Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Ophthalmology

Congenital Grouped Pigmentation of the Retina

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DOI: 10.36347/sjmcr.2024.v12i06.023 | **Received:** 01.05.2024 | **Accepted:** 05.06.2024 | **Published:** 08.06.2024

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Abstract **Case Report**

Congenital grouped pigmentation of the retina or bear track spots is an uncommon anomaly characterized by multiple, grouped, sharply circumscribed, pigmented spots and no visual dysfunction. We present a case of congenital grouped pigmentation of the retina in a 70-year-old woman. Ophthalmoscopy revealed multiple well-demarcated, flat, brownish-black pigmented lesions in superonasal quadrant in the right eye. We discussed congenital grouped pigmentation of the retina and congenital hypertrophy of retinal pigment epithelium.

Keywords: congenital grouped pigmentation of the retina, congenital hypertrophy of retinal pigment epithelium, bear tracks.

INTRODUCTION

Congenital grouped pigmentation of the retina or bear track spots is an uncommon anomaly characterized by multiple, grouped, sharply circumscribed, pigmented spots and no visual dysfunction [1, 2]. Detection is usually coincidental during routine ocular examination. Congenital hypertrophy of retinal pigment epithelium (CHRPE) is a benign, pigmented, flat lesion arising from the retinal pigment epithelium (RPE) [3, 4]. It is also discovered coincidentally on ocular examination because most patients are without related symptoms.

Herein, we report a patient with congenital grouped pigmentation of the retina.

CASE REPORT

A 70-year-old female presented for routine eye examination. Her personal and family history were unremarkable. On examination, her best corrected visual acuity was 1.2 in both eyes. Ophthalmoscopy revealed multiple well-demarcated, flat, brownish-black pigmented lesions in superonasal quadrant in the right eye (Figure 1). Fundus examination revealed no specific abnormalities in the light eye (Figure 2). We diagnosed our patient with congenital grouped pigmentation of the retina.

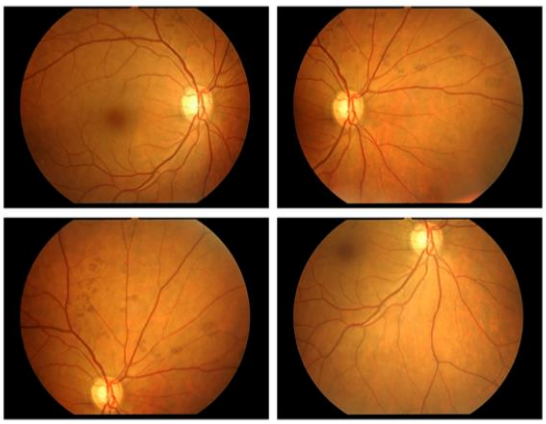

Figure 1. Fundus photographs in the right eye

Note multiple well-demarcated, flat, brownish-black pigmented lesions in superonasal quadrant.

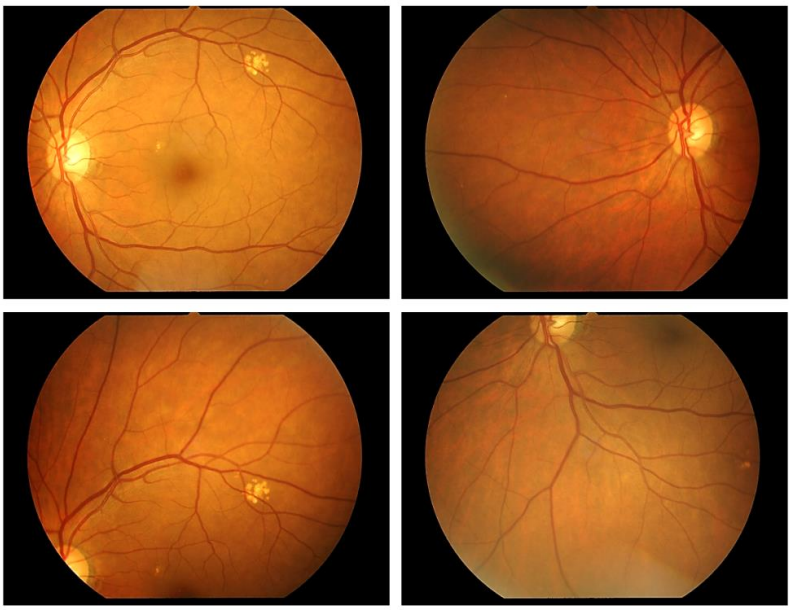

Figure 2. Fundus photographs in the light eye

Note no specific abnormalities.

DISCUSSION

We described a case of congenital grouped pigmentation of the retina in a 70-year-old woman.

The terms "congenital grouped pigmentation of the retina" and "congenital hypertrophy of the retinal pigment epithelium (CHRPE)" are often misinterpreted. CHRPE is a flat, well-circumscribed, benign lesion of the RPE with no malignant potential [5] and is usually solitary. The multifocal variant of CHRPE is known as congenital grouped pigmentation or bear track spots. The typical clinical features of solitary CHRPE and congenital grouped pigmentation have been clearly delineated in the literature. The histopathology of CHRPE has been described as a single layer of hypertrophy of the RPE, with an increase in the concentration of granules within the cells [6]. Shields JA and Tso [7] described a similar histopathological appearance of congenital grouped pigmentation of the retina. Both hypertrophy of the RPE and congenital grouped pigmentation are characterized by flat lesions with variable degrees of pigmentation, undeviated retinal vessels, similar histopathological features, and no evidence of inflammation; therefore, these appear to be different clinical expressions of the same condition, with the former being focal and the latter being multifocal.

Grouped pigmentation of the retina is considered to be sporadic, although familial occurrences have rarely been reported. Marmoy *et al.*, [8] reported a highly unusual case of bilateral, panretinal grouped CHRPE, or bear tracks, in infancy. Turell *et al.*, [9] described a case of grouped pigmented lesions with nonpigmented, punctate lesions located within the macula. Kadoi *et al.*, [2] described congenital grouped pigmentation of the retina in a girl and congenital albinotic spots of the retina in her sister. Renardel de Lavalette *et al.*, [10] reported a familial case of a mother and daughter who showed an autosomal dominant inheritance, while De Jong *et al.*, [11] described a father and son with the disease.

CONCLUSION

In conclusion, although our current findings were based on a single case, we speculated that this lesion remained unchanged for an extended period of time.

Conflicts of interest: The authors have no financial or proprietary interests related to this paper.

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