypertension.

Transfontanellarultrasonographydemonstratedtriventricularhydrocephalus.

Brain CT revealed a 52-mm leftvermian intra-axial hyperdense mass (Figure 1A), initially suggestive of a cerebellarhematoma, associatedwith active hydrocephalus, transependymaledema, and earlytonsillarherniation. Routine coagulation studieswereunremarkable. Because of signs of increasedintracranial pressure and impendingtonsillarherniation, an emergency ventriculoperitoneal shunt wasperformed. Cerebrospinalfluidanalysis, includingbacteriological, cytological, and biochemicalexaminations, wasunremarkable.

Post-shuntingmagneticresonanceimaging (MRI) revealed a large hemorrhagiccysticlesion in the posteriorfossa, withimagingfeatures suggestive of an intracranialcongenitalhemangioma (Figure 1B and 1C). No histopathologicalexaminationwasperformed, and the diagnosis of rapidlyinvolutingcongenitalhemangioma (RICH) wasestablishedbased on the characteristicclinicalpresentation and radiologicalevolution. The clinical course was favorable, with progressive regression of macrocephaly and normal age-appropriatepsychomotordevelopmentduring serial pediatricneurological and developmentalexaminations over 2 years of follow-up. Follow-up MRI performed 2 yearsafter the initial presentationdemonstratedcompleteresolution of the lesionwithoutresidualabnormalities. The final diagnosiswasintracranialrapidlyinvolutingcongenitalhemangioma (RICH)

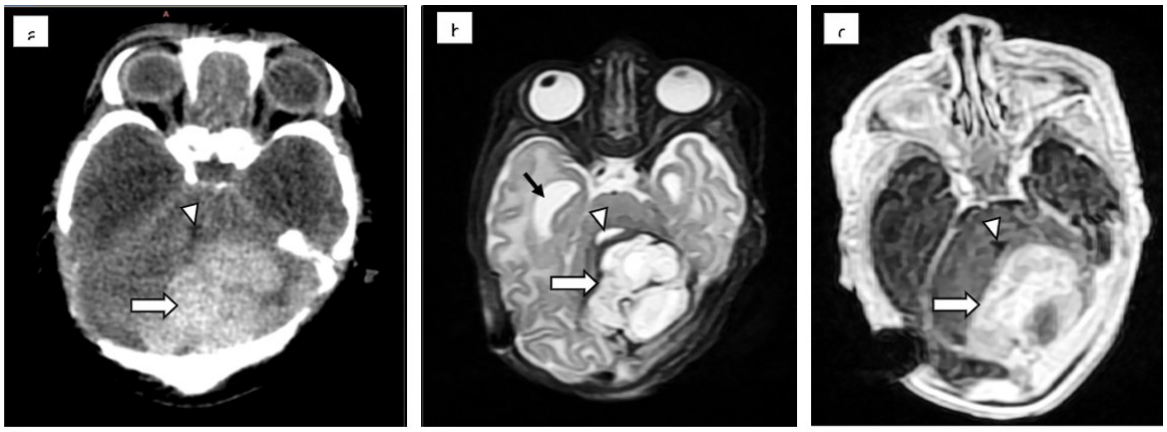


Figure 1. Neuroimagingfindings in a neonatewithintracranialrapidlyinvolutingcongenitalhemangioma (RICH).
(A) Axial non-contrastbrain CT showing an intra-axial, expansile, multiloculated, homogeneously hyperdense mass in the infratentorialcompartment, arisingfrom the leftcerebellarhemisphere (thickarrow). The lesion crosses the midline, compresses and displaces the fourthventriclesuperiorly (arrowhead), extendstoward the quadrigeminalcistern, and isassociatedwithmoderateventriculomegaly.

(B) Axial T2-weighted MRI showing a homogeneouslyhyperintenselesionwithcorrespondingrestricted diffusion on diffusion-weightedimaging and elevated apparent diffusion coefficient (ADC) values (arrows;arrowheadindicatesdisplacedfourthventricle).

(C) Axial post-contrast T1-weighted MRI demonstrating absence of enhancement of the cystic component (arrow;arrowheadindicates the fourthventricle).

Discussion

Pathophysiology and classification of congenitalhemangiomas

Congenital hemangiomas are benignvasculartumorsthat are present at birth, havingcompletedtheirgrowth in utero without a postnatal proliferative phase [1,2]. They differfrom infantile hemangiomas in boththeirnaturalhistory and theirindependencefrom postnatal growthfactors [1,2]. Threeclinical variants are recognized: RICH, NICH, and PICH [2–4]. RICH lesionsundergorapidspontaneous involution, oftenwithin the first year of life, with progressive replacement by fibrofatty tissue [2,3]; NICH lesionsremain stable withoutregression, whereas PICH lesions show only partial involution [2,4].

At the molecularlevel, congenitalhemangiomas have been linked to somatic, mosaic, activatingmissense mutations affecting codon 209 of GNAQ or GNA11, identified in both RICH and NICH specimens [8]. Because the same mutation can befound in lesionswithmarkedlydifferent postnatal trajectories, additionalgenetic, epigenetic, or local microenvironmentalfactors are thought to determinewhether a givenlesioninvolutesrapidly or persists [8]. To date, the molecular profile of intracranialcongenitalhemangiomas has not been specificallycharacterized. No genetic or molecularwork-up wasperformed in the present case due to the absence of available tissue samples.

Rarity of intracraniallocalization

Intracranialcongenitalhemangiomas are extremely rare and onlysporadicallyreported in the literature [2,5]. Thesetumors are typicallycutaneous or subcutaneous, with a predilection for the head and neck region [4,7]. Visceralinvolvement, particularlyhepatic, has also been described, but central nervous system involvementremainsexceptional [6,7]. Intracranial RICH isconsideredanecdotal, whichmakesitsdiagnosisparticularlychallenging in the neonatalperiod [5,7]. This rarityoften leads to misdiagnosis as neonatalcerebellarhemorrhage or posteriorfossatumors, as occurred in our case [5,7]. Vasculartumorsshouldthereforebeconsidered, despitetheirrarity, whenevaluatingneonatalintracranialhemorrhagic-appearinglesions [6,7].

Differentialdiagnosis

In neonatespresentingwith an intracranialhemorrhagic-appearing mass, cerebellarhemorrhageis the mostlikely initial diagnosis, particularlywhen CT demonstrates a hyperdense lesion in a high-riskperinatalcontext. In the present case, the onlyidentifiedperinatalriskfactorswereseverepre-eclampsia and macrosomia, with no history of prematurity, instrumental delivery, birth trauma, or neonatalcoagulopathy.

Intracranialvascular malformations, such as arteriovenous malformations and cavernomas, shouldalsobeconsidered, althoughthey are rare in neonates [2,5]. Congenital intracranialtumors, includingteratomas and primitive neuroectodermaltumors, maylikewisepresent as hemorrhagicposteriorfossalesions. Hemangioblastomamayalsobeconsidered in the differentialdiagnosis, althoughitisexceptional in thisage group [5]. A comparison of the clinical and imagingfeatures of theseentities highlights key differencesfromintracranial RICH, particularlyregardingonset, imagingcharacteristics, and lesionevolution over time.

Diagnostic value of neuroimaging

Neuroimagingplays a central role in the evaluation of neonatalintracraniallesions, althoughinterpretation can bechallenging in the acute phase. In our case, CT demonstrated a 52-mm hyperdense posteriorfossalesionassociatedwithtriventricularhydrocephalus, transependymaledema, and earlytonsillarherniation, whichinitiallysuggested a cerebellarhematoma. However, CT has limited tissue characterizationcapability in neonates, and hyperdensitymayreflecteitherhemorrhage or a vasculartumor [5].

MRI is the gold standard for lesioncharacterization, allowingbetterassessment of solid, cystic, and hemorrhagic components, as well as the relationship of the lesion to posteriorfossa structures [6]. Congenital vasculartumorsmay show heterogeneous T1 and T2 signal, withhemorrhagic and cystic areas reflectingtheirnaturalevolution [6,7];somereported RICH lesionsalso display areas of heterogeneity, prominent flow voids, and feeding-vesselaneurysms on MRI and angiography [3]. In our case, MRI demonstrated a hemorrhagiccysticposteriorfossalesionwithoutevidence of tumor progression, supporting a RICH lesion in the process of involution.

No single imagingmodalityisfullyspecific in the acute phase, and diagnosisultimately relies on the correlationbetweenimagingfindings, clinicalevolution, and the overallneonatalcontext [6]. This diagnostic uncertaintyisitself a key message of ourreport: in a neonatewith an atypicalposteriorfossahemorrhagic mass and no cleartraumatic or hematologicexplanation, serial imagingdemonstrating progressive involution — ratherthangrowth — shouldraise suspicion of a congenitalvasculartumorratherthan a truehematoma or a malignant process.

Management and outcome

Initial management wasdictated by triventricularhydrocephaluswithsigns ofintracranial hypertension and a risk of herniation [5,6]. Emergency ventriculoperitonealshuntingwasperformed to achieve effective cerebrospinalfluid diversion, consistent with the standard management of acute neonatalhydrocephalus [5,6].

The possibility of immediatebiopsy or surgicalresectionwas not pursued in the acute setting. This decisionwas made in a multidisciplinarycontext and wasprimarilybased on the criticalneurological condition requiring urgent intracranial pressure control, the deepposteriorfossa location of the lesion, and the

absence of a definitivepreoperativediagnosis. A conservative diagnostic approachwasfavored, withfurtheretiological clarification plannedaccording to clinical and radiologicalevolution.

Postoperativeevolutionwas favorable, withclinicalimprovement and resolution of intracranial hypertension. Follow-up imagingshowedevolutiontoward a cysticlesionwithhemorrhagicremodeling, consistent with an involutingcongenitalvascularlesion [2,4,7]. No shunt-related complications wereobserved.

At 2-year follow-up, the childhad normal psychomotordevelopment and normal MRI findings. The patient remainedclinically stable withoutevidence of raisedintracranialpressure;however, long-term shunt dependencycould not bedefinitivelyassessed, and no shunt revisionwasrequiredduring the follow-up period.

This evolution supports the diagnosis of rapidlyinvolutingcongenitalhemangioma (RICH), a benignvasculartumorcharacterized by rapidspontaneous involution during the first year of life [2–4].

Strengths and clinical implications of this report

This case adds to the verylimited body of literature on intracranialcongenitalhemangiomas. To ourknowledge, itisamong the rare reported cases in which the diagnosis of rapidlyinvolutingcongenitalhemangioma (RICH) wasestablishedwithouthistopathological confirmation, relyinginstead on characteristicradiologicalfindings and theirevolution over time.

It highlights the importance of consideringcongenitalvasculartumors in neonateswithintracranialhemorrhagic-appearinglesions and atypicalclinical or radiologicalfeatures, underscores the limitations of CT in the acute phase, and confirms the central role of MRI—ideallycomplemented by vascularimaging—in lesioncharacterization. Importantly, management of the acute, life-threatening complication (hydrocephalus) shouldremainprimarilysymptomatic and urgent, regardless of whether a definitiveetiologicaldiagnosis has been established. The favorable outcome in our patient is consistent with the generallybenignprognosis of RICH whenintracranial hypertension ispromptlyrecognized and managed.

Limitations

This report has several limitations. First, and mostimportantly, the diagnosis of intracranialrapidlyinvolutingcongenitalhemangioma (RICH) wasestablished on

clinical and radiological grounds withouthistopathological or immunohistochemical (GLUT1) confirmation, whichremains the diagnostic reference standard for congenitalvasculartumors. Histopathological confirmation would have been particularlyvaluablegiven the broaddifferentialdiagnosis of hemorrhagicposteriorfossalesions, includingmalignanttumors. Second, thisis a single-case report, whichinherentlylimits the generalizability of our observations. Third, follow-up waslimited to 2 years and thereforecannotexclude the possibility of late complications, particularly shunt-related complications or, more rarely, delayed local recurrence. Finally, because of the absence of tissue samples, no genetic or molecular analyses (including GNAQ/GNA11 mutation testing) wereperformed. In addition, the extremerarity of intracranial RICH limits direct comparisonwithpreviouslypublished cases.

Future directions

Furtherstudies are needed to bettercharacterizeintracranialcongenitalhemangiomas, particularlywith respect to theirclinicalpresentation, imagingfeatures, molecular profile, and long-termoutcomes. The accumulation of well-documented, ideallyhistologicallyconfirmed cases—potentiallythrough international vascularanomalyregistries—wouldimproveearly recognition and help refine diagnostic criteriaspecific to the intracranial location. A betterunderstanding of imagingcharacteristicsthatdistinguishcongenitalhemangiomasfromhemorrhages or malignanttumorscould help reduceunnecessary invasive procedures in this fragile population. Long-termneurodevelopmental follow-up usingstandardizedassessmenttoolsremains essential to fullycharacterizeoutcomesbeyondearlychildhood.

Conclusion

This case illustrates an exceptionalpresentation of intracranial RICH revealed by neonatalhydrocephalus. The main diagnostic challenge was the initial radiologicalmisinterpretation of the lesion as a cerebellarhemorrhage. This case emphasizesthatcongenitalvasculartumorsshouldbeconsidered in the differentialdiagnosis of neonatalintracranialhemorrhagic-appearinglesions, eventhoughsuchtumors are rare in this location. The spontaneous, favorable evolutionobserved in our patient confirms the generally good prognosis of RICH whenassociatedintracranial hypertension ispromptlyrecognized and managed.

Déclarations

Ethicsapproval and consent to participate

This case wasconducted in accordance with the ethical standards of Farhat Hached University Hospital and the Declaration of Helsinki. Formalethicsapprovalwas not required for this single anonymized case report.

Consent for publication

Writteninformed consent wasobtainedfrom the patient’s parents/legalguardian for publication of this case report and accompanying images.

Availability of data and materials

Data are availablefrom the correspondingauthoruponreasonablerequest.

Competinginterests

The authorsdeclare no competinginterests.

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Authors' contributions

Donia Brahemmanaged the patient and supervised the study. Chadha Chouchene collected data, reviewed the literature, and drafted the manuscript. Both authorsrevised and approved the final manuscript.

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