

Adult Chiari Type 1 Malformation with Holocord Syringomyelia Associated with Sagittal Synostosis

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Craniosynostosis associated with Chiari malformation (CM) is usually found in infants with an underdeveloped posterior fossa. We here present a case of adult craniosynostosis, CM, and symptomatic syringomyelia caused by the protrusion of the posterior rim of the foramen magnum without a tight posterior fossa. A 22-year-old woman with an abnormal head shape and forearm hypesthesia was given a diagnosis of sagittal suture synostosis with CM and syringomyelia caused by foramen magnum stenosis. She underwent foramen magnum decompression with a C1 laminectomy without cranial vault expansion or duraplasty. Her symptoms and radiographical findings improved after surgery. In cases of non-operative craniosynostosis with CM, clinicians should be alert to late-onset syringomyelia and choose surgical strategies according to the pathophysiology.

Keywords: adult craniosynostosis, Chiari type 1 malformation, syringomyelia

Introduction

Syndromic craniosynostosis is often associated with Chiari malformation (CM),^{1,2} and there are a few reports of non-syndromic sagittal suture synostosis associated with CM.^{2–4} CM associated with craniosynostosis is thought to result from abnormal skull development.¹ Patients with sagittal synostosis can have a disproportionate displacement of the occipital pole and a posteroinferior shift of the hindbrain.^{2,5} CM with craniosynostosis is usually diagnosed in infancy and treated by supratentorial cranial vault expansion with or without infratentorial cranial expansion such as foramen magnum decompression (FMD).^{1,2,6}

We here present a case of adult syringomyelia with CM and sagittal synostosis resulting from the protrusion of the posterior rim of the foramen magnum without a tight posterior fossa. This patient was treated successfully with FMD.

Case Report

A 22-year-old woman with an abnormal head shape but no previous diagnosis of craniosynostosis or developmental

delay presented with a few years' history of hypesthesia in both forearms. A computed tomography (CT) scan to detect head trauma revealed sagittal synostosis, Chiari type 1 malformation (CM-1), syringomyelia, and ventriculomegaly without a traumatic lesion. The sagittal synostosis was associated with oxycephaly rather than scaphocephaly. The skull was thicker than usual, especially in the occipital bone and cranial base. The foramen magnum was stenosed due to a protruding posterior rim (Fig. 1). The patient was referred to the neurosurgery department at University of Niigata Hospital. Physical examination revealed hypertelorism, proptosis, and midfacial hypoplasia. Magnetic resonance imaging (MRI) of the head revealed open cerebrospinal fluid (CSF) space and cerebellar sulci at the infratentorial supracerebellar lesion as well as the cerebellar convexity. However, the CSF space around the foramen magnum was too narrow to open the sulci (Fig. 2a). Her posterior cranial fossa volume was 185.6 ml.⁷ The size of posterior cranial fossa was within the normal range.⁷ There were reduction of both anterior-posterior diameter (27.6 mm), transverse diameter (22.6 mm), inferior outlet area (337.4 mm²), and superior outlet area (473.2 mm²) at the foramen magnum.⁷ Whole-spine MRI showed CM-1 and holocord syringomyelia (Figs. 2b and 2c). Cerebral angiography showed occlusion of the right transverse to the sigmoid sinus at the venous phase, and that the bilateral occipital sinuses were the main source of blood flow to the extracranial lesions (Fig. 2d). We treated the CM and the holocord syringomyelia by FMD with a C1 laminectomy and removal of the ossified fibrous band at the foramen magnum. The postoperative course was uneventful. The frequency of hypesthesia decreased in both forearms. The foramen magnum was successfully decompressed, and the size of the syringomyelia decreased (Figs. 3 and 4).

Discussion

Two important findings were obtained from this case: First, spontaneous skull remodeling caused the rim of the posterior foramen magnum to protrude, demonstrating that foramen magnum stenosis can contribute to CM and symptomatic syringomyelia in adults. Second, we should determine the most important factor in symptomatic syringomyelia based on the pathogenesis, and determine treatment accordingly.

CM is often associated with syndromic craniosynostosis,^{1,2} but rarely with non-syndromic single suture synostosis, especially sagittal synostosis.^{2–4} These conditions are usually diagnosed in infancy, or in pre-adolescence at the latest.^{2–4,8} This is the first report of adult symptomatic syringomyelia

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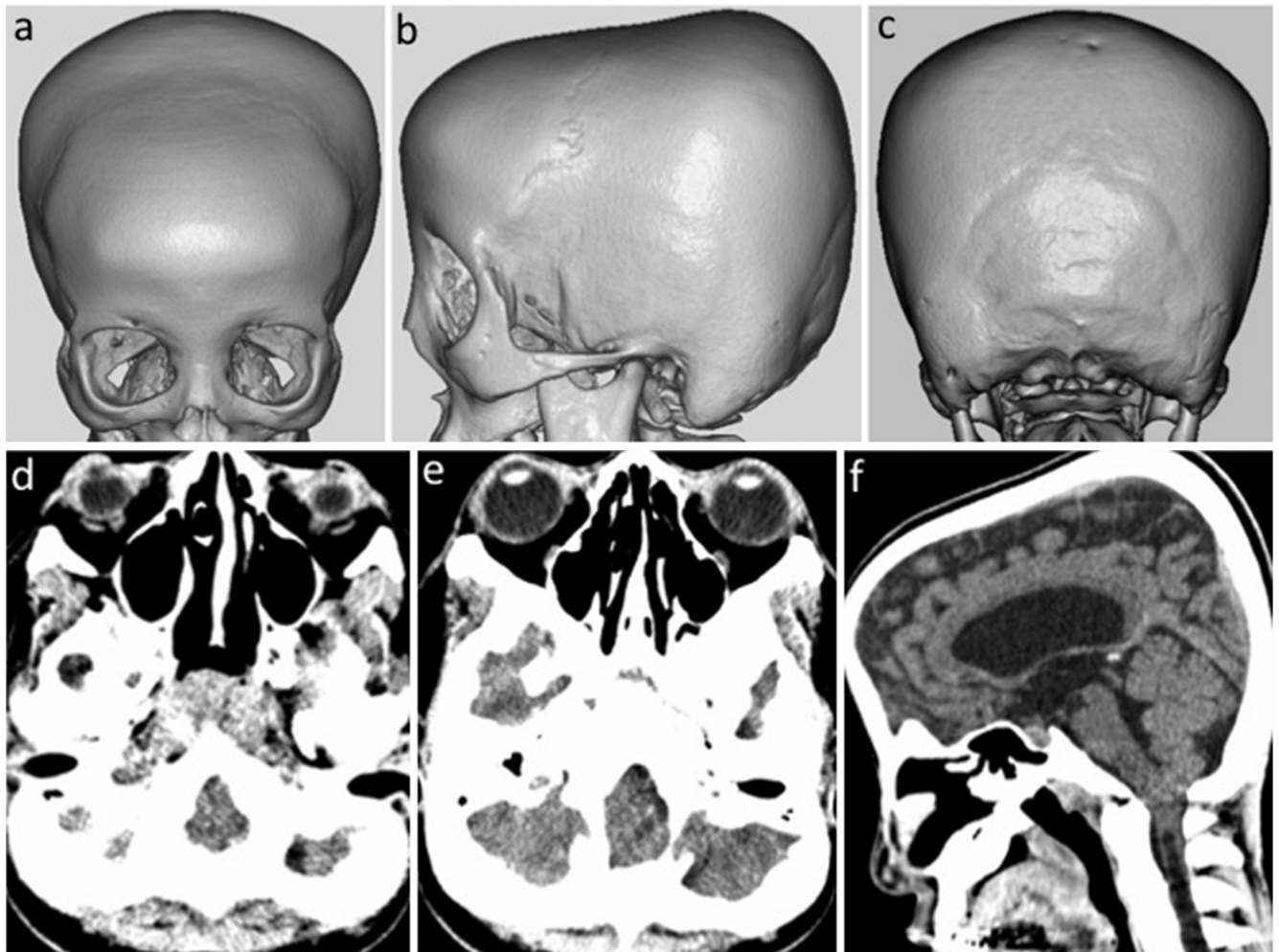


Fig. 1 (a–c) Preoperative 3-dimensional CT revealed the absence of a sagittal suture but no scaphocephaly. (d–f) Preoperative CT showed protrusion of the posterior rim of the foramen magnum and a tight foramen magnum.

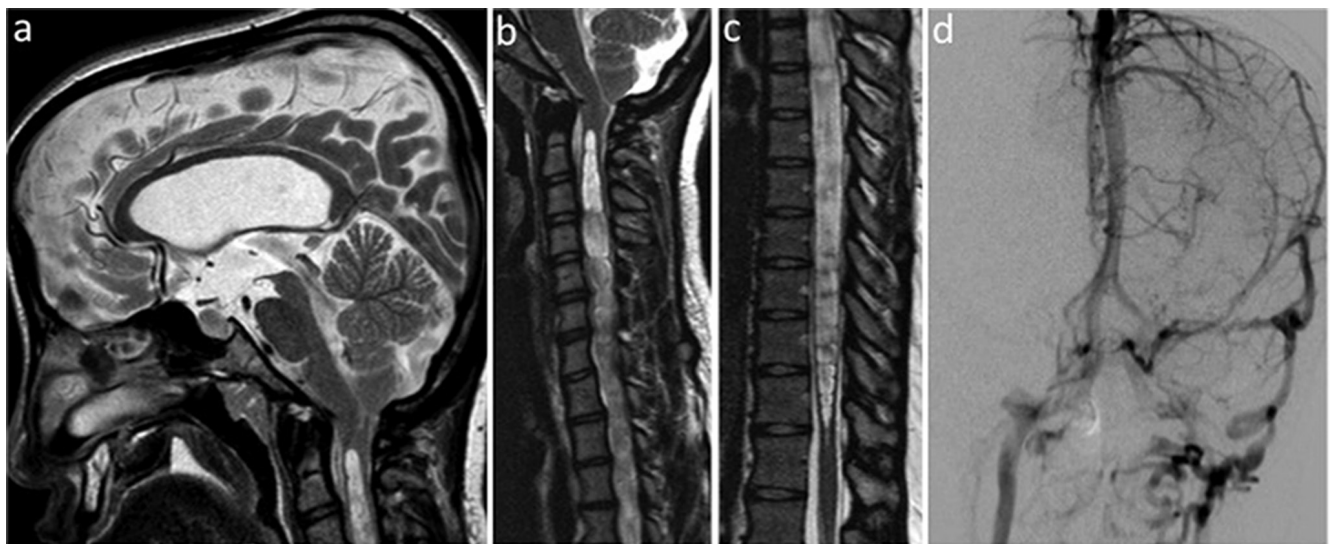


Fig. 2 Preoperative MRI showed tonsillar herniation to the foramen magnum, CM-1 (a), and holocord syringomyelia (b, c). Cerebral angiography showed occlusion of the right transverse to the sigmoid sinus at the venous phase, and that the bilateral occipital sinuses were the main source of blood flow to the extracranial lesions (d).

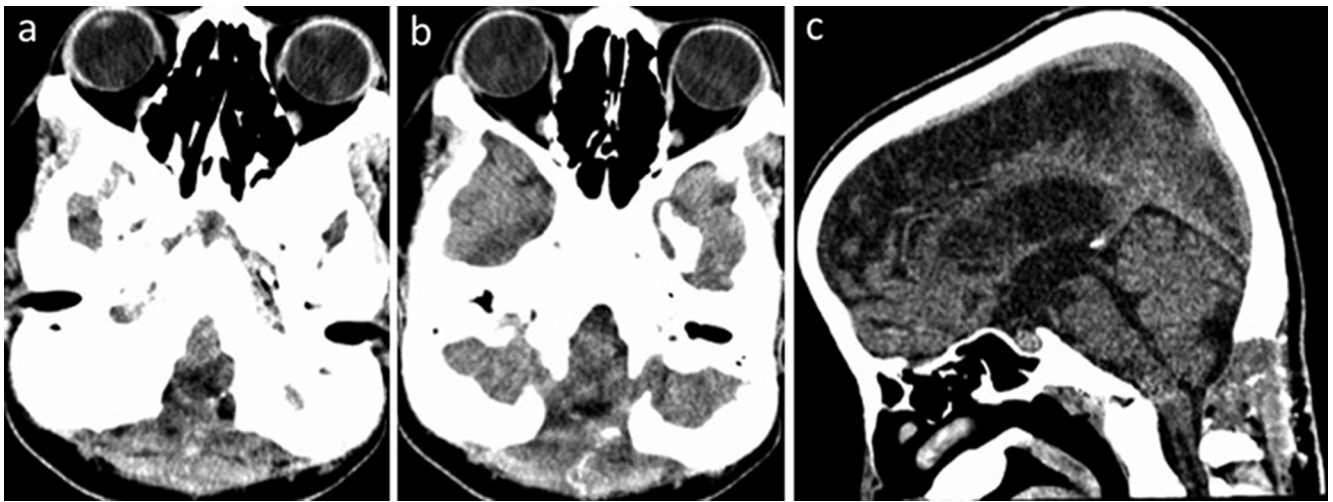


Fig. 3 (a–c) Postoperative CT showed successful decompression of the foramen magnum and open CSF space from behind the cerebellar tonsil to the medulla.



Fig. 4 Postoperative MRIs: 3 months after surgery, the syringomyelia had decreased in size (a, b), it was even smaller 2 years after surgery (c, d).

induced by CM coexisting with sagittal suture synostosis. We thought that cerebellar tonsil of this case herniated only a little into the foramen magnum in the preoperative sagittal MRI. Syringomyelia without tonsillar herniation that responds to posterior fossa decompression is diagnosed as Chiari type 0 malformation (CM-0).^{9,10} We observed carefully and judged this case as CM-1. CM with craniosynostosis is associated with an underdeveloped occipital bone and small posterior fossa.² In sagittal synostosis, compensatory growth can occur at the lamboid suture; this displaces the

cerebellar tentorium to the inferior and is thought to cause hindbrain herniation.⁴ In our case, however, the patient did not have a small posterior fossa, and she had sufficient cerebrospinal fluid space at the infratentorial supracerebellar cortex to open the cerebellar sulci. Her foramen magnum was stenosed due to the protrusion of the posterior rim of the foramen magnum. This foramen magnum stenosis produced a craniospinal fluid pressure gradient and caused the hindbrain to herniate into the foramen magnum.^{11–13} In addition, patients with craniosynostosis present venous hypertension

because of jugular foramen stenosis, result in intracranial hypertension and/or hydrocephalus.¹⁾ This clinical state of intracranial hypertension and/or hydrocephalus could enlarge the craniospinal fluid pressure gradient furthermore. Spontaneous remodeling of the skull occurs in infancy and is thought to continue until at least twenty years of age. Head CT images for our patient revealed thickening of the skull at the occipital bone and the cranial base.

We carefully analyzed the patient's pathophysiology and determined that the clinical condition was caused primarily by the protrusion of the posterior rim of the foramen magnum. Angiography revealed that the occipital sinuses on both sides were the main efflux route for blood flow from the brain, instead of the usual transverse to the sigmoid sinuses. Therefore, we judged duraplasty to be risky and performed FMD without duraplasty. To prevent cerebellar ptosis due to an overly large craniectomy,¹⁴⁾ we performed an FMD with a diameter of approximately 3 cm.

Surgical strategies should be chosen based on the underlying pathophysiology, including cranial remodeling and venous flow.^{15,16)} Cases of skull deformity or craniosynostosis in infants should be given careful attention and should be followed at least until spontaneous remodeling of the skull has ended.

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Conflicts of Interest Disclosure

We declare that we have no conflicts of interest.

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