MAK HILL Publications Research Journal of Medical Sciences

Research Article
doi: 10.59218/makrjms.2024.2.18.559.564

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Key Words
Raccoon eye, acute myeloid leukaemia, fungal sinusitis, amyloidosis, haemophilia, hypofibrinogenemia

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Received: 24 May 2024
Accepted: 18 June 2024
Published: 23 June 2024

Citation: Chirasree Sanyal, Kaustav Ghosh and Prakas Kumar Mandal, 2024. Raccoon Eye Sign, Spectrum of Manifestations of Diverse Haematological Diseases. Res. J. Med. Sci., 18: 559-564, doi: 10.59218/makrjms.2024.2.18.559.564

Raccoon Eye Sign, Spectrum of Manifestations of Diverse Haematological Diseases

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ABSTRACT
'The term "raccoon eyes" was first used in a surgical context to describe a clinical association between a skull fracture and trauma. It obviously indicates serious concern. Following this, a skillful understanding of the clinical profile started to emerge and be described in a number of specialty medical fields. In this regard, benign and malignant hematological diseases predominated in describing this entity. Understanding the hidden agenda underlying the classical description's expression and the associated responsibility is crucial when it comes to this matter. Five cases two each of acute myeloid leukemia, primary amyloidosis, severe hemophilia A and congenital hypofibrinogenemia all had the same clinical sign.

INTRODUCTION

The term Raccoon eyes" commonly used in the medical field to describe a distinctive discoloration or bruising around the eyes that resembles the dark markings seen on a raccoon's face. This condition is also known as periorbital ecchymosis. The dark circles or bruising typically appear as a result of blood pooling in the tissues around the eyes.

Periorbital ecchymosis, peri orbital hematoma and panda sign are other names for the raccoon eye sign. The "Raccoon eye Sign" is easily identifiable and it is associated with a significantly higher risk of surgery in cases involving basal skull fractures[1]. Nevertheless, patients with "raccoon eyes" may also develop from a variety of other nonsurgical disorders, including neuroblastoma, multiple myeloma, amyloidosis and Kaposi's sarcoma[2].

Clinical cases: Case 1 and 2, Acute Myeloid Leukaemia.

Case 1: A 28 year old female known case of Acute Myeloid Leukaemia (AML) The FLT3 ITD positive intermediate risk group underwent a "3+7" AML induction. The patient experienced periorbital ecchymosis, or Raccoon eyes, on day nine of treatment, along with febrile neutropenia (Fig. 1a). Her initial values for Hb were 6 g dL^{-1}, TLC (18 700) and platelet count (20,000). Following induction therapy, the platelet count remained consistently low (less than 10,000). Orbital bleeding without mucosal thickening is seen on a computed tomography (CT) scan orbit (Fig. 1b). Provided with supportive care, such as multiple units of transfusions using only platelets from random donors and packed red blood cells. It gradually vanished. Now the patient is doing well, her post induction marrow is in remission.

Case 2: A 43 year old female AML cases that were recently diagnosed fell into the intermediate risk category (normal karyotype) and they were treated with 3+7 induction therapy. The patient experienced swelling resembling raccoon eyes and febrile neutropenia on day 15 of treatment (Fig. 2a). A CT scan of the PNS and orbit showed mild mucosal thickening of the bilateral frontal and ethmoid sinuses along with polypoid mucosal thickening of the right maxillary sinus (Fig. 2b). Injection Amphotericin B was administered to the patient in addition to supportive care. The patient's symptoms have now completely resolved and they are in remission.

Case 3: About 53 years old female presented with progressively worsening dyspnea and bilateral leg swelling for the previous six months. Bilateral pedal oedema, Raccoon eyes (Fig. 3) and moderate pallor were found during the general examination. Upon

Fig. 1(a-b): (a) Raccoon eye at presentation and (b) CT PNS suggestive of Fungal sinus infection

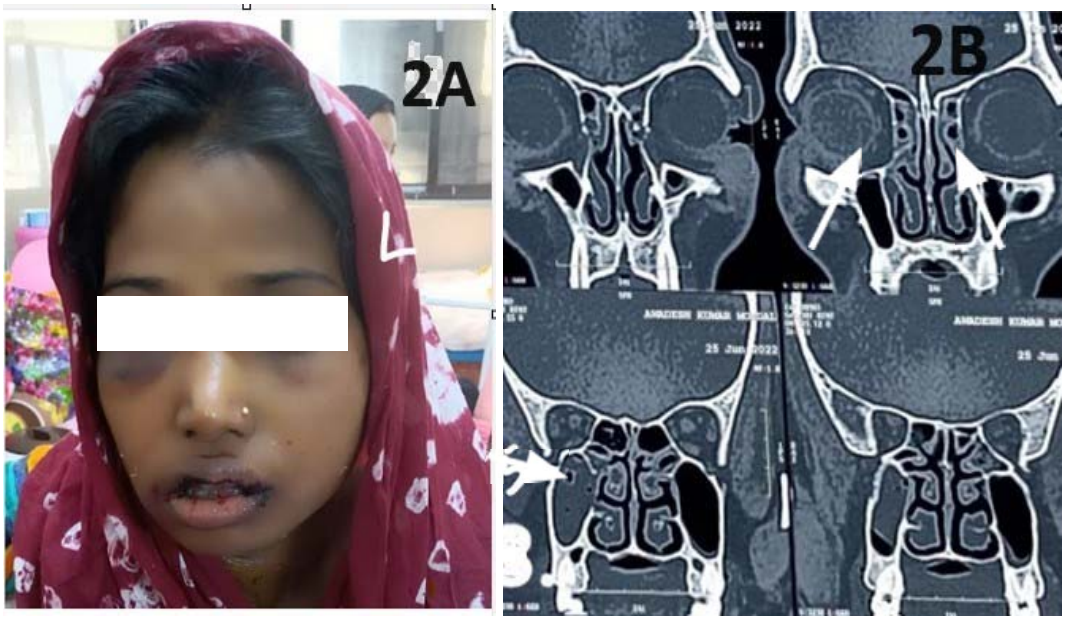

Fig. 2(a-b): (a) Raccoon eye at presentation and (b) CT PNS showing bilateral hypodense mucosal thickening and HALO sign suggestive of fungal sinusitis

Fig. 3: Raccoon eye bilateral dense purple periorbital ecchymosis

systemic examination, a 3 cm hepatomegaly and a short systolic murmur were found in the mitral area. The CBC revealed 1.4 lakh CU^{-1} mm of platelets, TLC 4500 CU^{-1} mm (N62, L30, M6, E2) and anemia (Hb 7.5 gm dL^{-1}). Urea was 34 mg dL^{-1} and creatinine was 1 mg dL^{-1}. A liver function test revealed 4 g dL^{-1} of total protein and 2.3 g dL^{-1} of albumin, or hypoalbuminemia.

The levels of calcium, potassium and sodium in the serum were within normal ranges. On mixing studies, the coagulation profile revealed prolonged APTT 62 seconds that were not corrected. A chest×ray revealed cardiomegaly and an echocardiogram showed a 38% ejection fraction and sparkling myocardial wall motion. After an abdominal fat pad biopsy suspected of having amyloidosis, amyloid protein aggregates stained positively with Congo red. Negative serum protein electrophoresis was obtained. NT Pro BNP was 3200 ng L^{-1} and troponin I was 2 mcg L^{-1}. As a result, cardiac amyloidosis was diagnosed and the patient was put on a protocol consisting of cyclophosphamide, dexamethasone and bortezomib in addition to intravenous albumin and diuretics for heart failure and hypoalbuminemia, respectively. Sadly, the patient passed away from sepsis and arrhythmia on day 12 of treatment.

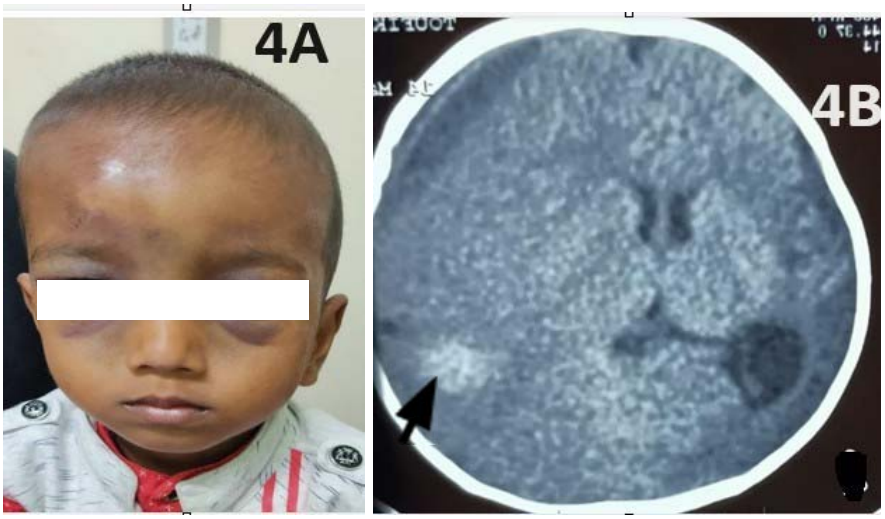

Fig. 4(a,b): (a) Raccoon eye at presentation of a case of severe haemophilia and (b) CT scan shows CNS bleed in the right parietal

Fig. 5: Raccoon eye bilateral periorbital bluish discolouration in a case of congenital afibrinogenemia

Case 4: A 2 year 5 month boy following a trauma, a known case of severe hemophilia A showed symptoms of swelling in the scalp. Upon general examination, there was no neurological deficit, bilateral Raccoon eyes and left hemarthrosis. It was suggested that he undergo inhibitor, brain and CT screening. High titre inhibitor positivity was found in the report and a CT scan revealed right parietal bleeding. The patient's raccoon eye sign subsided after 3 days of treatment with injection FEIBA@ 90 mcg kg^{-1} (Fig. 4).

Case 5: A 2 year child presented with unilateral periorbital ecchymosis and had been seen at a different pediatric facility for issues related to prolonged bleeding after minor trauma episodes. The mother had clearly disclosed to him the child's history of prolonged umbilical cord bleeding after delivery. Lacking a noteworthy family background. The coagulation profile showed prolonged PT and APTT and the fibrinogen level was less than normal (<190 mg dL^{-1}). For 2 days, the patient was treated for active bleeding episodes with an FFP transfusion at a dose of 10 mg kg^{-1} (Fig. 5).

DISCUSSION

Raccoon eyes were first linked to a base of the skull fracture but over time, they have been linked to a number of different clinical conditions. In the current article, six cases of hematological disorders that initially present as raccoon eyes are discussed.

The description of a case can vary from hypofibrinogenemia to acute leukemia. Identifying this clinical syndrome, then, offers a crucial hint based on clinical circumstances via research. This article's first case was identified as an AML case. Raccoon eyes, also known as periorbital ecchymosis, are not usually linked to acute myeloid leukemia (AML); however, there are certain AML related or AML treated complications that may cause this symptom.

Usually, bruising and bleeding around the eyes cause raccoon eyes. Raccoon eyes in the context of AML may result from a number of factors, such as: Thrombocytopenia: AML can cause thrombocytopenia, a low platelet count that makes people more prone to bleeding and bruising easily. Raccoon eyes may result from this, which may appear as bruises surrounding the eyes. Disseminated Intravascular Coagulation (DIC): AML may occasionally result in DIC, a condition that causes the clotting system to become widely activated, causing bleeding as well as clotting. DIC's bleeding component may be a factor in bruises, especially those near the eyes.

Leukemic infiltration: Rarely, leukemic cells may penetrate tissues, including facial bones, which may cause bruises and localized bleeding. AML manifestation.

Here, we display two AML cases that have comparable images. In the first instance, the patient's low platelet count resulted in raccoon eyes. RDP transfusion was used to treat an orbital bleed that was discovered on a CT scan without any mucosal thickening. Fungal sinusitis was the cause of the second case. After receiving intravenous (IV) amphotericin B therapy for fungal infections, a CT scan of the orbit revealed orbital bleeding and mucosal thickening.

Third instance Among patients with primary amyloidosis, raccoon eye sign was noted. In certain cases of amyloidosis, periorbital ecchymosis, also known as "raccoon eyes," can develop. A set of illnesses known as amyloidosis are defined by the buildup of aberrant proteins called amyloids in different tissues and organs, which causes organ dysfunction. Raccoon eyes are not a typical or characteristic symptom of amyloidosis but they can happen in some situations for the following reasons: Bleeding and Vascular Fragility: Blood vessels may become more fragile as a result of amyloid deposits. Fragile blood vessels are more likely to burst, which can cause bleeding or bruises in the tissues around the eyes and other surrounding areas[4-7].

Amyloid deposition in periorbital tissues: Amyloid deposits can occasionally build up in the tissues surrounding the eyes, resulting in localized harm and possibly bleeding. Secondary Complications: Bleeding disorders are among the many complications that amyloidosis can cause. Raccoon eyes and bruises can occur as a result of these complications. Hemophiliacs may experience periorbital ecchymosis, also known as "raccoon eyes." A blood condition known as hemophilia is caused by a lack or malfunction of clotting factors. Hemophilia may be connected to raccoon eyes: Hemophiliacs are more likely to experience spontaneous bleeding because they are deficient in certain clotting factors. This may cause bleeding in the skin surrounding the eyes as well as other tissues.

Trauma or injury: Even minor trauma or injury can cause bleeding in hemophiliacs. Intracranial Hemorrhage: In severe cases of hemophilia, spontaneous bleeding can occur in deeper tissues, including the brain. If bleeding occurs in the eye area or surrounding tissues, it can cause bruising and the appearance of raccoon eyes.

A traumatic event near the eyes may cause bruises and discolouration. In the fourth instance, severe hemophilia was noted. A patient who tested positive for titre inhibitors highly fifth instance congenital fibrinogenemia disorders, which include congenital afibrinogenemia, hypofibrinogenemia and dysfibrinogenemia, are uncommon pathologies of the hemostatic disorders with a highly variable clinical phenotype.

Although it's not a common or specific symptom of congenital fibrinogenemia, periorbital ecchymosis, also known as raccoon eyes, can occur in those with the bleeding disorder. The hallmark of congenital fibrinogenemia is a lack or insufficiency of fibrinogen, a vital protein involved in blood coagulation. One possible link between congenital hypofibrinogenemia and raccoon eyes is: Unplanned Bleeding: Congenital hypofibrinogenemia patients are more likely to experience spontaneous bleeding because they are less able to form stable blood clots.

This may result in bruises in a number of tissues, including the vicinity of the eyes. Bleeding into Soft Tissues: The skin surrounding the eyes is one area where bleeding into soft tissues, including it, can happen in severe cases of congenital hypofibrinogenemia.

This bleeding can cause discoloration, making raccoon eyes appear.

Trauma or injury: People with congenital fibrinogenemia are susceptible to bleeding from even small trauma or injury. Trauma to the area around the eyes can aggravate bruises and the development of fractures. The growth of eyes in raccoons.

Complications from raccoon eyes include meningitis, CSF leaks and palsies of the cranial nerves (cranial nerves I, II, III, IV, VI, VII and VIII).loss of hearing, Death from intracranial hemorrhage, vertigo

and cavernous sinus thrombosis Most CSF leaks go away on their own in 5 to 10 days, but some can linger for months.

Less than 5% of patients may develop meningitis, but the risk rises the longer the CSF leak lasts. In most cases, conductive hearing loss goes away in 7-21 days. Facial palsy and loss of smell are symptoms of deficits in the cranial nerve. Additionally, Mandal *et al*.[3] published a case of ITP with "Raccoon eyes," a presentation that was distinctive.

CONCLUSION

The significance of taking into account underlying hematologic conditions when evaluating individuals exhibiting this symptom is highlighted by the presence of raccoon eyes in hematology. To address the specific hematologic factors contributing to periorbital ecchymosis, hematologists and other specialists must collaborate to ensure timely diagnosis and appropriate management.

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