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Traumatic Optic Neuropathy: Surgical Versus Medical Treatment in Hemophilia- A Rare Case Report

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Abstract

Traumatic optic neuropathy (TON) refers to an injury to the optic nerve that results from a direct or indirect head and facial trauma. It can be classified as direct and indirect, based on the mechanism of injury causing the damage [1,2].

Hereditary bleeding disorders are conditions that occur due to the absence or deficiency of specific clotting proteins. Of these, Hemophilia A, hemophilia B and von Willebrand disease are the most common. Hemophilia is a genetic blood disorder caused by a missing or defective clotting factor that leads to an abnormality in clot formation. It is of two types; Hemophilia A (Due to deficiency or absence of Factor VIII) and Hemophilia B (Due to deficiency or absence of Factor IX).

In this case report we aim to showcase the necessity for surgical decompression in cases of Indirect TON even in patients with Hemophilia, as well as the possible benefits of preferring this modality of treatment to systemic steroids.

Keywords: Traumatic Optic Neuropathy (TON); Head and Neck; Trauma; People with Hemophilia (PWH); Optic nerve; Corticosteroids; Maxillary and Ethmoidal Sinuses

Abbreviations: TON: Traumatic Optic Neuropathy; PWH: People with Hemophilia; RGC: Retinal Ganglion Cell; RTA: Road Traffic Accident; CT: Computered Tomography

Introduction

Hemophilia A and B are the most common severe hereditary hemorrhagic disorders. Both hemophilia A and B are inherited via an X-linked recessive pattern, wherein males are commonly affected with women being carriers [3]. The estimated incidence of hemophilia is around 1 in 10000 live births, and the worldwide prevalence is nearly 400000 [4-6]. Hemophilia A is more prevalent than hemophilia B, making up 80-85% of the total hemophiliac population. When factor VIII and factor IX are essential in the intrinsic pathway of the coagulation cascade and their deficiency prevents proper activation of the mechanism, thus making clotting less effective. Thus, Hemophilia is considered a genetic disease in which the clinical manifestation is mainly the presence of hemorrhage [7].

Traumatic optic neuropathy (TON) is a relatively rare, but severe condition that can lead to irreversible vision loss. It is classified as Direct and Indirect TON, with the latter being the more common type. Direct TON is characterized by the presence of an open wound, leading to direct injury of the optic nerve, usually due to a traumatic injury by a sharp object [2]. Indirect TON, on the other hand, results from a blunt trauma to the head with the site of injury usually being the forehead or supraorbital ridge, and less commonly the temporal region [1,7]. The prevalence of indirect TON in closed-head trauma has been reported as 0.5% to 5% [1,2].

Frontal trauma can result in the deformation of the ipsilateral orbital roof near the optic foramen, and that such deformation of the orbital roof could damage not only the supporting vasculature

of the optic nerve but could also cause shear stress to the nerve [8]. According to the two-stage model, after the initial impact trauma, the Retinal Ganglion Cell (RGC) axons of the optic nerve are sheared at the level of the optic foramen. The subsequent oedema due to vascular ischemia and mechanical pressure then causes optic nerve swelling [9].

Case Report

The patient in question was presented to a Government District hospital following a Road Traffic Accident (RTA) with loss of consciousness for one hour, two discrete episodes of vomiting, nasal bleeding, and with multiple bruises on his face. He also had a history of Hemophilia-A diagnosed two years prior. His Computered Tomography (CT) investigations performed at a higher center showed segmental displaced fractures of the left zygoma, lateral wall of the left orbit and the left maxillary sinus. It also showed evidence of fracture of the left lateral pterygoid plate, left glenoid fossa, and the floor and lateral wall of the left orbit. Hemosinus of the right maxillary and ethmoidal sinuses was also noted. The Computered Tomography (CT) brain scan showed no abnormality.

Multiple evidence of blunt force trauma was noted. On examination, there was no gross facial asymmetry but the left eye showed periorbital ecchymosis, loss of continuity with respect to the lateral wall, and a restriction in movement. Pupillary reaction was also sluggish. The left infraorbital region and zygomatic buttress were mildly tender to palpation. Furthermore, there was a slight deviation to the right side when opening the mouth, which was itself more than 40mm.

Due to limited awareness in peripheral health centers regarding TON and its consequences, conservative management was initiated with a broad-spectrum oral antibiotic, antibiotic eye drops, as well as oral pain medication. Immediate steroid administration was, hence, delayed. The conservative treatment was continued and a hematoma accumulated retro-orbitally gradually compressing upon the optic nerve. The damage was identified after the patient complained of blurring of vision. Arrangements were made immediately to shift the patient to a specialized center. The patient's vision however, continued to deteriorate in the left eye and had descended into complete blindness by the time he was examined by a specialist. Megadose steroids were given intravenously and an epicanthic incision was considered to drain the hematoma. However, there was a concern about the management of subsequent bleeding considering his Hemophilia and a possibility of vision loss in the right eye that could lead to debilitating blindness. Furthermore, the left optic nerve had already been extensively damaged and probability of regaining any vision was extremely bleak.

At the end of the treatment course, the patient had unfortunately lost his vision completely in the left eye, which could not be rescued by steroids (Figure 1).

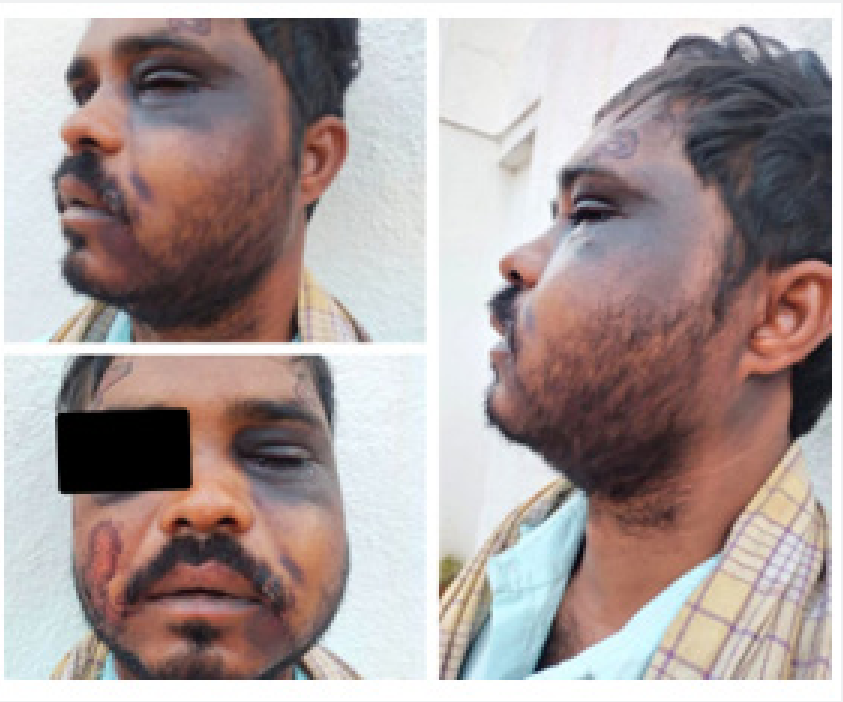

Figure 1: At the end of the treatment course, the patient had unfortunately lost his vision completely in the left eye, which could not be rescued by steroids.

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Discussion

The primary modalities of treatment of Indirect TON are 1) Systemic high dose steroid therapy; 2) Optic nerve decompression or a combination of the two [9]. However, in recent times, Erythropoietin (EPO) has been observed to be effective in reducing neural apoptosis and ischemic brain injury [10,11]. Of these treatments, early decompression is usually underperformed in patients with bleeding disorders in clinical practice, due to apprehensions regarding the risk of uncontrolled bleeding. Thus, these patients are often restricted to Megadose systemic steroids, particularly intravenous Methyl-Prednisolone [9].

In this case, however, surgical decompression was not considered due to apprehension regarding the patient's hemophilia history and high-dose Prednisolone was administered intravenously. However, due to continued collection periorbitally and local edema, the optic nerve continued to endure pressure-related damage. The long-term impacts of such situations require careful consideration. Deterioration of vision and possible irreversible blindness are debilitating and cause significant reduction in the quality of life of the patient.

With recent advancements in both the management of hemophilia and developments in head and neck and trauma surgery, surgical management should be promptly adopted in cases where it is indicated. This is especially true for time sensitive cases like the one we have discussed in this report [12].

Conclusion

The outcome of this case highlights the long-term consequence of inadequate timely management of Indirect TON. The prolonged conservative management and the lack of prompt administration of high dose corticosteroids or initiation of surgical management led to the build-up of pressure in the left orbital region. Thus, this evidences the need to educate clinicians and well and initiate a protocol for management of TON cases at both District-level as well as in specialized centers so as to prevent such cases of delay in the future (Figure 2).

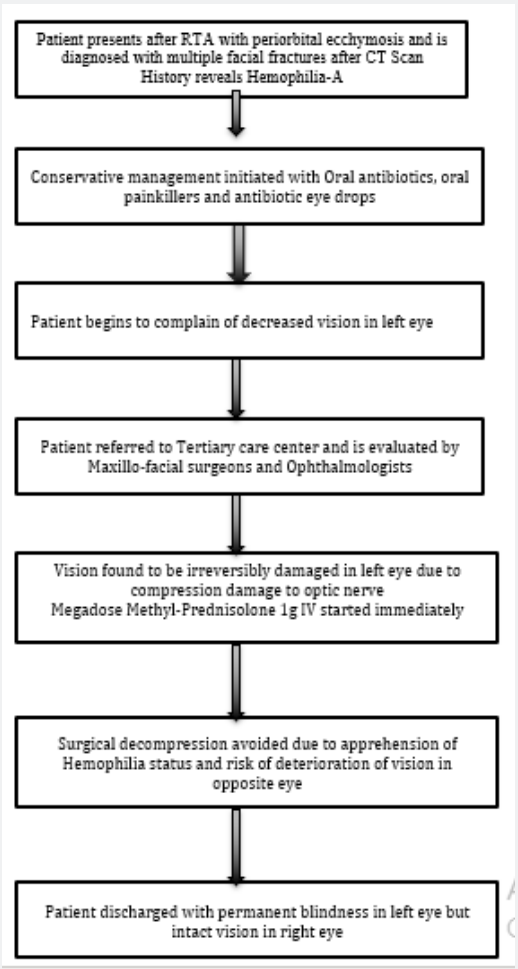

Figure 2: The outcome of this case highlights the long-term consequence of inadequate timely management of Indirect TON.

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