



Case report

Congenital geniculate quadruple sectoranopia with occipital heterotopia

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ABSTRACT

Purpose: To report a case of congenital geniculate quadruple sectoranopia associated with occipital heterotopia. **Observations:** A 51-year-old healthy woman was incidentally found to have a left incongruous quadruple sectoranopia. Analysis of the macular ganglion cell complex (GCC) revealed homonymous hemianopic thinning of the inner layer of the retina. Brain magnetic resonance imaging (MRI) showed congenital occipital heterotopia, characterized by hypertrophy of the right parahippocampal gyrus, lingual gyrus, and isthmus of the cingulate gyrus, with shrinkage of the white matter. In addition, serial coronal images on a short tau inversion recovery (STIR) sequence demonstrated an atrophic right optic tract. **Conclusion and importance:** Congenital geniculate quadruple sectoranopia is extremely rare and may be caused by congenital occipital heterotopia.

1. Introduction

Geniculate hemianopia is characterized by either a wedge-shaped homonymous hemianopia or a quadruple sectoranopia. The former visual field defect is produced by a lesion in the dorsal crest of the lateral geniculate nucleus (LGN) supplied by the lateral posterior choroidal artery, while the latter is produced by lesions of the medial and lateral horns of the LGN supplied by the distal anterior choroidal artery. In most cases the etiology of geniculate hemianopia is acquired, usually through arterial occlusion or hemorrhage.^{1–8} Congenital geniculate hemianopia is extremely rare. We report a case of congenital geniculate quadruple sectoranopia associated with congenital occipital heterotopia.

2. Case report

A 51-year-old healthy woman was referred to the ophthalmology section of our hospital with a chief complaint of myodesopsia in both eyes. On examination, visual acuity was 20/20 in each eye and there was no relative afferent pupillary defect. Intraocular pressure was 14 mm Hg in both eyes. Slit-lamp examinations showed no abnormalities. Fundusoscopic examinations showed large optic cups (Fig. 1A). Using spectral-domain optical coherence tomography (SD-OCT) (Cirrus 4000 HD-OCT; Carl Zeiss Meditec, Dublin, CA), retinal nerve fiber layer (RNFL) analysis showed slight nasal and temporal thinning on the left and

normal thickness on the right (Fig. 1B). By contrast, the ganglion cell complex (GCC), consisting of the ganglion cell layer (GCL) combined with inner plexiform layer (IPL), revealed thinning of the temporal hemimacula in the right eye and nasal hemimacula in the left eye, indicating homonymous hemianopic thinning (Fig. 1C). A 30–2.

Humphrey visual field examination showed left superior and inferior sectorial visual field defects, sparing the horizontal fields. The scotomas were larger in the left eye, consistent with an incongruous quadruple sectoranopia (Fig. 2). The patient had never noticed her visual field defects. Brain MRI revealed nodular hypertrophy of the right parahippocampal and lingual gyri and isthmus of the cingulate gyrus. There was shrinkage of the white matter surrounding the right lateral ventricle trigone toward the posterior horn, signifying occipital heterotopia (Fig. 3A). Additionally, serial coronal images (thickness, 2.5 mm) anterior to the LGN on a STIR sequence revealed an atrophic right compared to left optic tract (Fig. 3B).

3. Discussion

Based on the clinical findings, the quadruple sectoranopia in our case appears to have arisen from a geniculate lesion associated with occipital heterotopia. Congenital heterotopia is a cortical malformation in which neurons do not migrate properly during the early development of the fetal brain, from about the 6th to the 24th week of pregnancy.⁹ In most

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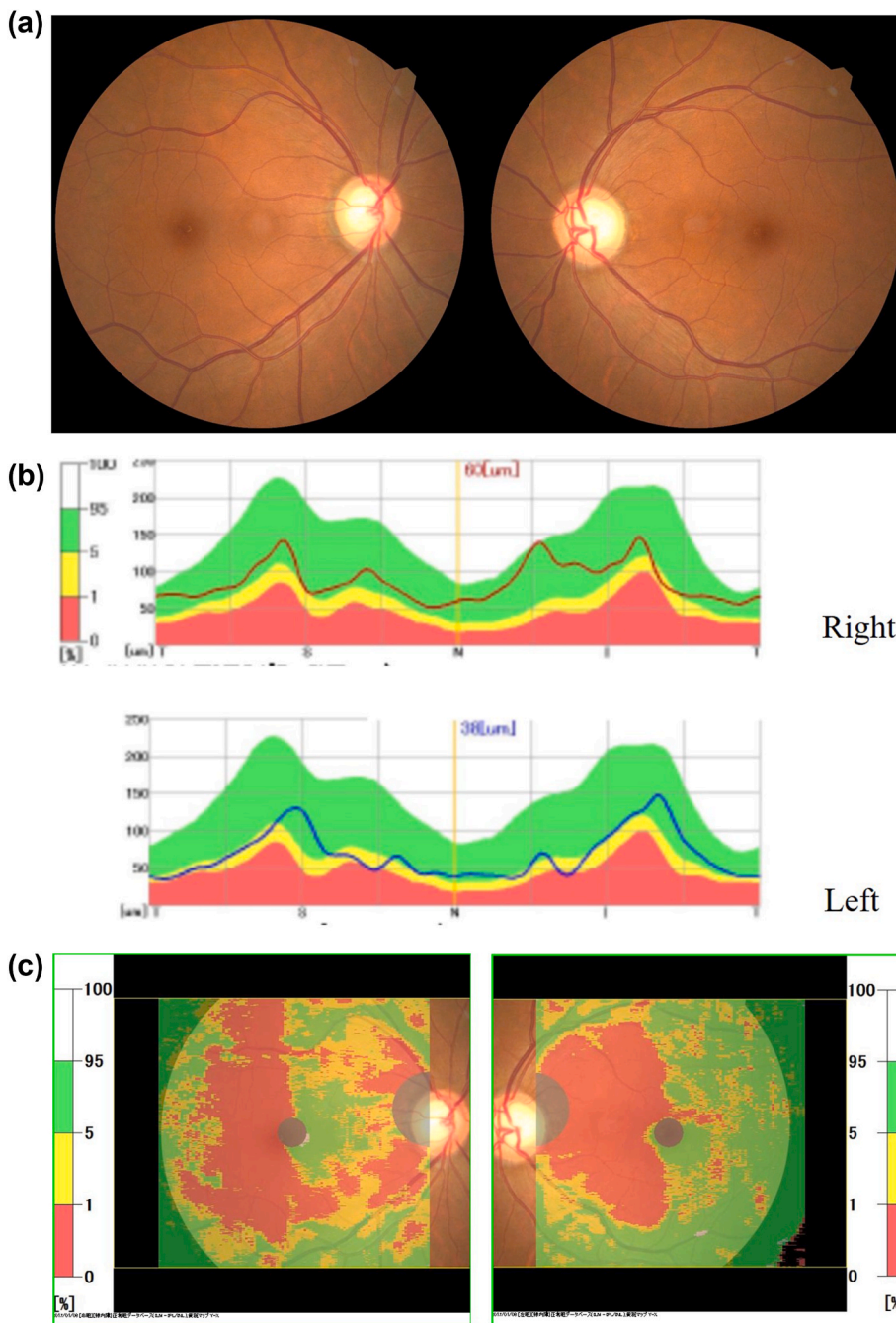


Fig. 1. Fundus photos and SD-OCT images. A) Fundus photographs showed large cups in both optic discs. B) RNFL analysis showed slight nasal and temporal thinning on the left and normal thickness on the right. C) Analysis of the macular GCC revealed the homonymous hemianopic thinning of the inner layers of the retina. RNFL, retinal nerve fiber layer; GCC, ganglion cell complex; SD-OCT, spectral-domain optical coherence tomography; GCL, ganglion cell layer; IPL, inner plexiform layer.

cases, heterotopia usually becomes evident when seizures first appear, often during adolescence. Affected individuals usually have normal intelligence, although some display mild intellectual disability. Our patient was asymptomatic.

There have been numerous previous reports of patients with geniculate quadruple sectoranopia.^{1,3,6-8,10-12} Most cases were caused by acquired vascular diseases, such as cerebral infarction or hemorrhage of the distal part of the anterior choroidal artery.^{1,3,6-8} No case reports appear to have described congenital geniculate quadruple sectoranopia. Hoyt¹⁰ described four patients with incongruous geniculate hemianopia and reported that in one case the patient was totally unaware of any visual field defect. That patient might have had a congenital hemianopia. Moreover, Hoyt highlighted the incongruity of homonymous hemianopia in patients with partial LGN involvement and explained these perimetric findings in terms of the retinotopic laminar anatomy of

the nucleus. The lesion in our patient involved the external portion of the lateral horn of the LGN, which mainly comprises lamina 6 receiving crossed projections from the contralateral lower peripheral retina and a large area of the medial horn of the nucleus receiving projections from the upper hemiretinas of both eyes. The postulated lesion may produce a left incongruous quadruple sectoranopia consisting of a larger visual field defect in the contralateral eye and inferior visual field defect larger than superior one in both eyes (Fig. 4).

Analysis of the macular GCC in our case demonstrated more strikingly homonymous hemianopic thinning of the inner layer of the retina as compared to a less prominent reduction in the thickness of the RNFL on OCT. Meier et al.¹³ reported a patient with acquired transsynaptic retrograde retinal degeneration due to occipital lobe abscess in which GCC of the macula on OCT demonstrated marked homonymous neuronal loss in contrast with slight bilateral temporal loss of the RNFL.

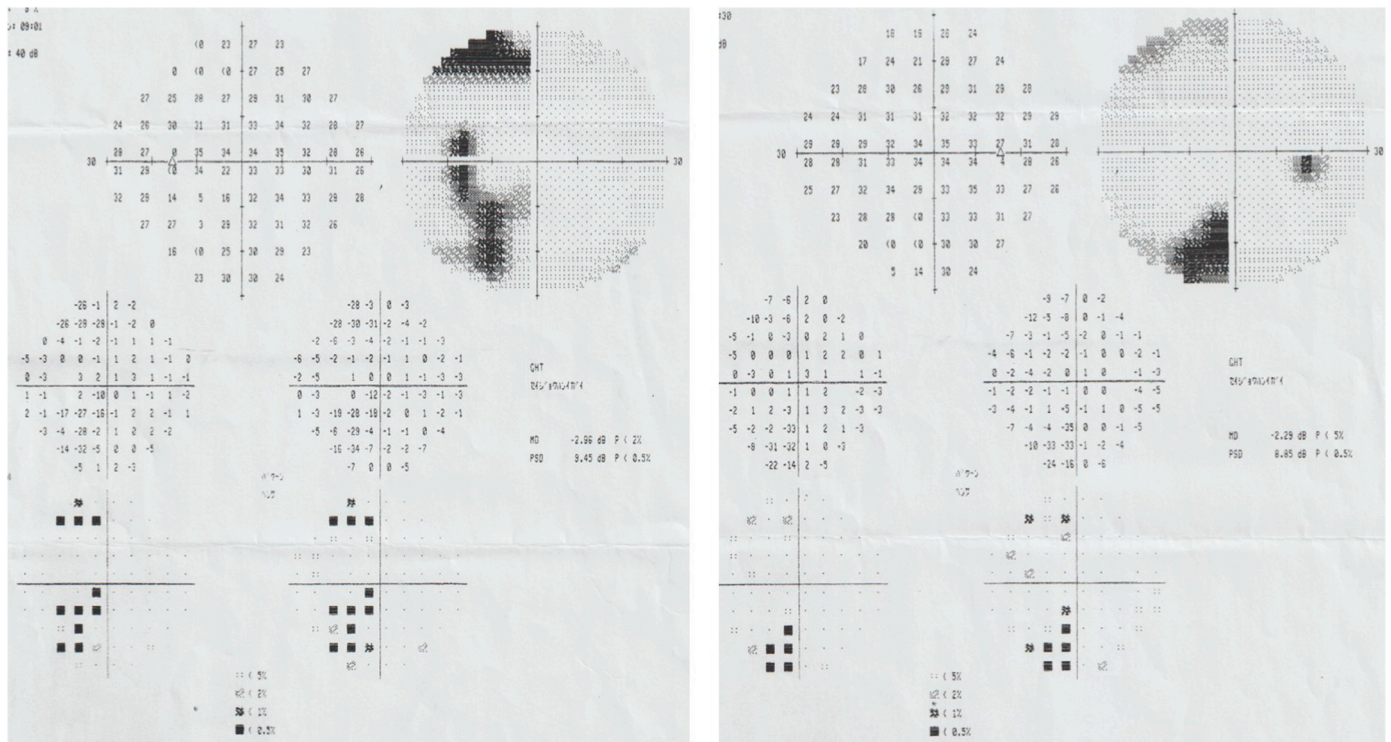


Fig. 2. A 30-2 Humphrey automated visual field examination demonstrating a left incongruous quadruple sectoranopia. The superior and inferior sectorial visual field defects along the vertical meridian spare the horizontal macular zone and consist of larger visual field defect in the left eye.

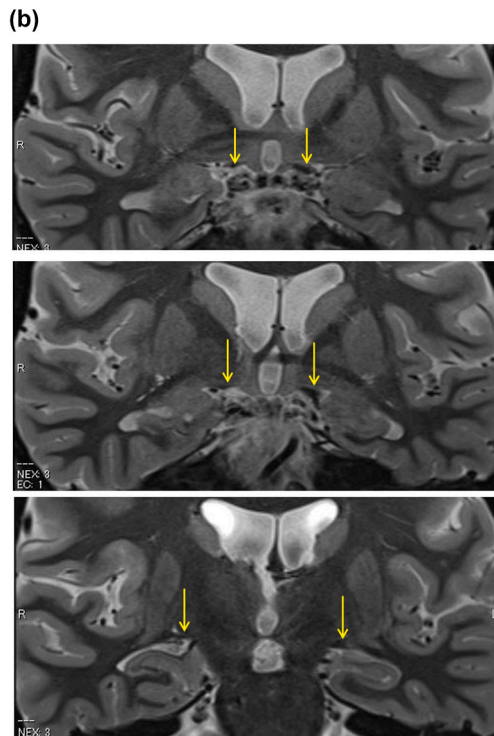
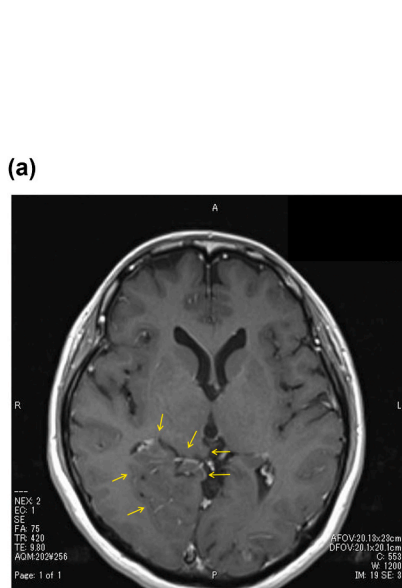


Fig. 3. MRI of the brain.

A) Axial T1-weighted MRI image of the brain demonstrated nodular hypertrophy of the right parahippocampal and lingual gyri and isthmus of the cingulate gyrus (arrows), in contrast with shrinkage of the white matter surrounding the right lateral ventricle trigone toward the posterior horn. There was no enhancement of the lesion after gadolinium injection. B) Serial coronal images (thickness, 2.5mm) on a short tau inversion recovery (STIR) sequence revealed the optic tracts anterior to the LGN (arrows). Note that the right optic tract is thinner than the left optic tract.

Rönnbäck et al.¹⁴ indicated that the analysis of the macular GCC was more sensitive than RNFL thickness for detecting structural loss in a cohort of patients with autosomal-dominant optic atrophy. Ganglion cell atrophy is more localized to the affected halves of the retina, whereas RNFL is contaminated by mixture with healthy fibers from the spared halves of the retina. As a result, differences between macular GCC

analysis and RNFL thickness may occur.

Serial STIR coronal images demonstrated the atrophy of the right optic tract anterior to the LGN. These findings suggest retrograde retinal degeneration. Horton¹⁵ imaged the LGN using MRI, first in brain specimens obtained at autopsy, and subsequently in living humans and determined the optimal imaging plane for precise depiction of the LGN

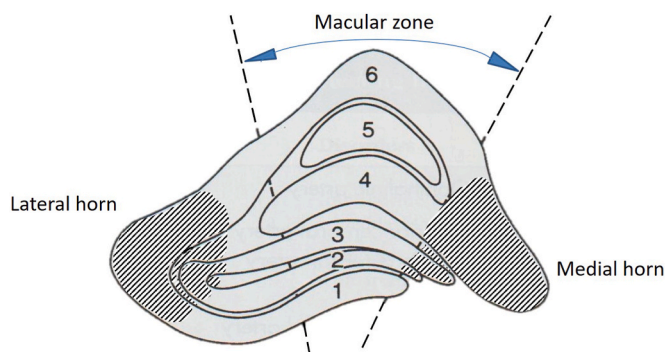


Fig. 4. The diagram depicts the right LGN laminae in mid-coronal section viewed from front. The outer lamina receives the crossed projection from contralateral retina (Laminae 6,4, and 1), whereas the laminae receiving the uncrossed retinal projection occupy the core of the nucleus (Laminae 5,3, and 2). The postulated lesion in our case involves the external portion of the lateral horn, which mainly comprises lamina 6 receiving crossed projections from the contralateral lower peripheral retina and a large area of the medial horn receiving projections from upper hemiretinas of both eyes (shaded area).

on MRI. Rispoli et al.¹⁶ reported a patient with congenital occipital hemianopia due to closed-lip schizencephaly with associated polymicrogyria and showed an abnormality of the retrochiasmatal visual pathway on diffusion tensor imaging. If we had performed MRI for our patient using these special techniques, we might have been able to demonstrate the LGN lesion or atrophic fibers in greater detail.

In conclusion, congenital geniculate hemianopia is extremely rare and may be produced by congenital occipital heterotopia.

Patient consent

Consent to publish the case reports was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors have no financial disclosures.

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