Nontraumatic subperiosteal orbital hemorrhage in a laboring patient with gestational immune thrombocytopenic purpura

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ABSTRACT

Purpose: To describe a case of nontraumatic subperiosteal orbital hemorrhage (NTSOH) in a laboring patient with gestational immune thrombocytopenic purpura.

Observations: A 28-year-old G3P2 laboring patient was urgently evaluated by our ophthalmology unit after she developed sudden onset left eye proptosis, headache, and diplopia in the final hour of pushing. The patient’s platelet count was markedly decreased at 45,000 and subsequent work-up established a diagnosis of gestational immune thrombocytopenic purpura. On examination, visual acuity was 20/25 bilaterally and there was a minus two restriction in upgaze in the left eye and a left hypotropia in primary position. Computed tomography demonstrated an elliptical, hyperdense collection at the left orbital roof consistent with NTSOH. The patient was deemed clinically stable through serial examinations and symptoms resolved with conservative management.

Conclusions and Importance: NTSOH is a rare, potentially sight-threatening condition that requires timely ophthalmological evaluation. To our knowledge, this is the first report in the literature of NTSOH in a laboring patient with gestational immune thrombocytopenic purpura. Consideration of the possibility of NTSOH as a complication in this population may allow for appropriate diagnosis, monitoring, and treatment when indicated.

1. Introduction

Immune thrombocytopenic purpura (ITP) is a clinical syndrome characterized by an isolated thrombocytopenia (<100 × 10^9 platelets/L) in the context of otherwise normal hematology. 1 It occurs as a result of auto-antibody production by B cells against platelet surface glycoproteins, resulting in platelet destruction by resident macrophages. As the rate of platelet destruction often outpaces the rate of thrombopoiesis, a marked thrombocytopenia results. 2 Clinically, this presents as mucocutaneous bleeding and easy bruising, however, ITP may sometimes lead to life-threatening intracranial hemorrhages or other systemic bleeds. 3 ITP is relatively common within the realm of hematological disorders, with a prevalence of approximately 12 per 100,000 in adults in the United States. 4 In the context of pregnancy, this prevalence increases substantially, to approximately 1–3 in 10,000. 5

Nontraumatic subperiosteal orbital hemorrhage (NTSOH) is a rare, potentially sight-threatening condition in which bleeding into the subperiosteal space causes local compression of ocular and adnexal structures. 6 NTSOH has been reported in the context of increased venous pressure as a result of various etiologies including coughing/straining, 7,8 physical exertion, 9 scuba diving, 10–12 strangulation, 13 and birth. 14,15 Systemic conditions such as coagulopathies, 16 liver disease, 17–18 cancer, 19 and sickle cell disease have also been implicated. 20,21 NTSOH has also been reported in the setting of sinusitis 22 and following non-ophthalmic surgical intervention. 23–26 Left untreated, NTSOH may lead to compression of the optic nerve and permanent visual loss. Patients with NTSOH require urgent evaluation, and there exist multiple treatment options ranging from observation to, steroid therapy, needle aspiration, and surgical evacuation. 27

Here, we describe a case of a laboring patient with gestational ITP, referred to the ophthalmology service with NTSOH.

2. Case report

A 28-year-old female, G3P2, presented in labor to an outside hospital in December 2018. During assessment for an epidural placement her platelet count was noted to be 45,000 (previously 212,000 in September 2018). INR and PTT were normal. The patient received tranexamic acid

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and oxytocin. In the final hour of pushing, the patient developed a sudden onset of left eye proptosis and complained of diplopia and headache without associated focal neurologic deficits. The remainder of the delivery was uneventful with no postpartum hemorrhage and no signs of hemodynamic instability. The baby was healthy with a platelet count of 385,000. Ophthalmology was urgently consulted following the delivery and the patient was transported to our tertiary care center where a CT head without contrast was performed revealing a 2.9 cm × 2.7 cm × 0.8 cm, biconvex, extra-conal, superior subperiosteal hematoma in the left orbit (Fig. 1).

Past ocular history was remarkable for a dermoid cyst removed from the right brow in infancy. Past medical history included mild iron-deficiency anemia and previous uncomplicated spontaneous vaginal deliveries in 2014 and 2016. The patient reported no allergies. Family history was negative for any bleeding disorders or autoimmune diseases. Social history was unremarkable. The patient denied any petechiae, purpura, spontaneous bruising, melena, hematochezia, or hematuria. Two weeks prior to the delivery the patient reported having an upper respiratory infection that was self-limiting and had resolved.

On examination, visual acuity was 20/25 bilaterally without correction at near, pupils were equal and reactive to light, with no evidence of relative afferent pupillary defect, and pressures by tonometry were 15 mm Hg in the right eye and 22 mm Hg in the left eye. Extra-ocular movements showed a minus two restriction in up gaze in the left eye as well as a left hypotropia in primary position. Red color saturation was equal. There was 2 mm of lid ptosis on the left. Anterior segment examination was unremarkable and posterior segment examination was deferred to allow for pupil monitoring. Serial ophthalmic examinations were performed over the next 3 h and the patient was deemed clinically stable. With no signs of optic neuropathy present and the resolution of the acute process of increased venous pressure, the decision was made to defer pupil monitoring. Serial ophthalmic examinations were performed over the next 3 h and the patient was deemed clinically stable. With no signs of optic neuropathy present and the resolution of the acute process of increased venous pressure, the decision was made to discharge the patient from the ophthalmology service and transfer her to the maternity ward. She was asked to monitor her visual acuity and color vision and nursing staff were instructed to conduct regular pupil checks.

The following morning the patient received a platelet transfusion and her platelet level increased to 89,000. The patient was brought to the ambulatory eye clinic where examination was overall stable. Additional testing at this time revealed full Ishihara color plates bilaterally, 4 mm of left sided proptosis by Hertel, and a normal dilated fundus examination. The patient was subsequently seen at days 3, 7, 14, 30, and 60 postpartum. The patient’s symptoms of diplopia and extra-ocular movement limitation improved to resolution over the first three weeks and the proptosis fully resolved at the 60-day follow up.

Fig. 1. Sagittal (A) and coronal (B) sections of noncontrast computed tomography (CT) scan on referral to ophthalmology service following onset of sequelae during labor. A) CT scan clearly shows an ellipsoid mass indicative of the hemorrhage in the left superior orbital space with proptosis of the globe. Compression of the optic nerve is not seen. B) Coronal view of the hemorrhage with compression of the globe and proptosis visible. In both panels, the green arrow indicates the hemorrhage. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

Subsequent investigations by internal medicine, rheumatology, and hematology were negative apart from mildly elevated single-stranded binding protein. The patient’s hemoglobin remained stable and normal throughout the admission. Her platelets slowly normalized over the course of three months to her baseline of 200,000–230,000 and a diagnosis of gestational ITP was made.

3. Discussion

Nontraumatic orbital hemorrhages can be classified according to their anatomic location as diffuse intraorbital, localized intraorbital, subperiosteal, related to extraocular muscles, or related to orbital floor implants. NTSOH has previously been reported in the context of pregnancy, with ITP may be regarded as a milder coagulopathy of ITP and a ‘perfect storm’ is created to allow for a subperiosteal bleed. To date, there have been 3 cases of NT SOH associated with labor, and 4 cases in the peripartum setting. In the context of labor, all patients presented with unilateral findings and were managed conservatively without any permanent sequelae upon resolution. Of the four peripartum cases of NT SOH, believed to be related to either vomiting or post-partum hemorrhage, 2 required orbitotomy and drainage with resultant optic neuropathy, while 2 were managed conservatively and had complete resolution of sequelae. Notably, permanent sequelae from optic nerve compromise were seen only in the two patients with significant coagulopathy from suspected disseminated intravascular coagulation. ITP may be regarded as a milder coagulopathy than disseminated intravascular coagulation and in our patient did not result permanent damage.

Given that the physiologic changes concurrent with pregnancy often induce a coagulopathy, and especially, thrombocytopenia, it may be prudent to consider NT SOH as a possibility in laboring patients. The reason for the increased prevalence of ITP in pregnancy is unknown, however, the pathophysiology of the disorder remains the same. It is possible that physiologic changes associated with pregnancy unmask a low-grade thrombocytopenia, or that the increased screening that pregnant women are subject to allows for detection of otherwise asymptomatic disease. Obstetricians should pay close attention to patients complaining of vision loss, and should be mindful to check for any evidence of proptosis of the globe. Work-up of visual complaints in the laboring patient should include NT SOH on the differential, among other common diagnoses such as pregnancy-induced hypertension, especially if the patient has an underlying coagulopathy. Although in our case the hemorrhage resolved under careful monitoring, an urgent ophthalmologic referral and intervention is warranted in these patients. Pregnant patients with underlying coagulopathies should be educated on the symptoms of NT SOH and instructed to report to their nearest emergency room if they experience symptoms of diplopia and extra-ocular movement limitation.

To our knowledge, this is the first reported case of NT SOH in a laboring patient with gestational ITP. Diagnosis of NT SOH is by radiographic imaging, usually computed tomography (CT) without contrast, where a biconvex mass of varying hyperintensity (corresponding to the age of blood) can be noted with associated displacement of surrounding tissue. Diagnosis is supported by clinical evaluation, with patients reporting diplopia, pain, and visual disturbances and visible proptosis, eyelid edema and periorbital ecchymosis present on exam. The superior orbit remains the most commonly reported location of NT SOH and hemorrhage is believed to occur more commonly in children and young adults as their nascent connective tissue is less adherent to bony structures, easily separating and allowing for hemotoma formation.

It is expected that straining in labor acts similar to other Valsalva maneuvers in increasing intraabdominal and intrathoracic pressures. Plausibly, the resulting increase in jugular venous pressure is then transmitted to the orbit by the venous supply, where distension of valveless diploic veins may increase the likelihood of rupture. Similar to subdural hematomas, the bridging vessels to the periorbita, in proximity to the bony surface of the orbital roof, may be sheered or burst due to this anatomical relationship. Couple this with the underlying coagulopathy of ITP and a ‘perfect storm’ is created to allow for a subperiosteal bleed. To date, there have been 3 cases of NT SOH associated with labor, and 4 cases in the peripartum setting. In the context of labor, all patients presented with unilateral findings and were managed conservatively without any permanent sequelae upon resolution. Of the four peripartum cases of NT SOH, believed to be related to either vomiting or post-partum hemorrhage, 2 required orbitotomy and drainage with resultant optic neuropathy, while 2 were managed conservatively and had complete resolution of sequelae. Notably, permanent sequelae from optic nerve compromise were seen only in the two patients with significant coagulopathy from suspected disseminated intravascular coagulation. ITP may be regarded as a milder coagulopathy than disseminated intravascular coagulation and in our patient did not result permanent damage.

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department or ambulatory ophthalmology clinic should any sequelaecome present.

4. Conclusions

To the best of our knowledge, this is the first reported case of NTSOH
in a laboring patient with ITP. As thrombocytopenia is a common
complication associated with the physiologic changes of pregnancy, and
increased venous pressure is a known consequence of straining, it is
imperative that obstetricians be on alert for any symptoms of vision loss
and proptosis. Similarly, colleagues in ophthalmology should be suspi-
cious of NTSOH in laboring patients referred for visual complaints.
Given the potential for permanent impairment, timely assessment,
diagnosis, and intervention can mean the difference between functional
vision and blindness for the patient.

Patient consent

Consent to publish was not obtained. This report does not contain
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