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Clinical and imaging features of SMACH

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Abstract

Background/Obejectives To describe clinical and multimodal imaging features of patients with stellate multiform amelanotic choroidopathy (SMACH).

Subjects/Methods This retrospective multicenter study included patients with SMACH at Severance Eye Hospital and Seoul National University Bundang Hospital from January 2005 to October 2024.

Results The mean age of SMACH at initial diagnosis was 30.48 ± 22.52 years from 8 eyes of 8 patients. The cohort comprised 3 males and 5 females. Fluorescein angiography (FA) demonstrated a core filled with angiomatous vessels accompanied by mild leakage, irrespective of the presence of subretinal fluid (SRF). Indocyanine green angiography (ICGA) revealed prominent filling defects corresponding to the choroidal lesions, and near-infrared reflectance imaging further showed marked hyperreflectivity along the surface of the dendritic mass lesions, which distinctly highlighted the stellate configuration of the tumors. Three patients (3/8) had SRF. Whether treated or not, the SRF was almost completely absorbed, and visual acuity fully recovered. Most patients with SMACH maintained good visual acuity above 0.1 (logMAR).

Conclusions In patients with SMACH, typical findings in FA and ICGA are presented to facilitate differentiation from other diseases. Understanding the disease entities and favorable prognosis of SMACH should aid clinicians in differentiating these lesions from other tumor diseases.

Keywords SMACH, Choroidal tumor, Ocular oncology

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Introduction

van Dijk EHC and Boon CJF first described "serous maculopathy due to aspecific choroidopathy (SMACH)" in 2021, characterized by subretinal fluid (SRF) accumulation overlying the lesion with unique multimodal imaging features [1]. Since then, fewer than 20 additional cases have been reported, predominantly from Western countries [2]. Ramtohul P, Pellegrini M, Pichi F, et al. later proposed renaming the condition "stellate multiform amelanotic choroidopathy" while retaining the original acronym, SMACH, to emphasize the typical stellate configuration of the lesions and to account for the absence of SRF in some cases [3].

SMACH typically presents as a unilateral orange choroidal lesion underlying nonspecific hypo- and hyperpigmentary changes of the retinal pigment epithelium (RPE). On cross-sectional OCT, the choroidal lesion demonstrates a hyperreflective core within the inner choroid, accompanied by distinctive finger-like hyperreflective projections extending radially in a stellate configuration. ICGA reveals delayed filling and persistent hypofluorescence corresponding to these characteristic projections [3]. Although these imaging features differ markedly from those of other macular choroidopathies, SMACH is often misdiagnosed due to its extreme rarity.

Herein, we report eight SMACH cases with multimodal images, representing the first case series on Asian patients.

Materials and methods

Study population and basic tumor inspection

A retrospective chart review was performed for all patients diagnosed with choroidal osteoma, choroidal mass and choroiddal disorder, other (ICD-10: H31.8 and D31.3) from January 2005 to October 2024, at Severance Eye Hospital, affiliated with Yonsei University College of Medicine, Seoul, Republic of Korea, and National University Bundang Hospital, affiliated with Seoul National University College of Medicine, Seongnam-si, Gyeonggi-do, Republic of Korea. All patients underwent a global ophthalmic evaluation, including best-corrected visual acuity (BCVA) measurement, anterior segment examination, and dilated fundoscopy. All OCT images were obtained using SD-OCT (Spectralis OCT; Heidelberg Engineering GmbH, Heidelberg, Germany). A cube-imaging protocol consisting of 10 scans, five horizontal and five vertical (8× averaged), centered on the fovea and tumor, was performed. Fundus photography (iCare Eidon, Revenio group, Finland), ocular ultrasonography (A-scan and B-scan, Ellex, Adelaide, Australia), SD-OCT, and fluorescein angiography (FA) / indocyanine green angiography (ICGA, Spectralis scanning laser angiography, Heidelberg Engineering GmbH, Heidelberg, Germany) were performed at the initial visit. The width and height diameters were measured in the OCT to define the size of the tumor core beneath the choroid. The greatest linear distance (GLD) including the longest dendrite was measured in infrared reflectance views of OCT or spectralis ICGA.

Statistical analysis

Statistical analyses were performed using SPSS software (version 28.0; SPSS Inc., Chicago, IL, USA). Continuous variables are presented as means ± SD, and categorical variables are presented as numbers (n) and relative frequencies (%). Significance was determined by the Mann–Whitney U test. Statistical significance was set at p-value < 0.05.

Results

Case selection and patient demographics

The baseline characteristics are summarized in Table 1. The mean age at initial diagnosis was 30.5 ± 22.5 years (range, 4.5–65 years), and 5 patients (62.5%) were female. Consecutive follow-up was available for 5 of the 8 patients, with a median duration of 24 months (range, 3–102 months). The referring or suspected disgnoses included atypical osteoma in three patinets, unspecified choroidal mass in three, uveal lymphoma in one, and SMACH in one. Best corrected visual acuity (BCVA) was

Table 1 Demographics and list of the Stellate Multiform Amelanotic Choroidopathy (SMACH) patients

Age (Year)	Eye	Referring or Suspected diagnosis	F/U (mo)	BCVA initial (Snellen)	BCVA_final (Snellen)	Treatment	Tumor core size, Width*Height (μm)	The longest dendritic span, (μm)	SRF height, (μm)	Concurrent finding
4.5	OS	SMACH	1st visit	20/100	N/A	N/A	1055* 100	2038	N/A	Cataract
37	OS	Atypical osteoma	24	20/20	20/25	Observation	1533* 158	3168	N/A	None
24	OS	Atypical osteoma	3	20/25	20/20	Bevacizumab	1132* 113	3172	202	None
55	OD	Atypical osteoma	9	20/20	20/20	Observation	1615* 143	2455	76	None
5.8	OD	Choroidal mass	24	20/25	20/20	Observation	4035* 403	5731	254	None
5.5	OS	Choroidal mass	102	20/25	20/25	Observation	1237* 612	2240	N/A	Acc. ET
65	OS	Choroidal mass	1st visit	20/63	N/A	N/A	4033*478	8600	N/A	Cataract
47	OS	Uveal lymphoma	1st visit	20/25	N/A	N/A	1838*289	2502	N/A	None

F/U: follow up, BCVA: best corrected visual acuity, GLD: Greatest linear distance, SRF: subretinal fluid, Acc. ET: accommodative esotropia

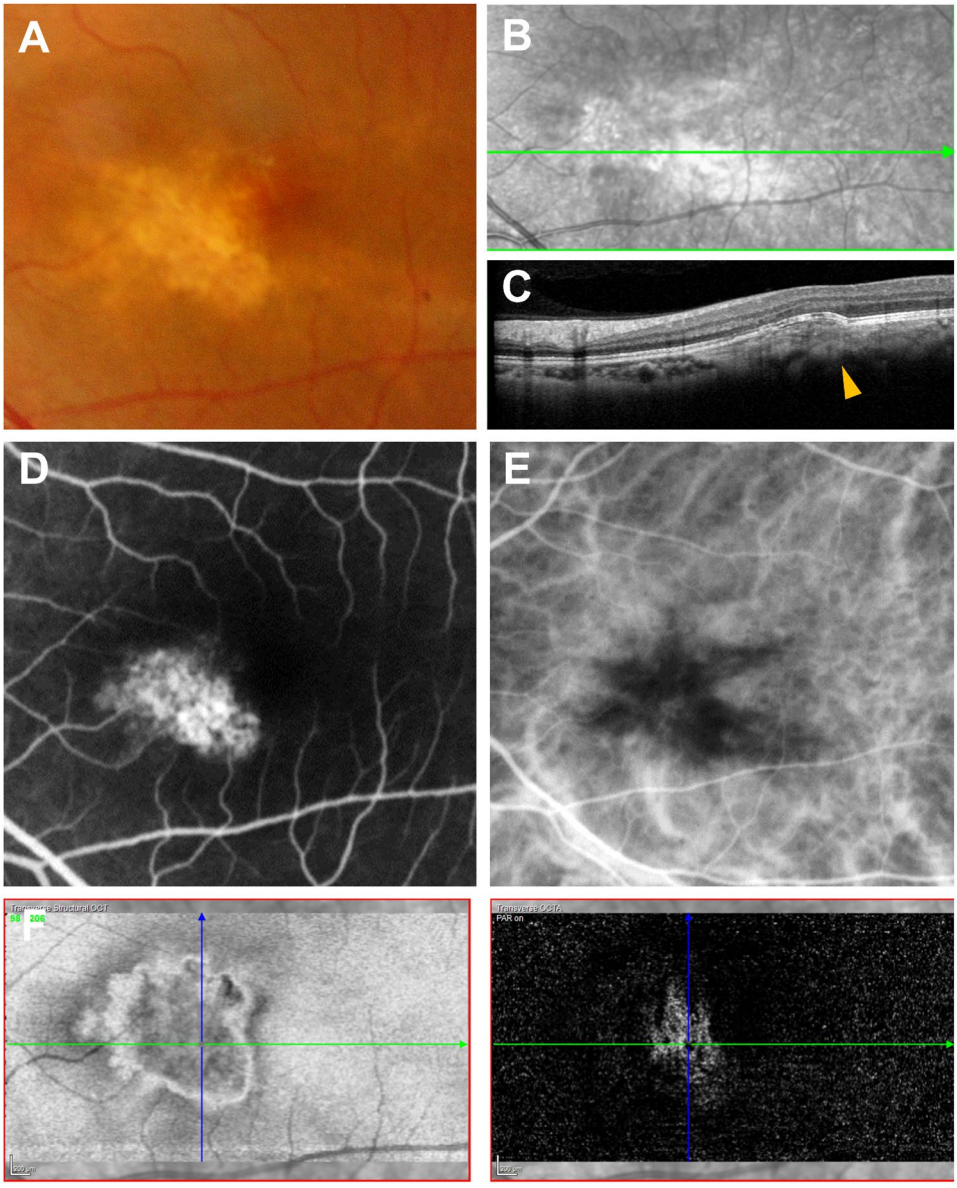

Fig. 1 Representative multimodal images of the SMACH. **A**. fundus photography shows a parafoveal whitish mass lesion. **B** and **C**. near IR and OCT show the solid mass lesion. **D**. Fluorescence angiography shows a hyper-fluorescence lesion of the tumor core. **E**. The Indocyanine green angiography shows the prominent star-shaped choroidal filling defect. **F**. The OCTA showed the tumor margins and the core vasculature

≥20/25 in 6 of 8 patients (75%); decreased vision in the remaining 2 patients was primarily attributable to congenital and senile cataracts, respectively. No significant changes in visual acuity were observed during follow-up (from 0.06 to 0.04 (logMAR), *p*-value:0.58).

Subretinal fluid (SRF) was detected in 3 of 8 patients (37.5%), with a mean height of 177.3 ± 74.7 μm. SRF resolved spontaneously in two cases; in one of these, recurrence was observed two years later but again resolved spontaneously. In another case, SRF resolved one month after a single intravitreal bevacizumab injection (1.25 mg/0.05 mL) and remained absent for the following 2 months. Choroidal neovascularization (CNV) was not detected in any patient.

All cases demonstrated macular involvement, characterized by whitish, stellate lesions on fundus photography (Fig. 1A). OCT revealed a small solid mass in the superficial choroid or Sattler's layer, accompanied by hyperreflective signals (Fig. 1B and C). The mean mass core size was 2059.8 ± 1165.5 μm (range, 1055.0–4035.0 μm) by 287.0 ± 179.7 μm (range, 100.0–612.0 μm), and the longest dendritic span measured 3738.3 ± 2137.9 μm. FA and ICGA were performed in 5 of 8 patients (62.5%). FA typically revealed angiomatous vasculature at the mass core with hyperfluorecence (Fig. 1D), while ICGA

prominently showed a finger-like projections filling defect in the choroid (Fig. 1E). OCTA demonstrated hyperreflective signals at the mass core (Fig. 1F), and B-scan ultrasonography showed the mass as hypoechoic compared with the sclera.

Discussion

The clinical demographics of patients in the present study were not markedly different from those of the 18 patients previously reported in the literature, showing a comparable mean age (30.5 years vs. 28 years in previous studies), a relatively good mean visual acuity of approximately 20/25, and no definite sex predilection. Decreased vision in some of our patients was attributed to other causes, such as cataract. A notable feature of our study was the inclusion of three younger patients aged 4.5, 5.5, and 5.8 years, whereas the minimum age in the previous study was above 5 years including recent report [4].

SMACH was frequently misdiagnosed as atypical choroidal osteoma, atypical central serous chorioretinopathy (CSCR), or unspecified maculopathy (Table 1). Although SMACH demonstrates relatively distinct phenotypic features, characterized by stellate lesions with flecked and honeycomb-shaped cores on FP, it can still be misdiagnosed as choroidal osteoma or other mimicking entities when evaluation is limited to conventional imaging tools restricted to FP and OCT. Therefore, multimodal imaging, including FA, ICGA, B-scan ultrasonography, and OCTA, is recommended for the differential diagnosis of SMACH and other choroidal diseases. A finger-like projection shaped choroidal filling defect observed in ICGA, and a bundle of mild hyperfluorescence within the lesion core on FA are typical characteristics of SMACH (Fig. 1). Additionally, whereas the choroidal osteoma typically presents as a hyperechoic lesion with posterior shadowing on B-scan, SMACH tends to exhibit a hyporeflective signal on B-scan and hyperreflectivity within the core lesion on OCTA (Fig. S1). In patients with SRF, the central portion of the mass may not be clearly visualized, which can lead to confusion with other choroidal maculopathies such as CSCR. In such cases, careful evaluation of the finger-like projections is helpful. On near IR imaging, choroidal wrinkles were observed in some patients ($n = 3/3$ in this cohort), and on indocyanine green angiography, filling defects of finger-like projections are clearly visible in areas without SRF. These features may aid in differential diagnosis.

Subretinal fluid was noted in 37.5% of patients at initial presentation. SRF either resolved spontaneusly and recurred or completely resolved after a single session of intravitreal anti-VEGF in one case (case #3) (Fig. S2). Whether anti-VEGF therapy had a role in this case remains uncertain, since other patients demonstrated spontaneous resolution without treatment, and multimodal imaging did not reveal definite signs of choroidal neovascularization or significant vascular leakage in any case. The source of SRF in SMACH remains undermined. It may result from direct leakage related to vascular malformations within the lesion or secondary effects of RPE dysfunction overlying the lesion. The spontaneous occurrence and resolution may of SRF may support the latter hypothesis. Regardless of the presence of SRF or treatment, all cases showed relatively good visual outcomes.

The exact etiology of the whitish mass lesion in SMACH has not yet been elucidated [1]. The most clinically similar choroidal lesion appears to be choroidal osteoma, which may share certain developmental mechanisms. However, the distinct multimodal imaging features described above, relative stability of lesion size over time (Fig. S3), and the strong female predilection observed in choroidal osteoma distinguish it clinically from SMACH. Considering the early onset and the non-progressive nature of the focal lesion, we hypothesize that SMACH may result from a congenital abnormality of vascular progenitor cells. During development, these progenitor cells, originating from large choroidal vessels, may differentiate into tertiary branches of the choroidal vasculature, forming the typical net-like patterns observed in SMACH [5, 6].

This study was limited by the small number of patients; however, it represents the second-largest case series to date. Because these patients were collected over a relatively short period from only two tertiary centers, and most were diagnosed recently following the recognition of SMACH in literature, this entity may not be as rare as previously believed. Further investigation is warranted to determine whether SMACH is more prevalent among Asian patients. Other limitations include variable and short follow-up periods, which precluded the evaluation of treatment efficacy and lesion size progression. Nevertheless, our study expands the demographic scope to different racial groups and provides valuable insight into this newly recognized choroidal entity, SMACH.

Abbreviations

SMACH	Stellate Multiform Amelanotic Choroidopathy
BCVA	Best-corrected visual acuity
OCT	Optical coherence tomography
SRF	Subretinal fluid
FA/ICGA	Fluorescein and indocyanine angiography

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12886-026-04810-z.

Supplementary Material 1

Supplementary Material 2

Supplementary Material 3

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Author contributions
JJ was involved in collecting clinical data, the literature search, and the preparation of the manuscript. JJK requested cases #1 to us and helped write the manuscript's discussion section. HJS helped to collect clinical data about choroidal osteoma. JK provided valuable discussions about the pathophysiology and medical illustration of the choroid. SSK and SHB advised and supervised the interpretation of clinical data. SJW requested case #6 to us and advised and supervised the interpretation of clinical data. CSL and SJW helped manage the patient, editing, and review of the manuscript, are the guarantor. All authors have read and approved the manuscript.

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Data availability
The datasets analyzed during the current study are not publicly available due to rarity of the disease but are available from the corresponding author on reasonable request.

Declarations

Ethical approval
The study protocol was approved by the Institutional Review Board of Severance Hospital (No. 4-2024-0084), and the study was conducted following the principles of the Declaration of Helsinki.

Consent for publication
Not applicable.

Consent to participate
Informed consent was obtained from each patient after a detailed explanation of the nature and possible consequences of the study procedures. Individuals younger than the age of 16, consent to participate were obtained from their parents or legal guardians.

Meeting presentation
Not applicable.

Competing interests
The authors declare no competing interests.

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