Spontaneous Orbital Hematoma-A Case Report

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Abstract

To report a case of a 4 months old girl presenting with massive spontaneous proptosis, due to a large hematoma resulting from systemic vitamin K deficiency. Systemic features resolved with appropriate medical management, but proptosis was persistent and needed surgical intervention to prevent visual loss. Such a presentation of vitamin K deficiency has not been previously reported in the literature.

Keywords: Proptosis; Hemorrhagic disease of newborn; Vitamin K deficiency; Infraclival approach; Tarsoraphy; Ocular motility; Conjunctival prolapse

Introduction

Spontaneous intraorbital hematoma is a rare condition and is usually attributed to trauma, neoplasm, acute sinusitis or vascular malformations [1]. It was reported to occur spontaneously in very few cases [2]. Rarely, it presents as an early manifestation of hematological disorders.

Vitamin K deficiency can cause bleeding in an infant in the first few weeks of life. This is known as Hemorrhagic Disease of the Newborn (HDN). HDN is divided into three categories: early, classic and late HDN. Early HDN occurs within 24 hours post-partum. Classic HDN occurs on day one to seven; common bleeding sites are gastrointestinal, cutaneous, nasal and from a circumcision. Late HDN occurs from week 2-12; the most common bleeding sites are intracranial, cutaneous, and gastrointestinal [3].

Here we report the first case of HDN with spontaneous intraorbital bleeding.

Case report

A 4 months old female, presented with severe, sudden, spontaneous, exophthalmos of the left eye (Figure 1). This was preceded with one day history of multiple bruises involving upper limbs, lower limbs and the back, followed by profuse conjunctival bleeding. However, there was no history of trauma, or previous bleeding diathesis. There were no concomitant ear, nose, or throat symptoms and the child was not reluctant to feed. There was no family history of bleeding or clotting disorders. There was no parent's consanguinity.

Systemic review excluded the presence of preceding diarrhea, jaundice, or difficulty in breathing.

There was severe conjunctival chemosis, ecchymosis, inferior conjunctival prolapse and profuse conjunctival bleeding, pupil was poorly reacting to light, proptosis was measured to be 28 mm and ocular motility was restricted in all directions of gaze. The eye was displaced superiorly and inward, liver was palpable 2 cm below right costal margin. There was no lymphadenopathy.

Laboratory findings revealed deranged bleeding and clotting profile, prothrombin time and activated partial thromboplastin time both were prolonged to 120 seconds. Bleeding time was 3 mins and clotting time was 4:30 minutes. Hemoglobin (Hb) was 7.8 mg/dl and blood smear showed microcytic hypochromic red blood cells with anisocytosis and the presence of elliptocytes. Leukocytes and platelets were normal in count and morphology. The rest of baseline investigations were within normal limits.

Figure 1: Initial presentation.
Fresh frozen plasma (120 ml) was transfused followed by 1 pint of packed cell volume. Vitamin K injections were administered intravenously OD for 5 days.

Computed tomography (CT) revealed a well-defined soft tissue mass in the left orbit measuring 34x40 mm in dimensions with gross exophthalmos, thinning of the lateral aspect of the left orbital roof. Optic nerve and extraocular muscles couldn’t be identified Figure 2. A diagnosis of a huge rhabdomyosarcoma involving the globe, optic nerve, and the extraocular muscles was made by the consultant radiologist.

As this did not correlate clinically magnetic resonance imaging of the orbit and brain was ordered. This revealed a large well circumscribed, ovoid lesion in the left orbit inferolaterally occupying the whole orbital cavity. The lesion was predominantly isointense to brain parenchyma on T1W images (Figure 3). On T2W images the lesion shows heterogeneously hypointense contents and fluid signals from its least dependent part (Figure 4). Post Gadolinium images showed rim enhancement of the lesion and these findings gave the impression of a large orbital hematoma.

Bruises and fever were relieved but proptosis persisted and needed surgical drainage of hematoma. Surgical drainage was delayed for 5 days due to poor bleeding profile. Moreover the patient had auto-decompression of the orbit due to gross anterior displacement of the lower lid and globe with laxity of lids. The procedure was performed under general anesthesia when the patient's bleeding and clotting profile stabilized after medical management, allowing for general anesthesia and surgical intervention. Moreover, the Cornea remained protected from exposure under the cover of upper eyelid, due to the inferolateral location of the hematoma, which displaced the eyeball supeomedially.

A lateral Infraciliary approach was used. A huge organized hematoma was seen posterior to the orbicularis muscle. It was drained by vacuum suction. A drain could not be placed due to the small age of the child. Examination under anesthesia after one week showed clear cornea and reduction of proptosis to 19 mm. Optic disc and macula were within normal limits. Retinoscopy revealed no error of refraction and intraocular pressure was 15 mm Hg. Conjunctival features and proptosis resolved over the following 15 days (Figure 5). The child was prescribed patching in the right eye to treat the amblyopia due to the globe displacement. The child was lost to further follow up even after repeated requests.
Discussion

Vitamin K deficiency bleeding (VKDB) in infancy is an acquired coagulopathy secondary to reduction of vitamin K (VK)-dependent coagulation factors (II, VII, IX, X), below hemostatic levels; 30-60 % of cases are associated with intracranial haemorrhage [4]. In a bleeding infant a prolonged PT together with a normal fibrinogen level and platelet count is almost diagnostic of VKDB; rapid correction of the PT and/or cessation of bleeding after VK administration are confirmative [5]. A normal PT for age excludes the diagnosis of VKDB [6].

Our case seems to fall in the category of late VKDB. Late VKDB begins on or after day 8, most often between weeks 2 and 8 and rarely after 3 months [4,7]. In co-operative studies the upper age limit was set arbitrarily at the end of week 12 [8]; but infants presenting with VKDB between weeks 13 and 26 should also be reported [9]. Our case presented at 18 weeks of age.

Late VKDB occurs almost exclusively in breast-fed infants, more often in boys than girls [4]; these infants often have history of diarrhea lasting for more than one week or antibiotic consumption within a week of the onset of the presenting bleeding which is commonly an intracranial one [10]. Its incidence ranges widely; for reasons which may include racial variation, maternal diet, and different VK prophylaxis regimens and compliance.

The child discussed in this report did not receive a prophylactic vitamin K intramuscular injection after birth. Child birth occurred at home under the care of insufficiently educated midwife. Here we point that the obstetric and neonatal care along with the compliance of prophylactic measures in Pakistan does not seem to be satisfactory. Emphasis upon initial prophylactic vitamin K supplementation and an additional intramuscular dose or oral supplementation of vitamin K especially for exclusively breast-fed infants may reduce this catastrophic problem [11].

APTT was also prolonged to 120 seconds in this child. This suggested coexisting deficiency of other clotting factors including factor V, factor X and factor 1. However plasma assays for these factors were not performed due to cost issues, especially when APTT level returned to normal after administration of vitamin K. We wanted to perform these assays later, but the child was lost for follow-up despite repeated requests and calls.

The late onset of VKDB can point to the presence of some secondary cause of the deficiency. Such possible causes include: chronic diarrhea, hepatitis, celiac disease, alpha1-antitrypsin deficiency and cystic fibrosis. These causes were clinically ruled out due the absence of suggestive symptom including diarrhea and jaundice.

The initial symptoms in a child with alpha1-antitrypsin deficiency include cough, sputum production, and wheezing. The child may have history of treatment with multiple courses of antibiotics and evaluation for sinusitis.

In cystic fibrosis, the first feature to be noticed by the parents, in a baby having the disease, may be salty skin when kissed. Other features appear as the disease progressively involves the pulmonary and the digestive systems. The child may have chronic diarrhea and failure to gain weight or grow, due to malabsorption resulting from the blockage of pancreatic ducts. Frequent chronic coughing and difficulty in breathing results from respiratory tract blockage with thick mucus.

In early reports on cases of VKDB, intracranial bleeding (ICH) was observed in 65-100%, [12] but in more recent reports in 30-60% [4]. Other common sites being: gastrointestinal mucosa& cutaneous [3]. Late VKDB has been also reported to manifest as nodular cutaneous purpura [13]. However, no cases of orbital bleeding are reported to occur as a manifestation of VKDB.

Most of the cases of non-traumatic orbital hemorrhages are subperiosteal. A subperiosteal orbital hemorrhage detected on computed tomography is usually superior in site, and results in no visual compromise. These features allow conservative management [14]. Orbital hematomas rarely cause visual deterioration, but reportedly an irreversible one, unless treated with early surgical decompression [15].

In the case of this report immediate medical management of bleeding diathesis was lifesaving. However, conservative management alone was found to be ineffective to treat the intraorbital, large, organized hematoma. Surgical intervention was required to restore visual function or prevent further deterioration.

Conclusion

Emphasis upon vitamin K prophylactic injections for every newborn should be increased. Neonatal spontaneous hemorrhages as well as hemorrhages in any infant should be suspected to be due to VKBD, the late type should be kept in mind.

References


