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CASE REPORT

Hypomelanosis of Ito presenting with unilateral dilation of Virchow-Robin spaces: a case report

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Abstract

Hypomelanosis of Ito is a rare heterogeneous neurocutaneous disorder often associated with central nervous and musculoskeletal system involvement. Herein, we report the first case of hypomelanosis of Ito in the literature presenting with unilateral dilation of Virchow-Robin spaces (VRS). A girl aged 16 years old presented with a 1-year history of headache. Her physical and neurological examinations were normal, except for the presence of unilateral cutaneous macular hypopigmented whorls and streaks on lower side of the right trunk and lower limb, termed as Blaschko's lines. She had mild deficits in cognitive and adaptive functioning. Hearing, renal, dental, ophthalmologic, metabolic, and cardiac assessments were normal. Brain magnetic resonance imaging (MRI) showed markedly unilateral hemispheric enlarged VRS without contrast enhancement and diffusion restriction. To the best of our knowledge, our case is the first report describing the unilateral hemispheric enlarged VRS in a patient with hypomelanosis of Ito. Our report suggested that hypomelanosis of Ito may have unilateral dilation of VRS in brain MRI.

Keywords Hypomelanosis of Ito · Virchow-Robin spaces · Magnetic resonance imaging · Unilateral · Blaschko's lines

Introduction

Hypomelanosis of Ito is a rare heterogeneous neurocutaneous disorder often associated with central nervous and musculoskeletal system involvement. Typically, it is a sporadic condition, but there are few reports of autosomal dominant and recessive inheritance [1]. It is a rare disease with an estimated prevalence is approximately 1 case per 7500 births [2]. The main clinical manifestation is skin abnormalities in the form of unilateral or bilateral cutaneous macular hypopigmented whorls, streaks, and patches, termed as Blaschko's lines. Neurological involvement is the most common and serious noncutaneous manifestation and may include the following: intellectual disability, epilepsy, microcephaly, macrocephaly, neurosensory deafness, speech disorder, hypotonia, and ataxia [3, 4]. Other various systemic manifestations include musculoskeletal (scoliosis, chest wall deformity, finger abnormalities, etc.), dental (molar agenesis, tooth malformations, dental dysplasia, etc.), ophthalmologic (retinal hypopigmentation, strabismus, amaurosis, microphthalmia, etc.), cardiac (congenital heart disease, congenital heart block, etc.), and genitourinary anomalies (renal agenesis, ureteral duplication, cystic kidney disease, microphallus, hypospadias, etc.) [4].

The neuroimaging findings are normal in approximately 30–70% of patients with hypomelanosis of Ito. The brain magnetic resonance imaging (MRI) commonly shows diffuse white matter lesions mainly in the parietal and periventricular areas of bilateral hemispheres [5]. Moreover, brain MRI abnormalities include the following: malformations of cortical development, cerebral infarcts, hypoplasia of the corpus callosum, brainstem and cerebellar atrophy, cyst lesions, hemispheric asymmetry, and enlarged Virchow-Robin spaces (VRS) [6].

Herein, we report the first case of hypomelanosis of Ito in the literature presenting with unilateral dilation of VRS.

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Case presentation

A 16-year-old female patient presented to our pediatric neurology department with a 1-year history of headache. There were asymptomatic generalized hypopigmented streaks on the right hemi-trunk and lower limb since birth. She was born at term after an uneventful pregnancy and delivery, with non-consanguineous marriage of her parents. Her early developmental milestones were within normal limits. There was no history of seizures, hearing or visual deficits or dental problems. Family history was unremarkable. Her weight, height, and head circumference had normal percentages. Her physical and neurological examinations were normal, except for the presence of unilateral cutaneous macular hypopigmented whorls and streaks on lower side of the right trunk and lower limb (Fig. 1). On neuropsychiatric evaluation, she had mild deficits in cognitive and adaptive functioning. Laboratory tests including hemogram, kidney, liver, and thyroid function tests were within normal limits. Metabolic tests such as plasma and urine amino acids, and urine organic acids were unremarkable. Hearing, renal, dental, ophthalmologic, and cardiac assessments were normal. Brain MRI showed markedly enlarged subcortical, parietal, temporal, occipital, and periventricular VRS in the right hemisphere without contrast enhancement and diffusion restriction (Fig. 2). Cranial and cervical magnetic resonance angiography were normal. The electroencephalogram demonstrated normal background activity and no epileptic discharge. Based on the clinical manifestations, examinations, and radiological findings, she was diagnosed with hypomelanosis of Ito.

Discussion

Virchow-Robin spaces are perivascular spaces that surround small cerebral arteries and arterioles as they course from the subarachnoid space through the brain tissue [7]. Use of high-resolution MRI has facilitated the visualization of VRS. They occur in typical anatomical areas in one or both hemispheres, can be single or multiloculated, and may provoke non-specific neurological symptoms such as headache and dizziness. Virchow-Robin spaces have been associated with the aging process and occur in various conditions. Lacunar infarcts, cystic periventricular leukomalacia, multiple sclerosis, mucopolysaccharidoses, cystic neoplasms, neurocysticercosis, cryptococcosis, arachnoid cysts, neuroepithelial cysts, and traumatic head injuries have also been reported to be associated with or have similar appearances to VRS on brain MRI [8]. Size (millimetric to several centimeters) and shape (spheric to oval) of VRS on MRI are extremely variable. They have characteristic features on MRI: occurring throughout the path of perforating vessels, isointense to CSF on all sequences, no contrast enhancement, and no diffusion restriction [8, 9].

Virchow-Robin spaces are often detected bilaterally. However, there are also a few cases with unilateral VRS in literature [10–12]. To date, bilateral VRS have been described in several cases with hypomelanosis of Ito. In a pediatric case series, Steiner et al. showed two cases

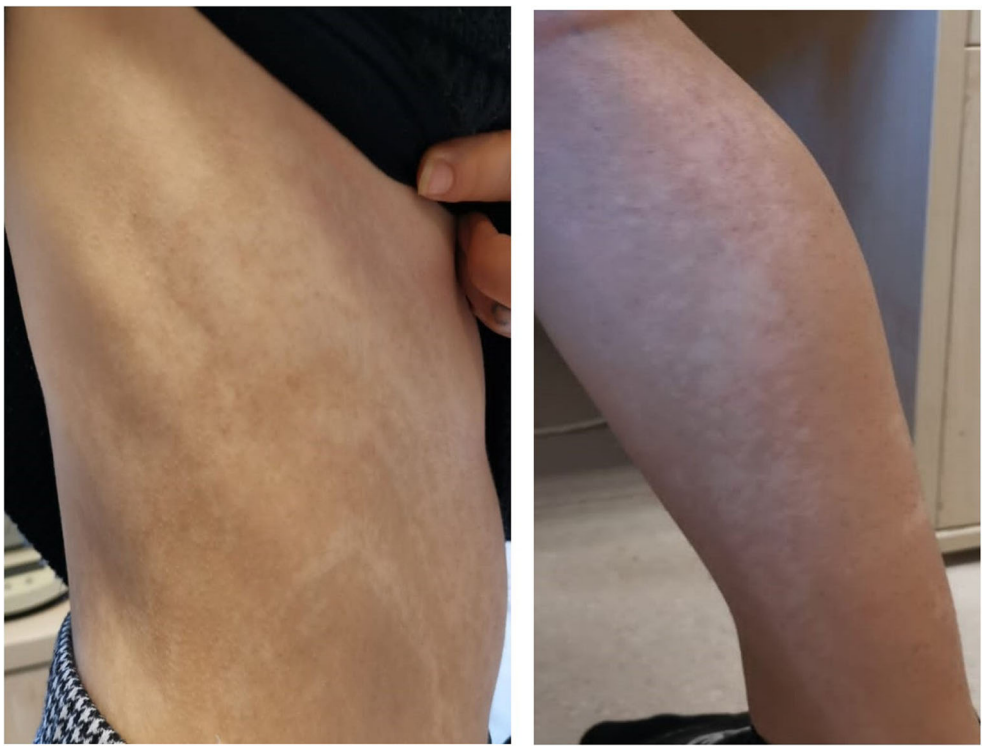

Fig. 1 Unilateral cutaneous macular hypopigmented skin lesions displaying a linear pattern of streaks on the unilateral lower side of the right and lower limb, termed as Blaschko's lines

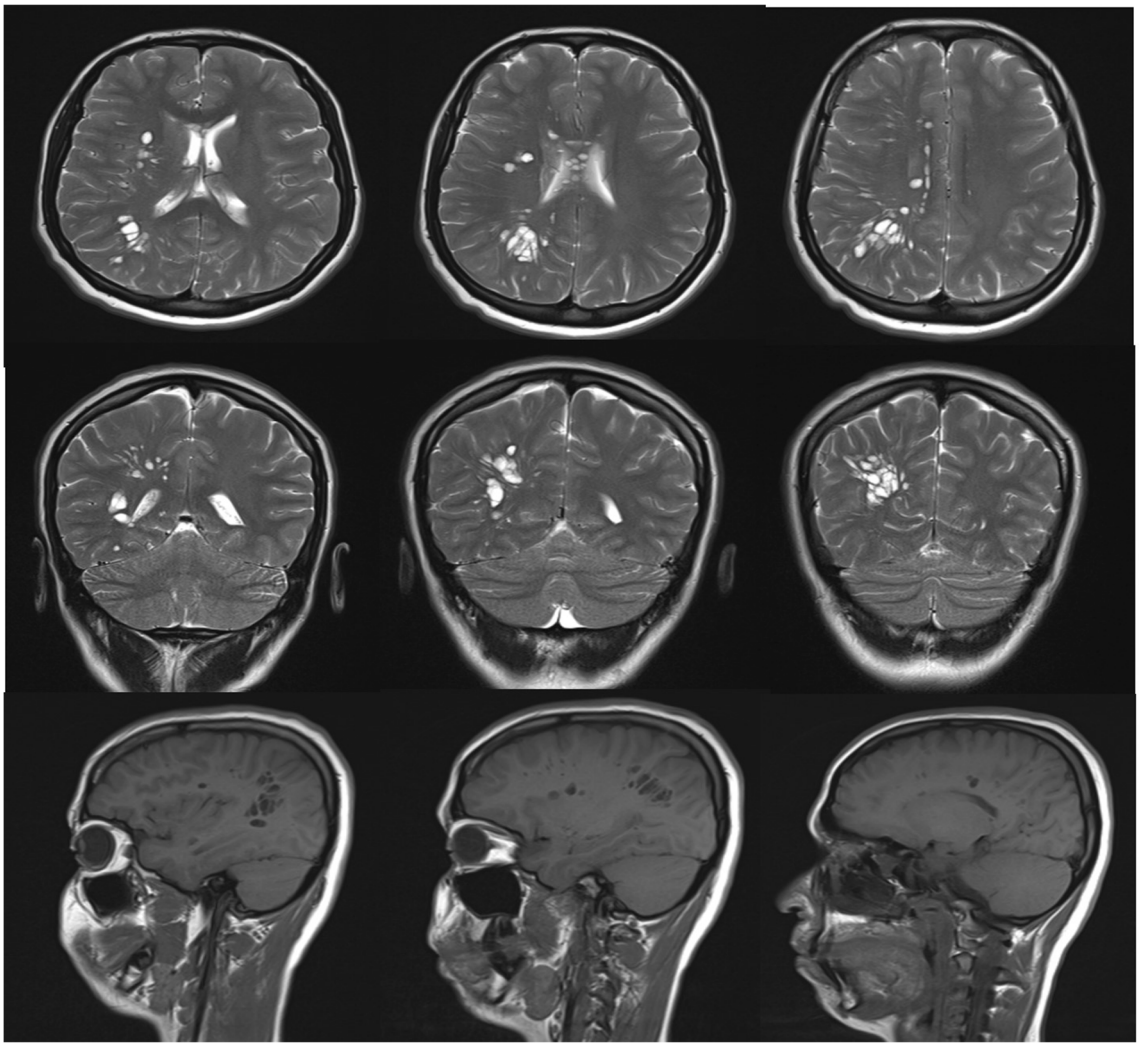

Fig. 2 Enlarged Virchow-Robin spaces. Axial T2-weighted and coronal T2-weighted images show multiple, isointense to cerebrospinal fluid, confluent, oval, well defined, hyperintense (hypointense in sagittal T1-weighted images) lesions in right hemispheric (parietal, temporal, and occipital) subcortical and periventricular white matter

with hypomelanosis of Ito had bilateral VRS [6]. One of the cases had additional cerebellar atrophy and hypoplasia of the corpus callosum, and the other had isolated VRS. Moreover, in another report, Souza et al. described an adult case with hypomelanosis of Ito had bilateral marked enlarged VRS [13]. To the best of our knowledge, our case is the first report describing the unilateral (ipsilateral with the skin lesions) hemispheric enlarged VRS in a patient with hypomelanosis of Ito.

Conclusion

This report expands on the underlying causes of unilateral dilation of VRS and suggests that hypomelanosis of Ito may have unilateral dilation of VRS in brain MRI. If the clinical findings are convenient in cases with unilateral dilation of VRS, hypomelanosis of Ito should also be kept in mind.

Declarations

Conflict of Interest No conflict of interest was declared by the authors.

Informed Consent Written informed consent was obtained from patients' parents.

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