

Thalamic Massa Intermedia Duplication in a Dysmorphic 14 month-old Toddler

Matthew T Whitehead^{1*}

1. Department of Neuroradiology, Children's National Medical Center, Washington, D.C., USA

* **Correspondence:** Matthew T. Whitehead, MD, Department of Radiology, 111 Michigan Ave N.W., Washington, D.C. 20010, USA
(✉ MWhitehe@childrensnational.org)

Radiology Case. 2015 Jun; 9(6):1-5 :: DOI: 10.3941/jrcr.v9i6.2297

ABSTRACT

The massa intermedia is an inconstant parenchymal band connecting the medial thalami. It may be thickened in various disease processes such as Chiari II malformation or absent in other disease states. However, the massa intermedia may also be absent in up to 30% of normal human brains. To the best of my knowledge, detailed imaging findings of massa intermedia duplication have only been described in a single case report. An additional case of thalamic massa intermedia duplication discovered on a routine brain MR performed for dysmorphic facial features is reported herein.

CASE REPORT

CASE REPORT

Clinical History

A 14 month-old female was referred for imaging evaluation of the brain because of dysmorphic facial features, developmental delay, and seizures. She is a fraternal twin born at 39-weeks via emergent cesarean section for breech presentation and delayed interval delivery (78 minutes) to a 32 year-old G6P6 mother. Her fraternal twin was born via spontaneous vaginal delivery without incident, and is currently asymptomatic and developmentally normal.

Feeding and development were normal from birth to 3-months of age. Subsequently, developmental milestone achievement was arrested, with some evidence of developmental regression. Additionally, the patient has a history of poor feeding, bouts of laughing/babbling, bruxism, hand wringing, and seizures. Physical examination showed frontal bossing, low set, cupped, posteriorly rotated ears, down-slanting palpebral fissures, and delayed teeth eruption with hypertrophic gums. Hypotonia was present affecting the lower greater than upper extremities. Ophthalmologic exam was normal. A chromosomal microarray was consistent with a normal female.

Imaging Findings

A routine brain MRI was performed on a 1.5T magnet (General Electric, Milwaukee, WI) with and without intravenous gadolinium (1.4 mL Gadopentate Dimeglumine). The following pulse sequences were prescribed: sagittal Fast Spoiled Gradient-Recalled-Echo (FSPGR) T1 weighted-images (WI) reformatted into axial and coronal planes, axial T2 spin echo, axial T2 Fluid Attenuated Inversion Recovery (FLAIR), axial Susceptibility Weighted Angiography (SWAN), coronal fat-saturated T2WI, axial arterial spin labeling, and post contrast axial T1 spin echo and coronal T1 CUBE images reformatted into axial and coronal planes. Short and long TE single voxel MR spectroscopy (MRS) was also performed with voxels of interest placed over the left cerebral deep gray nuclei (relaxation time (TR) 1500, echo time (TE) = 35 and 144).

Sagittal midline T1WI demonstrates two distinct round soft tissue structures interposing the medial thalami within the 3rd ventricle (Figure 1). Incidentally, a small intrasellar pars intermedia and pineal cysts are present (Figure 1). Reformatted axial T1WI confirms continuity of the 3rd ventricular structures across midline, adjoining the medial thalami (Figure 2). The smaller, posterior parenchymal band measures 2 x 2 x 2 mm, while the larger, anterior band measures 3 x 3 x 3 mm.

These structures are isointense to the thalami on all pulse sequences. There is no abnormal contrast enhancement or perfusion abnormality. No additional structural abnormality is present. The thalami and hypothalami are otherwise normal. Short and long TE MRS performed through the left basal ganglia show normal metabolic ratios.

Management

Genetics, neurology, ophthalmology, and gastrointestinal consultations were requested. A surgical gastric tube was placed and Nissen fundoplication performed for weight loss in the context of poor oral intake. Otherwise, management was largely supportive. Seizures were treated with antiepileptic medications. Physical therapy was instituted.

Follow-up

The patient had outpatient follow-ups with neurogenetics, neurology, and ophthalmology. Based on the clinical symptomatology and physical examination findings, MECP-2 related disorders and lysosomal storage disorders were posited in the differential diagnosis. However, ocular examination performed by ophthalmology was reportedly normal, without signs of macular pathology. An abdominal ultrasound was requested to evaluate for hepatosplenomegaly. The patient was also referred for early intervention services with physical, occupational, and speech therapy. However, the patient failed to return for any further follow-up.

DISCUSSION

Etiology & Demographics

The thalamic massa intermedia (MI) is a band of contiguous parenchymal tissue comprised of neurons and axons extending from the medial thalami across the 3rd ventricle [1]. Because the bulk of the axons therein do not connect the thalami to one another, it is not considered a commissure. In the majority of patients, the MI is centrally located between quadrants of the 3rd ventricle [2]. Some believe the MI to be a functionally void vestigial remnant in humans, as it may be absent in up to 20-30% of normal individuals [1,3].

Massa intermedia absence may be congenital or age-dependent; older subjects are more likely to have absent or small MI antero-posterior and craniocaudal lengths compared with younger subjects [3-5]. Its more frequent presence in female brains indicates a sexual dimorphism [3,4]. It may also be absent in certain disease states such as pituitary duplication syndrome [6] and schizophrenia [7]. In other diseases such as Chiari II malformation [8], L1 syndrome with X-linked hydrocephalus [9], and 6q terminal deletion syndrome [10], the MI may be enlarged (greater than 1 cm in diameter). In 2010, Nayak described a case of an enlarged MI found at autopsy in an otherwise normal approximately 70 year-old brain [3].

Only a few reports of duplication of the massa intermedia have been described in the literature. Malobabic and colleagues reported a case of duplication of the massa

intermedia discovered at autopsy [5]. Accessory massa intermedia were identified in 2/20 patients who underwent MR evaluation for Chiari II Malformation [8]. Tubbs et. al. demonstrated the first and, until now, only imaging correlate of massa intermedia duplication in a 14 month-old child who had additional findings of an incomplete Dandy-Walker disease spectrum [11].

Clinical & Imaging findings

This patient has an undiagnosed, presumed genetic aberration. The clinical symptoms including developmental regression, failure to thrive, poor feeding, laughing and bruxism bouts, hand wringing, and seizures in this female child raise suspicion for MECP-2 related disorders such as Rett syndrome. Lysosomal storage disorders are additional differential diagnostic considerations given the frontal bossing, coarse facial features, and gum hypertrophy. However, ophthalmic exam was negative for the "cherry-red macula" which can be seen in storage diseases such as Tay Sachs and Niemann-Pick.

The brain MR reveals an isolated structural abnormality: duplication of the massa intermedia. There are two distinct similar appearing but different sized structures that represent the massa intermedia duplication connecting the medial thalami by traversing the 3rd ventricle horizontally. The duplicated MI is isointense to the thalami on all pulse sequences and demonstrates no contrast enhancement. The brain parenchyma is otherwise normal in volume, morphology, and signal intensity for the patient's age. No focal signal changes are present to support a poliodystrophy or leukodystrophy. The bone marrow is normal in volume and demonstrates age-appropriate signal intensity.

Treatment & Prognosis

Massa intermedia duplication is a congenital abnormality that is not treated. With only a few case reports of MI described in the literature to date, the long-term prognosis for a patient in which it is discovered remains unclear. Although it may be present in some congenital diseases, it has also been previously noted as a presumed incidental variation at autopsy.

Differential Diagnoses

The differential diagnosis for midline transthalamic tissue is limited. A thalamic hamartoma could demonstrate similar imaging characteristics (a nonenhancing, isointense thalamic lesion), however, communication with the contralateral thalamus across the 3rd ventricle would be unusual. Infiltrative neoplastic diseases such as bithalamic gliomas and germ cell tumors would be distinguishable by altered signal intensity with respect to normal thalamus, and some cases, contrast enhancement.

Broadening the lesional location to include paraventricular and intraventricular lesions would introduce subependymal nodular heterotopia, subependymal nodules and giant cell tumors in the setting of tuberous sclerosis, and intraventricular neoplasms into the differential diagnosis. Although a subependymal heterotopian may be essentially isointense to the thalami, it does not communicate with the contralateral thalamus across the 3rd ventricle. Subependymal

nodules, giant cell tumors, and intraventricular neoplasms are of altered signal intensity with respect to the thalamus and often enhance after contrast agent administration.

TEACHING POINT

Duplication of the thalamic massa intermedia is a rare structural abnormality that may occur in patients with additional congenital defects.

REFERENCES

1. Nieuwenhuys R, Voogd J, Van Huijzen C. The Human Central Nervous System, 4th Ed. Berlin, Germany: Springer; 2008:255. ISBN-13: 9783540346845.
2. Samra KA, Cooper IS. Radiology of the Massa Intermedia. Radiology 1968. 91:1124-1128. PMID: 5303380.
3. Nayak BS, KV S. Unusually large interthalamic adhesion and its clinical importance. International Journal of Anatomic Variations 2010. 3:174-175. ISSN: 13084038.
3. Sen F, Ulubay H, Ozeksi P, Sargon MF, Tascioglu AB. Morphometric measurements of the thalamus and interthalamic adhesion by MR imaging. Neuroanatomy 2005. 4: 10-12. ISSN: 13031783.
4. Rosales RK, Lemay MJ, Yakolev PI. The development and involution of massa intermedia with regard to age and sex. J. Neuropat. 1968. 27:166. PMID: 5301516.
5. Malobabic S, Puskas L, Blagotic M. Size and position of the human adhaesio interthalamica. Gegengaur's Morphol. Jahrb. 1987. 133:175-184. PMID: 3569816.
6. Vittore CP, Murray RA, Martin LS. Case 79: pituitary duplication. Radiology 2005. 234(2):411-414. PMID: 15670998
7. Napoulos PC, Rideout D, Crespo-Facorro B, Adreasen NC. Sex differences in the absence of massa intermedia in patients with schizophrenia versus healthy controls. Schizophren. Res. 2001. 48:177-185. PMID: 11295371.
8. el Gammal T, Mark EK, Brooks BS. MR Imaging of Chiari II Malformation. AJR Am J Roentgenol. 1988. 150(1):163-70. PMID: 3257116.
9. Kanemura Y, Okamoto N, Sakamoto H, Shofuda T, Kamiguchi H, Yamasaki M. Molecular mechanisms and neuroimaging criteria for severe L1 syndrome with X-linked hydrocephalus. J Neurosurg. 2006. 105(5 Suppl):403-412. PMID: 17328266.
10. Elia M, Striano P, Fichera M, et al. 6q terminal deletion associated with a distinctive EEG and clinical pattern: a report of 5 cases. Epilepsia 2006. 47(5):830-838. PMID: 16686647.
11. Tubbs RS, Smyth MD, Oakes WJ, Blount JP. Duplication of the massa intermedia in a child. Pediatr Neurosurg 2004. 40(1):42-43. PMID: 150017230.

FIGURES

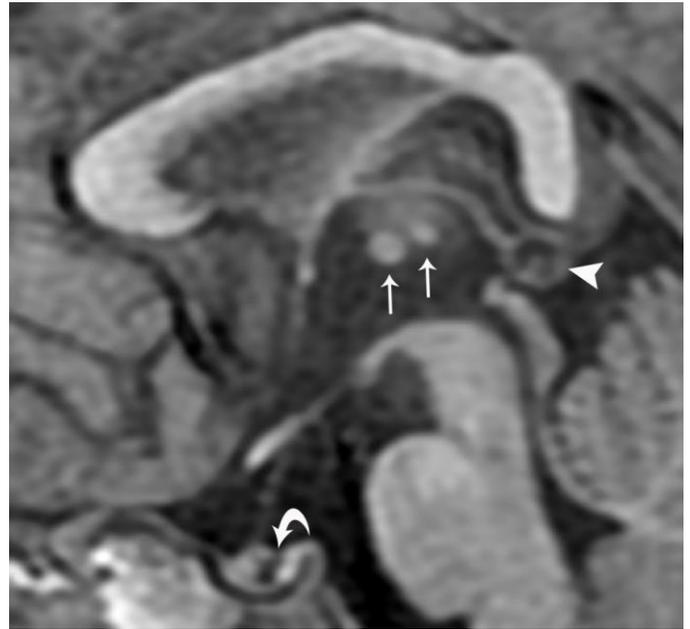


Figure 1: 14-month-old female with duplication of the massa intermedia. FINDINGS: Sagittal T1WI demonstrates two distinct round structures in the 3rd ventricle (straight arrows) representing duplication of the massa intermedia. The anterior massa intermedia measures 3 x 3 x 3 mm while the posterior MI measures 2 x 2 x 2 mm. Small pineal cyst (arrow head) and intrasellar pars intermedia cyst (curved arrow) are incidentally present. TECHNIQUE: 1.5T MR (General Electric, Milwaukee, WI). Sagittal midline T1 Fast Spoiled Gradient Echo (FSPGR) image (TR/TE/IT = 11/2/500).

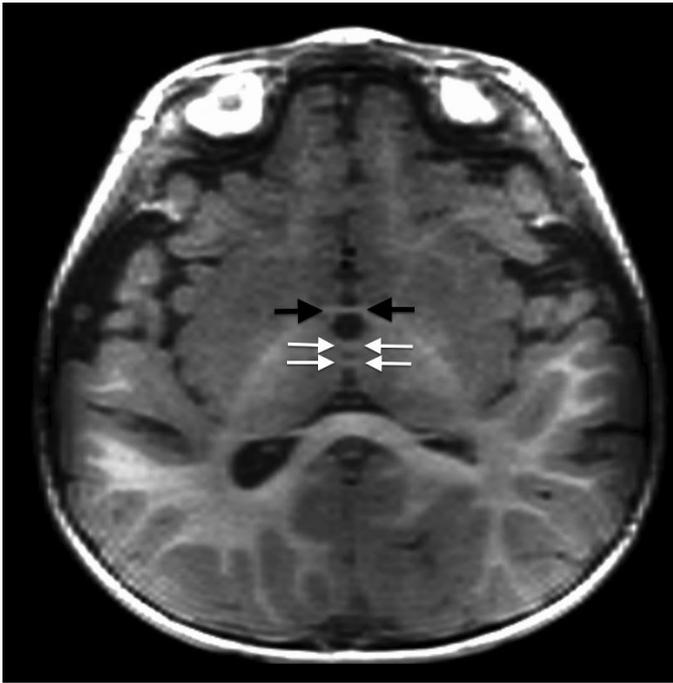


Figure 2 (left): 14-month-old female with duplication of the massa intermedia. FINDINGS: axial T1WI demonstrates 2 distinct horizontally oriented parenchymal bands crossing the 3rd ventricle connecting the thalami consistent with massa intermedia duplication (white arrows). The normal anterior commissure is visible anteriorly (black arrows). TECHNIQUE: 1.5T MR (General Electric, Milwaukee, WI). Fast Spoiled Gradient Echo (FSPGR) image (TR/TE/IT = 11/2/500).

Etiology	Congenital. Unknown underlying defect.
Incidence	Unknown but rare (less than 10 reported cases)
Gender ratio	Unknown. Both case reports with images were female.
Age Predilection	Congenital.
Risk Factors	Unknown.
Treatment	None.
Prognosis	Unclear. Likely related to additional concurrent developmental abnormalities
Findings on Imaging	Two horizontal parenchymal bands connecting the thalami across the 3 rd ventricle. Isointense to thalami on T1WI and T2WI; no contrast enhancement

Table 1: Etiology, incidence, demographics, risk, treatment, prognosis, and imaging factors associated with massa intermedia duplication.

	T1W MR	T2W MR	T1W post con	DWI MR	Location
Duplicated MI	Isointense	Isointense	Isointense	Isointense	Connecting thalami
Hamartoma	Isointense	Isointense	Isointense	Isointense	Thalamus
Glioma	Hypointense	Hyperintense	Variable +/- enhancement	Variable	Thalami
Heterotopia	Isointense	Isointense	Isointense	Isointense	Sub-ependymal
Giant cell tumor	Variable	Variable	Hyperintense + enhancement	Variable	Foramen of Monroe
Intraventricular neoplasm	Variable Hypointense	Variable Hyperintense	Hyperintense + enhancement	Variable	Ventricle

Table 2: Differential diagnosis (1st column) and MR imaging appearance (columns 2-6) for a duplicated massa intermedia. Displayed intensity is relative to the normal thalamic signal intensity. MI = massa intermedia, W = weighted, con = contrast, DWI = diffusion weighted images.

ABBREVIATIONS

FLAIR = Fluid Attenuated Inversion Recovery
 FSPGR = Fast Spoiled Gradient-Recalled-Echo
 MI = massa intermedia
 MRI = magnetic resonance imaging
 MRS = MR spectroscopy
 SWAN = Susceptibility Weighted Angiography
 T = tesla
 TE = echo time
 TR = relaxation time
 WI = weighted-images

KEYWORDS

Massa intermedia duplication; interthalamic adhesion; thalamus; brain MRI; congenital

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/2297

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
 Available online at www.RadiologyCases.com

Published by EduRad



www.EduRad.org