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CASE REPORT

Enhancing nodular lesions in Chiari II malformations in the setting of persistent hindbrain herniation: case report and literature review

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Abstract

Background Chiari II malformation includes concomitant cerebellar tonsillar herniation, hydrocephalus, and myelomeningocele. Rarely, pediatric patients with persistent hindbrain herniation develop a new enhancing nodule at the cervicomedullary junction as adults. These new lesions may be suspicious for neoplastic growth, but it remains unclear if neurosurgical intervention is necessary.

Case Report A 27-year-old female patient with history of Chiari II malformation and persistent hindbrain herniation presented with a 3-month history of headache and upper extremity weakness and numbness. Neuroimaging revealed a new enhancing nodule near the cervicomedullary junction suspicious for neoplasm. Following posterior fossa decompression and excision of the enhancing lesion, pathological analysis demonstrated only benign glioneural heterotopia.

Results New enhancing nodules at the cervicomedullary junction in Chiari II malformation are exceedingly rare and are likely benign, reactive changes rather than a neoplastic process. Biopsy or surgical excision of these lesions is likely unnecessary for asymptomatic patients.

Keywords Chiari II malformation · Pediatrics · Neurodevelopmental disorders

Introduction

Chiari II malformation is characterized by the herniation of hindbrain structures through the foramen magnum, a small posterior fossa, hydrocephalus, and myelomeningocele; its etiology remains largely unknown [3, 14]. Initial management typically involves treatment of hydrocephalus and meningomyelocele repair [4, 7]. Infants with life-threatening symptoms of brainstem compression may also undergo surgical decompression of the posterior fossa via cervical laminectomy and/or suboccipital craniectomy [14]. Children with asymptomatic or equivocal presentations may not need surgical decompression and so have persistent hindbrain herniation on neuroimaging as adults. There are no clear guidelines for follow-up surgical management of these patients due to variability in natural history and clinical presentation [4, 15].

Chiari II malformation patients with persistent hindbrain herniation may develop additional associated pathologies [4]. Commonly associated lesions, such as syringomyelia or cystic lesions, have classic, recognizable features on neuroimaging [7]. However, solid, contrast-enhancing nodules at the cervicomedullary junction have a more complex differential diagnosis, one which must include neoplasms such as pilocytic astrocytoma, ganglioglioma, low-grade glioma, or hemangioblastoma [1, 6]. A recent review of these pediatric cervicomedullary tumors (CMTs) recommends either surgical excision or open biopsy for suspicious lesions, depending on symptomatology [6]. The decision to intervene on radiographic abnormalities in Chiari II malformation patients is also largely based on symptom severity [15]. This approach falls short in the case of a newly discovered lesion that is asymptomatic but suspicious for neoplasm.

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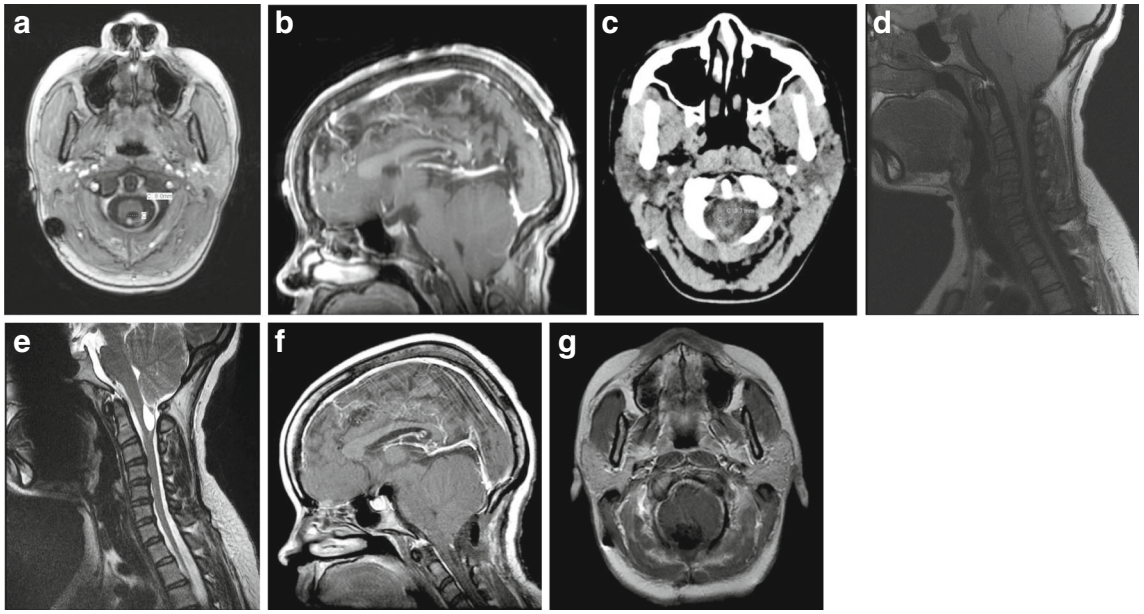

Fig. 1 **a-e** Pre- and **f-g** post-operative imaging for 27-year-old female with Chiari II malformation and enhancing nodular mass at the cervicomedullary junction. **a** Axial and **b** sagittal T1-weighted MRI with contrast showing a 2.2 × 1.1 cm non-enhancing intramedullary cystic lesion at the C1-2 spinal level and 3.5 × 3.0 mm enhancing nodule. **c** Axial computed-tomography scan of the head obtained 1.5 years prior showing interval growth of the cystic lesion. **d** Preoperative T2-weighted MRI of the cervical spine showing the cystic lesion and enhancing nodule. Postoperative **e** axial and **f** sagittal T1-weighted MRI of the brain showing resection of enhancing nodule and decompression of the cystic lesion at C1-2

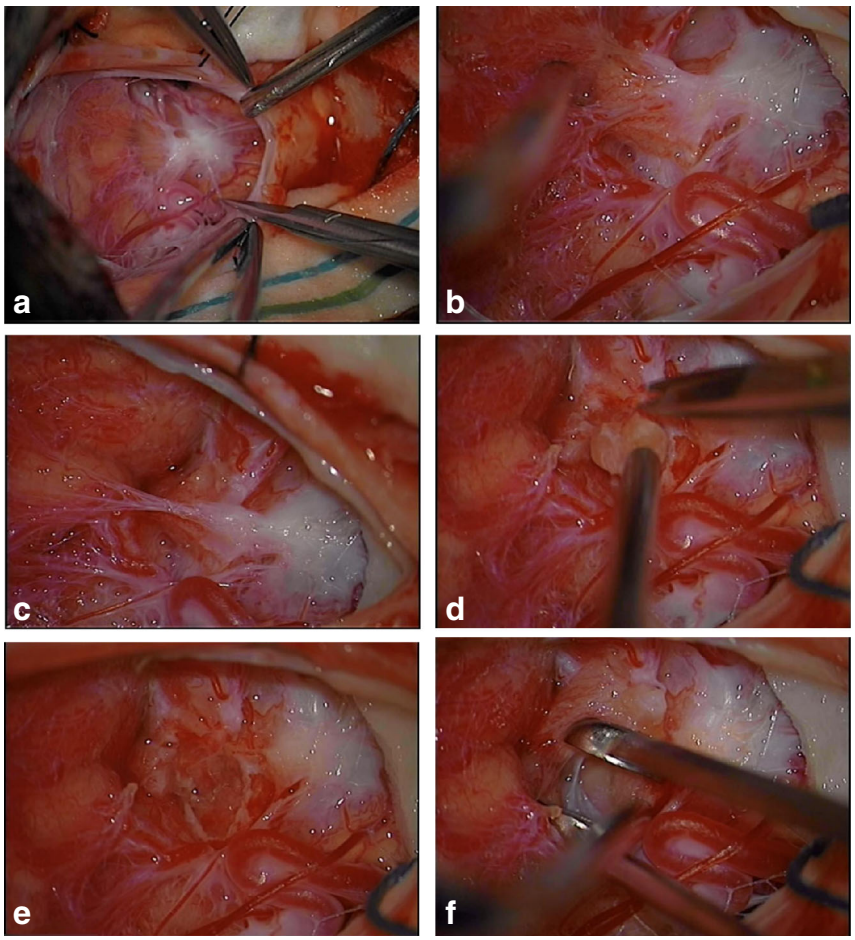

Fig. 2 Intraoperative images obtained from surgical microscope showing **a** thickened arachnoid band from cerebellum to upper cervical spinal cord, covering the cerebellar tonsils and foramen of Magendie, **b** dissection of enhancing nodules overlying the obex and cervical spinal cord (left posterior inferior cerebellar artery seen in operative field), **c** final dissection of arachnoid bands, **d** en bloc removal of enhancing nodule, **e** resection cavity, and **f** fenestration of the cystic lesion

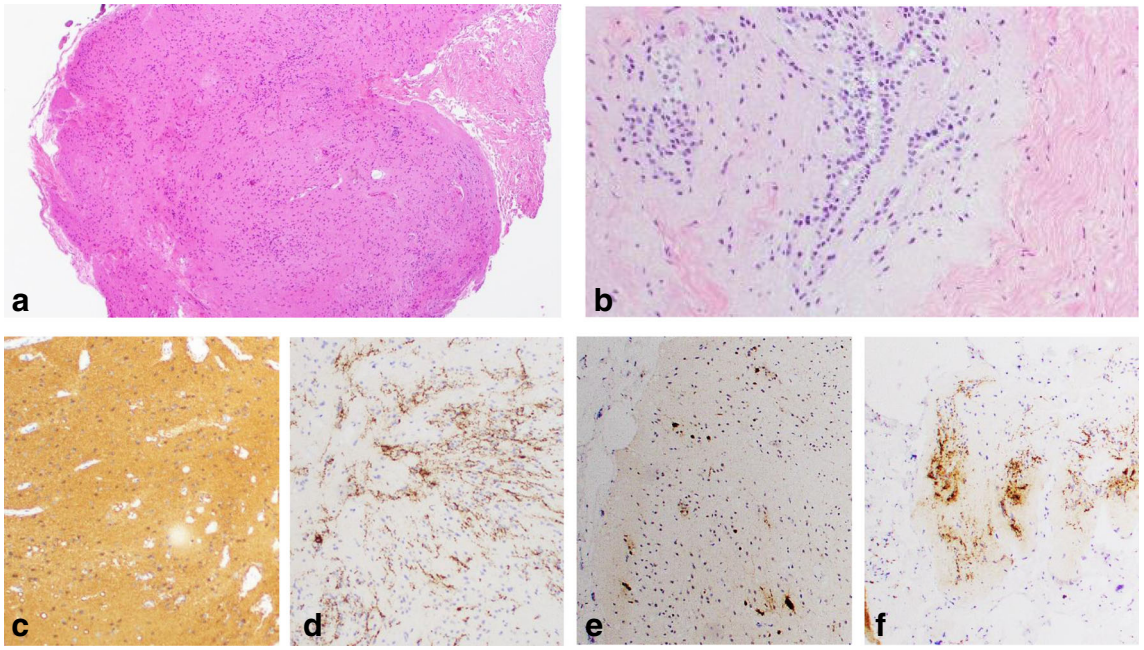

Fig. 3 Histopathology of the surgical specimen obtained from the enhancing nodule. **a** Hematoxylin and eosin staining shows a variably cellular lesion with lobular architecture, surrounded by fibrous tissue, × 40 magnification. **b** Same preparation at × 200 magnification shows haphazardly distributed glial cells and rare neurons in a dense background of neuropil, with focal ependymal tubule formation. **c** Glial fibrillary acidic protein (GFAP) immunohistochemistry demonstrates diffuse distribution of astrocytes at × 100 magnification, while **d** synaptophysin (× 200 magnification) and (**e**) HuD (× 100 magnification) immunostaining labels scattered neurons. **f** Immunohistochemistry for neurofilament protein labels neuritic processes at the edge of the nodular lesion (× 200 magnification)

We present the case of a patient with history of Chiari II malformation who was found to have a new, enhancing nodule near the cervicomedullary junction in the setting of persistent hindbrain herniation. Literature review shows these cases are extremely rare; implications for pathophysiology and surgical management are discussed.

Case report

A 27-year-old female with past medical history of Chiari II malformation, spina bifida, and hydrocephalus, status post ventriculoperitoneal (VP) shunt placement, presented with progressively worsening headache and bilateral upper extremity weakness and numbness over 3 months. Magnetic resonance imaging (MRI) of the cervical spine revealed a 3 mm solidly enhancing nodule associated with a 2.2 cm non-enhancing intramedullary cystic lesion at the C2 level; interval growth was demonstrated in comparison with prior computed tomography (CT) head scan obtained 1.5 years prior (Fig. 1a–e). Differential diagnosis included hemangioblastoma, astrocytoma, or ependymoma. Previous cervical spine imaging was not available for comparison. Shunt tapping and nuclear medicine studies ruled out shunt malfunction. Given the progressive neurological symptoms, the patient was brought to the operating room for suboccipital craniectomy, C1-C2 laminectomy, expansile duraplasty, and excision of the nodule. The cystic lesion was found to be a syrinx at the base of the fourth ventricle that extended inferiorly into the cervical spine; the nodule was located at its dorsal aspect. The lesion included a thick, fibrous webbing of arachnoid covering the cerebellar tonsils and foramen of Magendie, preventing outflow from the fourth ventricle (Fig. 2a–c). The nodule was resected, and the syrinx was fenestrated (Fig. 2d-f). Pathological analysis of frozen sections was most consistent with benign glioneural heterotopia (Fig. 3a-f). Postoperative imaging demonstrated gross total resection of the enhancing nodule and decompression of the posterior fossa and cystic lesion (Fig. 1f-g). The patient recovered well from the procedure and reported complete resolution of her upper extremity weakness and numbness. Patient remains asymptomatic at 6 months of follow-up.

Discussion

We present a patient with Chiari II malformation with persistent hindbrain herniation who was found to have an enhancing nodule near the cervicomedullary junction. Although there was suspicion for neoplasm, pathology demonstrated benign

Table 1 Review of literature on enhancing nodular changes in Chiari II malformation

Study	# of Cases	Age (range, years)	Presentation (age, years)	Preoperative imaging	Management	Histopathology	Conclusion
Stark et al. 1992 [13]	3	2–11	Suspected syringomyelia or tethered cord.	T1-weighted MRI post-contrast: enhancing nodular tissue at the caudal tip of the cerebellar vermis.	Associated cyst resection (2), shunting of cervical syrinx (1). No resection of nodule.	Not performed.	Ectopic choroid plexus tissue from fourth ventricle
Singla et al. 2012 [12]	1	22	Progressive pain and weakness in neck, back, upper extremities for 3 months.	T1-weighted MRI post-contrast: Partially enhancing lesion within upper cervical cord extending into fourth ventricle.	C1-C2 laminectomy, resection of nodular mass	Papillary fronds of collagenous tissue lined by ependymal epithelium, occasional ependymal rosettes	Dysplastic-reactive choroid plexus
Piatt and D'Agostino, 1999* [11]	5	2–26	Case 1 (26): Progressive weakness and atrophy of hands Case 3 (13): Kyphoscoliosis Case 4 (13): Kyphoscoliosis Case 6 (4): Scoliosis Case 8 (2): Syringomyelia	Not seen Not seen Not seen T2-weighted MRI post-contrast: multiple nodular lesions in fourth ventricle isointense to brain Not seen	C1-C3 laminectomy, nodule discovered within fourth ventricle and resected Not described Not described C1-C4 laminectomy, nodular lesions resected from the roof Not described	Clusters of glial cells surrounded by dense glial matrix Atrophic cerebellum Not described Collagen with cuboidal ciliated epithelial cells, hydropic avascular choroid plexus villi Not described	Subependymoma Atrophic cerebellum Subependymoma Atrophic choroid plexus with ependymal features Gliotic cerebellar tissue

* Article [11] describes eight cases in total, but only five discussing nodular lesions are included

glioneuronal heterotopia. Previous studies support an association between Chiari II malformation and benign lesions at the cervicomedullary junction, such as cysts, syringes, arachnoid adhesions, and scar tissue, among others [2, 4, 8, 10, 16]. However, few (< 5) reports of enhancing solid nodules exist, highlighting their rarity.

Literature review via database search of PubMed identified nine cases of solid nodular lesions in Chiari II malformation from three articles published between 1992 and 2018 (Table 1**)** [11–13]. Prior surgical decompression had not been performed in any patient. The nodules were classified as ectopic choroid plexus, subependymoma, or glial tissue. In Piatt and D'Agostino [11], some nodular lesions were discovered intraoperatively, not on preoperative imaging. Nonetheless, these cases reinforce that such nodular lesions are likely benign. In Stark et al. [13], biopsy and histopathological analysis were not performed, yet the authors conclude that the enhancing nodular lesion was most likely benign ectopic choroid plexus.

Of note, the enhancing nodule in our patient consisted of glioneuronal heterotopia, unlike previous reported cases. While this pathology is associated with other neurodevelopmental disorders such as encephalocele and meningocele, it has not yet been described in Chiari II malformation [5, 9].

We see two potential etiologies for our patient's enhancing nodule. One theory of glioneuronal heterotopia is that it is an early developmental aberration of neuroepithelial cell migration or forebrain vesicle development [5, 9]. However, the lesion in our patient was a new finding in an adult. Alternatively, the nodule may be a reactive change to longstanding hindbrain compression and ischemia [12]. A combination of both theories could be consistent with our patient's history of persistent hindbrain herniation as compression may cause ectopic tissue displacement into the cervical cord. This explanation has been proposed previously to explain nodules comprised of ectopic choroid plexus in Chiari II malformation [11]. Similarly, we propose that solidly enhancing nodules at the cervicomedullary junction in Chiari II malformation are most likely benign, reactionary changes secondary to prolonged hindbrain compression.

Although our patient underwent surgical decompression for her progressive symptoms, serial imaging may be preferable in asymptomatic patients with enhancing nodules over biopsy or surgical excision since these lesions are likely benign. Long-term follow-up on the natural history of these lesions should be performed to continue this discussion.

Conclusion

Enhancing lesions near the cervicomedullary junction in patients with Chiari II malformation and persistent hindbrain

herniation may raise concern for neoplastic process. Here we conclude that: (1) longstanding compression of hindbrain structures may result in pathological fibrous tissue changes and thus (2) the presence of an enhancing posterior fossa lesion in Chiari II malformation should not be presumed to be a tumor. In asymptomatic patients with these lesions, serial imaging may be preferred over surgical intervention.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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