

'Spontaneous' Periocular Hemorrhage and Macrohematuria

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Key Words

Rhabdomyosarcoma • Bone marrow infiltration • Periocular hemorrhage • Macrohematuria

Abstract

Purpose: To report on a 19-year-old man with 'spontaneous' periocular hemorrhage and macrohematuria as the first signs of a bone-marrow-infiltrating rhabdomyosarcoma of the left superior rectus muscle. **Procedures:** Clinical eye examination, magnetic resonance imaging, laboratory workup and bone marrow biopsy were performed. **Results:** Magnetic resonance imaging showed a mass lesion in the left superior rectus muscle. While funduscopy had initially been normal, optic disc swelling, retinal hemorrhage and vitreous cell infiltration could be seen in the further course of disease. Laboratory work-up showed macrohematuria, thrombocytopenia and decreased concentration levels of plasma fibrinogen. Thromboplastin time was decreased. A bone marrow biopsy revealed small cell infiltration; using immunohistochemistry, a rhabdomyosarcoma was diagnosed. Unfortunately, the patient died few months later, in spite of chemotherapy. **Conclusion:** 'Spontaneous' periocular hemorrhage and macrohematuria were the first signs of a bone-marrow-infiltrating rhabdomyosarcoma in this young and otherwise healthy patient.

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Introduction

Rhabdomyosarcoma (RMS) was found to be the most frequent malignant orbital neoplasm in young patients, representing about 3% of all orbital masses in children [1]. Bone marrow infiltration by RMS is not unusual, and can lead to different types of bone marrow failure, mimicking other hematological disorders such as leukemia [2]. Even in cases without manifest bone marrow failure, infiltrations by RMS were found in 7 cases out of 32 in a prospective study [3]. Therefore, bone marrow assessment at the moment of RMS diagnosis is recommended by some authors in order to ameliorate the prognosis [4]. The orbit as the primary localization of RMS is not common, but has been reported [5]. Although RMS is a highly malignant neoplasm, patients treated with combined chemo- and radiotherapy showed a survival rate of 80% over 7 years in a small series [6].

To the authors' knowledge, this is the first report on a primary orbital RMS combined with bone marrow infiltration causing thrombocytopenia.

Unfortunately, the patient died a few years before submission of this paper. As a result, there was no opportunity to get signed approval for image publication.

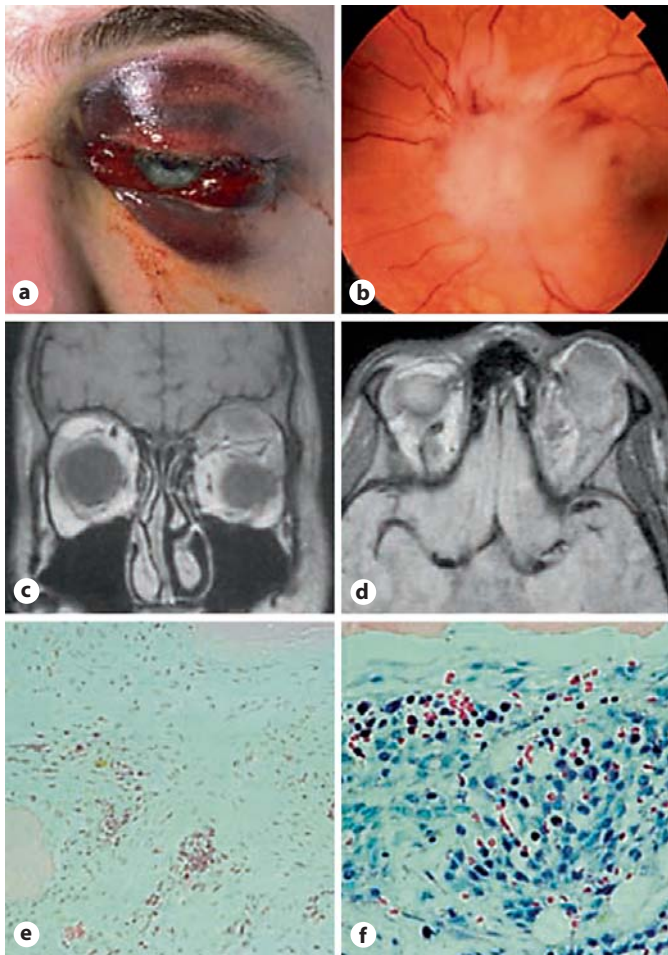
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Case Report

A 19-year-old man presented with acute proptosis and periocular hemorrhage in the left eye after experiencing double vision for 2 weeks (fig. 1a). The patient was otherwise in good health. Magnetic resonance imaging showed a mass lesion in the superior part of the left orbit (fig. 1c, d). The laboratory workup was remarkable for macrohematuria, thrombocytopenia (70,000/mm³), decreased fibrinogen plasma concentration (0.8 μmol/l) and decreased thromboplastin time (51%). Plasma thrombin time and partial thrombin time as well as the hemoglobin concentration and white cell count were within the normal range. Measurements of liver transaminase concentration, α₁-fetoprotein and carcinoembryonic antigen were also normal. A bone marrow biopsy was performed, revealing diffuse small cell infiltration (fig. 1e). The following immunohistochemical workup met the criteria for an RMS (fig. 1f). Visual function as well as fundus examination was normal for both eyes.

Several weeks later the visual acuity in the left eye decreased and funduscopy demonstrated optic disc swelling and vitreous cell infiltration (fig. 1b). The patient died within 5 months, in spite of chemotherapy.

Discussion

In this case, the key to diagnosis was the early bone marrow assessment, which allowed the diagnosis of RMS. The performance of a primary biopsy of the orbital mass would have been an option, but it was considered to be dangerous for a patient with thrombocytopenia, as optic nerve compression, following bleeding into the orbit, is a worrying potential complication of this procedure.

The combination of rare or unusual symptoms, such as nontraumatic periocular hemorrhage and macrohematuria in a young patient, should alert the physician to the possibility of an unusual diagnosis, and prompt him to conduct further, even invasive, examinations. A highly interdisciplinary procedure is mandatory.

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