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Case Report

Bilateral Nonarteritic Anterior Ischemic Optic Neuropathy in Glanzmann Disease

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Abstract

Purpose: To describe our experience of bilateral sudden vision loss in a rare autosomal recessive (AR) platelet aggregation disorder, that is Glanzmann´s thrombasthenia (GT).

Observation: We report nonarteritic anterior ischemic optic neuropathy (AION) as a cause, following severe attack of gynecological bleeding in a 15-years-old girl. She had recurrent attacks of blood loss and subsequent hypovolemia requiring blood transfusion. This form of optic neuropathy is diagnosed due to the presence of bilateral pale swollen optic discs and proved by altitudinal pattern of perimetry along with exclusion of other reasons and normal visual evoked potential.

Conclusion: Nonarteritic AION should be considered in young patients diagnosed with bleeding disorders, whenever vision loss is preceded by hypovolemia. Reversibility of vision loss is possible

Keywords: Blood Loss; Hypovolemia; Glanzmann; Anterior Ischemic Optic Neuropathy; Case Report

Abbreviations

AR: Autosomal Recessive; GT: Glanzmann´s Thrombasthenia; AION: Anterior Ischemic Optic Neuropathy; CBC: Complete Blood Count; MRI: Magnetic Resonance Imaging; MRV: Magnetic Resonance Venography; CSF: Cerebrospinal Fluid; pVEP: Pattern Visual Evoked Potential; fERG: Flash Electroretinogram

Introduction

Anterior ischemic optic neuropathy results from hypoperfusion of the optic nerve head through the posterior ciliary arteries. Few reports of children and young adults with AION make perception of diagnosis really difficult especially in rare condition as GT.

Materials and Methods

Patient consent

This report does not contain any personal information that could lead to the identification of the patient.

Case Presentation

Fifteen years old female was referred to ophthalmology outpatient clinic from pediatric department. She presented with an attack of sudden painless drop of vision of 1 week duration affecting the right eye more than left eye. Her condition started by multiple ecchymosis and severe gingival bleeding at the age of 3 years old [1]. She also had a positive family history of similar condition with her sister [2]. Flow cytometry assay of platelets showed deficient CD61 and CD41 expression confirming the diagnosis of GT (Figure 1) [3,4]. Past surgical history was unremarkable. The patient had recurrent attacks of blood loss requiring multiple blood and plasma transfusions. She had gynecological bleeding attacks 4 years ago with puberty and the latest was the most severe, prior to her presentation to the ophthalmology clinic. She had increased frequency of blood and plasma transfusions since the attack [4].

General examination revealed fully conscious patient with pallor and multiple ecchymosis, blood pressure of 90/55 mm Hg, pulse

Figure 1: Flow cytometry assay showing deficient CD61 (GpIIIa) and (GPIIIb) expression consistent Glanzmann thrombasthenia.

of 100 bpm and mild gingival bleeding. Ophthalmic examination revealed initial decimal visual acuity of 0.08 in the right eye and 0.05 in the left eye. Pupils were reactive, anterior segment examination was unremarkable. Fundus examination showed bilateral swollen optic nerve heads, blurring of disc margins, and pale discs more evident in the right eye (Figure 2). Investigations were done by sequence in accordance with clinical thinking. Complete blood count (CBC) showed red blood cells count of 3.11 x 106/µl, Hemoglobin of 8.1 g/dl, hematocrit value 24.5%, white blood cells count of 6.7 x 103/µl, and platelet count of 214 x 103/µl. Platelet aggregation to ADF was 6% and to ristocetin was 40% [4,5]. Erythrocyte sedimentation rate was normal and c- reactive protein was negative.

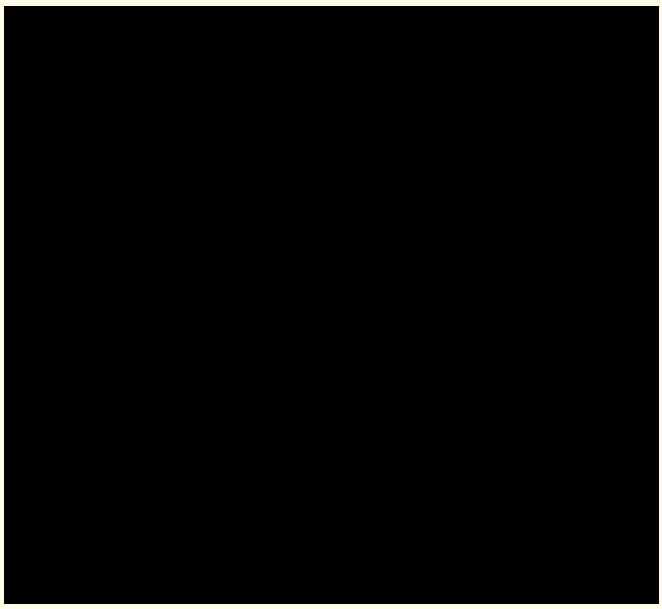
Figure 2: Presenting bilateral pale optic discs edema more evident in the right eye.

Magnetic resonance imaging (MRI) of the brain, orbits and MRV (magnetic resonance venography) of cerebral vessels showed no abnormality (Figure 3). Lumber puncture was performed for the patient showed cerebrospinal fluid (CSF) opening pressure of 14 cm H_2O, normal glucose and protein levels. Electrophysiological studies were performed after some improvement of VA. Pattern visual evoked potential (pVEP) was done for large and small checks showing within normal P100 latency and amplitude (Figure 4). Flash electroretinogram (fERG) showed normal photopic and scotopic a and b waves, with normal latencies and amplitudes, normal oscillatory potentials, and flicker response (Figure 5). Field examination could not be performed initially due to poor visual acuity.

Figure 3: Normal MRV.

Figure 4: Normal pVEP (large and small checks).

a



b

Figure 5: (a-b): Normal fERG showing photopic (5a) and scotopic (5b).

The differential diagnosis included high intracranial tension either benign or due to space occupying lesion as intracranial hemorrhage. Other reason of the condition could be thrombosis of the major venous sinuses or superficial and deep veins of the brain. Differential diagnosis includes also other types of optic neuropathy whether inflammatory, postinfectious, infiltrative, vitamin b deficiency or autoimmune. Ischemic optic neuropathy due to hypovolemia was our most likely diagnosis.

Results and Discussion

Outcome and follow-up

Patient was reassured and advised to continue repeated blood and plasma transfusions in pediatric department. Her vision was subjectively better over the subsequent 8 weeks. At the end of the second month, her vision improved to decimal visual acuity of 0.2 (right eye), 0.25 (left eye). Fundus examination showed less swelling of both optic nerve heads with pallor affecting left optic disc more than the right (Figure 6). Complete blood count (CBC) showed red blood cells of 4.37 x 106/µl, Hemoglobin of 11.2 mg/dl, white blood cells of 6.5 x 103/µl, and platelets of 203 x 103/µl. MRI was repeated with MRV of cerebral vessels and both were normal. Field examination using the Humphrey Field Analyzer (HFA II, Carl Zeiss Meditec, Dublin, CA) showed superior altitudinal and lower temporal quadrant defects with central encroachment (right eye), and inferior hemifield affection with nasal step together with enlarged blind spot (left eye) (Figure 7 a-b). Her vision continued to improve to Decimal visual acuity of 0.8 each eye.

Figure 6: Less swelling of both optic discs with pallor affecting left more than the right disc 2 month after the attack.

Figure 7a: Superior altitudinal and lower temporal quadrant defects with central encroachment (right eye).

Figure 7b: Inferior hemifield affection with nasal step together with enlarged blind spot (left eye).

Discussion

Non-arteritic anterior ischemic optic neuropathy (AION) is the most common acute optic neuropathy in patients over 50 years due to astherosclerosis, with increased risk in ischemic heart diseases, hypercholesterolemia and diabetes. It results from hypoxia of the optic nerve head anterior to lamina cribrosa [6]. Posterior ischemic optic neuropathy has been reported more commonly after spinal surgery and radical neck dissection [7]. Non-arteritic AION, which is usually bilateral and up to 10 days from the onset of bleeding, results from acute anemia or hypotension. This is explained by decrease in optic nerve perfusion pressure (6) or increase in endogenous vasoconstrictors (as epinephrine) due to stimulation of the sympathetic adrenergic system by the vasomotor center or increase in platelet aggregation [8].

We report Non-arteritic AION in a 15 years old girl with GT, a dysfunctional platelet disorder impairing proper clotting mechanism. Her presentation to ophthalmology was bilateral sudden drop of vision with bilateral pale edematous optic discs. Our differential diagnoses list included high intracranial tension, either benign or due to intracranial hemorrhage. This possibility was excluded by normal MRI brain. Thrombosis of superficial and deep cerebral veins or intracranial venous sinuses was excluded by normal MRV. Other causes of optic neuropathy as infection or autoimmune affection were excluded by blood tests.

We considered the diagnosis of nonarteritic AION relying on the history of bilateral sudden painless drop of vision 1 week following an attack of severe blood loss, the fundus picture of bilateral pale swelling of optic nerve heads, and her reported field defects [8].

Our case denotes that the proposed mechanism of increased platelet aggregation (8) is not possible, because GT is characterized by decreased platelet aggregation. This is supported by the list of risk factors for AION that did not include any role for thrombophilia [6]. In our case there was a clear improvement of visual acuity upon increasing the rate of blood transfusion. This denotes the importance of early diagnosis and management of ischemic changes before they become irreversible.

A previously reported case of vision loss in a healthy woman with history of menorrhagia described nonarteretic AION as the cause. The reported vision loss was unilateral, and it did not improve. This may be explained by more severe anemia (hemoglobin of 3.6 g/dL) and the patient refusal for blood transfusion [9].

Another case was reported as unilateral nonarteretic AION with irreversible vision loss following hysterectomy. Patient had severe anemia (hemoglobin 7.2 g/dl), refused blood transfusion, and re-operated on with extraction of a 1.5-liter blood clot [10]. Two cases of young children on peritoneal dialysis were also reported with bilateral AION [11]. In our case, pupils were reactive, pVEP and fERG were normal. These investigations were not done in the previously mentioned case reports to exclude bilateral optic neuritis. Non-arteritic AION is not accompanied by neurologic defect [11]. Similar to our case variable field defects were reported in AION [12].

The time interval and the bilaterality of vision loss in our case report were attributed to blood loss related optic nerve ischemia [8].

We suggest that the mechanism of non-arteritic AION in our case is reduced blood oxygen carrying capacity and decreased arterial perfusion pressure [10]. Another proposed mechanism is volume depletion and its effect on releasing vasoconstrictor substances affecting posterior ciliary vessels, which supply the optic nerve head [11]. Up till now, there is no effective treatment that improves vision loss in nonarteretic AION in older age group [13]. In young adults, similar to our case report, vision can be improved to a good extent, if the blood loss is promptly treated [8].

Conclusion

Nonarteretic AION due to massive blood loss and hypovolemia, although not the most common, should be considered and can be salvageable if diagnosed and managed early. This could be applied in any pediatric blood dyscrasias.

Acknowledgements

None.

Conflict of Interest

No financial interest or any conflict of interest exists.

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