American Journal of Ophthalmology Case Reports 20 (2020) 100986

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

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

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ARTICLE INFO

Keywords:
Subperiosteal hemorrhage
Labor
Immune thrombocytopenic purpura

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 ($<100 \times 10^9$ 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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https://doi.org/10.1016/j.ajoc.2020.100986
Received 24 July 2019; Received in revised form 1 July 2020; Accepted 25 October 2020
Available online 28 October 2020
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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 x 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. Extraocular 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 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.

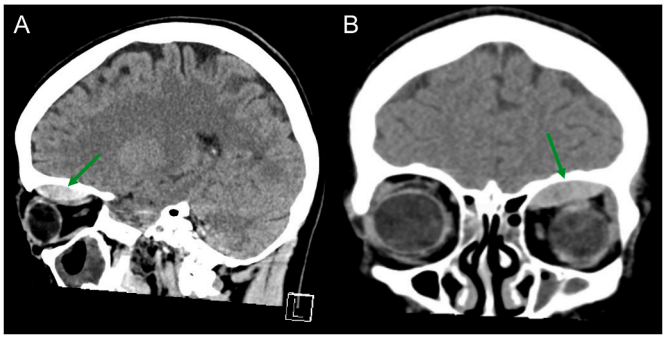

Fig. 1. Sagittal (A) and coronal (B) sections of noncontrast computed tomography (CT) scan on referral to ophthalmology service following onset of sequelae during labor. A) CT scan clearly shows an ellipsoid mass indicative of the hemorrhage in the left superior orbital space with proptosis of the globe. 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.[27] NTSOH has previously been reported in the context of pregnancy[6,28–30] as well as in the setting of bleeding disorders.[6,16,27] To our knowledge, this is the first reported case of NTSOH in a laboring patient with gestational ITP. Diagnosis of NTSOH 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.[6] 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 NTSOH 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 hematoma formation.[27]

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 NTSOH associated with labor,[6,28–30] and 4 cases in the peripartum setting.[6,29–31] 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 NTSOH, 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 sequalae from optic nerve compromise were seen only in the two patients with significant coagulopathy from suspected disseminated intravascular coagulation.[6,30] 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 NTSOH 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 NTSOH 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 ophthalmological referral and intervention is warranted in these patients. Pregnant patients with underlying coagulopathies should be educated on the symptoms of NTSOH and instructed to report to their nearest emergency

department or ambulatory ophthalmology clinic should any sequelae become 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 suspicious 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 any personal information that could lead to identification of the patient.

Funding

No funding or grant support.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The following authors have no financial disclosures: CS, SN.

Acknowledgements

None.

References

1. Rodeghiero F, Stasi R, Gernsheimer T, et al. Standardization of terminology, definitions and outcome criteria in immune thrombocytopenic purpura of adults and children: report from an international working group. *Blood*. 2009;113(11):2386–2393.
2. Toltl LJ, Arnold DM. Pathophysiology and management of chronic immune thrombocytopenia: focusing on what matters. *Br J Haematol*. 2011;152(1):52–60.
3. Neunert C, Noroozi N, Norman G, et al. Severe bleeding events in adults and children with primary immune thrombocytopenia: a systematic review. *J Thromb Haemostasis*. 2015;13(3):457–464.
4. Terrell DR, Beebe LA, Neas BR, Vesely SK, Segal JB, George JN. Prevalence of primary immune thrombocytopenia in Oklahoma. *Am J Hematol*. 2012;87(9):848–852.
5. Care A, Pavord S, Knight M, Alfirevic Z. Severe primary autoimmune thrombocytopenia in pregnancy: a national cohort study. *BJOG*. 2018;125(5):604–612.
6. Atalla ML, McNab AA, Sullivan TJ, Sloan B. Nontraumatic subperiosteal orbital hemorrhage. *Ophthalmology*. 2001;108(1):183–189.
7. Reid JR, Rajak SN, Good CD, Brittain GP. Emesis-induced sub-periosteal haematoma: a case report. *Orbit*. 2014;33(3):226–228.
8. Elia MD, Shield D, Kazim M, et al. Spontaneous subperiosteal orbital hemorrhage. *Orbit*. 2013;32(5):333–335.
9. Katz B, Carmody R. Subperiosteal orbital hematoma induced by the Valsalva maneuver. *Am J Ophthalmol*. 1985;100(4):617–618.
10. Chen JC, Kucharczyk W. Nontraumatic orbital subperiosteal hematoma in a scuba diver: CT and MR findings. *J Comput Assist Tomogr*. 1985;12(3):504–506.
11. Woo D, Rogers S, Leong J, Clement CI, Kourt G. Non-traumatic subperiosteal orbital hemorrhage secondary to barotrauma. *Orbit*. 2012;31(5):347–349.
12. Crawford C, Mazzoli R. Subperiosteal hematoma in multiple settings. *Digit J Ophthalmol*. 2013;19(1):6–8.
13. Subiras X, Koch KR, Schrittenlocher S, Cursiefen C, Heindl LM. Spontaneous unilateral subperiosteal hematoma in the orbit due to self-induced asphyxia: unusual cause of unilateral exophthalmos. *Cas Rep Ophthalmol*. 2017;8(1):232–236.
14. Skull SA, Sullivan TJ. Subperiosteal orbital haematoma presenting as proptosis at birth. *J Paediatr Child Health*. 1997;33(3):256–258.
15. Angkasa F, Mohammadi L, Taranath D, Taranath A, Brecht M. Orbital subperiosteal hematoma in the newborn causing unilateral proptosis: ultrasound as first-line imaging modality. *AJP Rep*. 2017;7(2):e138–e143.
16. Case of bilateral non-traumatic subperiosteal orbital haematomas. *J Med Imaging Radiat Oncol*. 2013;57(2):202–204.
17. Moorthy RS, Yung CW, Nunery WR, Sondhi N, Fogle N. Spontaneous orbital subperiosteal hematomas in patients with liver disease. *Ophthalmic Plast Reconstr Surg*. 1992;8(2), 150–02.
18. Griffeth MT, Dailey RA, Ofner S. Bilateral spontaneous subperiosteal hematoma of the orbits: a case report. *Arch Ophthalmol*. 1997;115(5):679–680.
19. Capua JK, Stiner ES, Li TG. Spontaneous subperiosteal orbital hematoma as initial presentation of metastatic lung adenocarcinoma to the skull: case report. *Orbit*. 2014;33(2), 152–05.
20. Sundu C, Dinç E, Sari A, Ünal S, Ö Dursun. Bilateral subperiosteal hematoma and orbital compression syndrome in sickle cell disease. *J Craniofac Surg*. 2017;28(8):e775–e776.
21. Alghamdi A. Recurrent orbital bone sub-periosteal hematoma in sickle cell disease: a case study. *BMC Ophthalmol*. 2018;18(1):211.
22. Choi S, Lawson W, Urken ML. Subperiosteal orbital hematoma. An unusual complication of sinusitis. *Arch Otolaryngol Head Neck Surg*. 1988;114(12):1464–1466.
23. Mangla D, Carruth BP, Britton LW, Wladis EJ. Nontraumatic subperiosteal bilateral orbital hemorrhages after mitral valve repair. *Ophthalmic Plast Reconstr Surg*. 2014;30(1):e1–2.
24. Diatchuk V, Rehany U, Rumelt S. Spontaneous subperiosteal orbital hemorrhage following thrombolytic therapy for myocardial infarction. *Ophthalmic Plast Reconstr Surg*. 2007;23(6):482–484.
25. Gore SS, Rathi N, Ganvir AY, et al. Nontraumatic subperiosteal orbital hemorrhage following upper gastrointestinal endoscopy. *Indian J Ophthalmol*. 2018;66(6):877–879.
26. Wu HP, Tsai CJ, Tsai JP, Hung CL, Kuo JY, Hou CJ. Spontaneous retro-orbital subperiosteal hemorrhage with complete resolution following percutaneous coronary intervention for acute myocardial infarction. *Acta Cardiol Sin*. 2013;29(2):181–184.
27. McNab AA. Nontraumatic orbital hemorrhage. *Surv Ophthalmol*. 2014;59(2):166–184.
28. Jacobson DM, Itani K, Digre KB, Ossoinig KC, Varner MW. Maternal orbital hematoma associated with labor. *Am J Ophthalmol*. 1988;105(5):547–553.
29. Harrie RP, Digre KB. Nontraumatic subperiosteal orbital hemorrhage during labor. *Ophthalmology*. 2001;108(11):1937–1938.
30. Tan JC, Chan EW, Looi A. Bilateral orbital subperiosteal hemorrhage following labor. *Ophthalmic Plast Reconstr Surg*. 2011;27(3):59–63.
31. Riggs MJ, Coston NH, Teneyuque NR, Keyser EA. The eyes do not lie: a case of bilateral subconjunctival hemorrhages and orbital hematoma postpartum. *Mil Med*. 2019;184(1-2). e272–274.