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Periorbital Purpura: A Key Finding in Identifying Hematological Disorders

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ABSTRACT

Periorbital purpura, also known as “raccoon eyes,” is the key finding in identifying hematological disorders such as amyloid light chain amyloidosis, multiple myeloma, or monoclonal gammopathy of undetermined significance.

1 | Introduction

Periorbital purpura, also known as “raccoon eyes,” is the cutaneous manifestation of periorbital ecchymosis caused by increased vascular fragility due to vascular infiltration of amyloid fibrils and is usually associated with amyloid light chain (AL) amyloidosis [1]. However, the relationship between periorbital purpura and other hematological disorders is still poorly understood.

2 | Case History

A 72-year-old man with a history of heart failure, hypertension, and dyslipidemia presented with a two-week history of periorbital purpura. He had never had any hematological problems or trauma. Physical examination revealed purpura around both eyes (Figure 1). Laboratory findings showed elevated levels of serum creatinine (1.21 mg/dL), N-terminal pro-brain natriuretic peptide (560 pg/mL), and immunoglobulin G (IgG) (1939 mg/dL), but no anemia, hypercalcemia, troponin elevation, proteinuria, or coagulation abnormalities. In addition, IgG- κ type monoclonal antibody (1530 mg/L; free light chain κ/λ ratio of 71.5) in protein electrophoresis and Bence Jones protein were detected. Radiologic examinations showed no skeletal lytic lesions. He underwent a bone marrow biopsy, which revealed 9.6% of monoclonal plasma cells. Congo red staining of bone marrow

showed no evidence of amyloidosis. Furthermore, because he had a history of heart failure, cardiac investigations were also performed. Transthoracic echocardiography showed a left ventricular ejection fraction of 61% without hypertrophy (interventricular septal thickness of 8 mm and posterior left ventricular wall thickness of 9 mm) or significant valvular abnormalities. Enhanced cardiac magnetic resonance imaging showed no late gadolinium enhancement. The native T1 values were 1250 to 1300 ms throughout the myocardium, which were not as high.



FIGURE 1 | Physical examination showing periorbital ecchymosis (raccoon eyes).

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Myocardial biopsy showed no amyloid deposition. Therefore, he was finally diagnosed with monoclonal gammopathy of undetermined significance (MGUS) and will be followed by a hematologist.

3 | Discussion

MGUS is an asymptomatic premalignant condition characterized by a serum monoclonal immunoglobulin concentration of less than 3g/dL and a bone marrow plasma cell proportion of under 10%. It is diagnosed in the absence of lytic bone lesions, anemia, hypercalcemia, or renal dysfunction attributable to the proliferation of monoclonal plasma cells. MGUS is also a known precursor of multiple myeloma or AL amyloidosis [2]. Although blood vessels throughout the body are affected, purpura is more likely to occur in areas of thin skin where the vessels are particularly fragile; therefore, the periorbital area is particularly susceptible [3]. Some cases of raccoon eyes have been reported to be associated with multiple myeloma or prior to the diagnosis of AL amyloidosis [4]. Periorbital purpura in MGUS has not been reported, but even a few monoclonal plasma cells may affect vulnerable vessels. To the best of our knowledge, this is the first reported case of raccoon eyes associated with MGUS. Periorbital purpura is the key finding in identifying hematological disorders such as AL amyloidosis, multiple myeloma, or MGUS.

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Author Contributions

Ryohei Ono: conceptualization, formal analysis, investigation, writing – original draft. **Togo Iwahana:** formal analysis, investigation, writing – review and editing. **Kaoruko Aoki:** formal analysis, investigation, writing – review and editing. **Hirotoishi Kato:** formal analysis, investigation, writing – review and editing. **Yoshio Kobayashi:** supervision, writing – review and editing.

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The authors have nothing to report.

Consent

Written patient consent has been signed and collected.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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