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Major review

Nontraumatic orbital hemorrhage

*Alan A. McNab, MB, BS, FRANZCO, DMedSc**

Director, Orbital Plastic and Lacrimal Clinic, Royal Victorian Eye and Ear Hospital, Melbourne, Australia

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ABSTRACT

Nontraumatic orbital hemorrhage (NTOH) is uncommon. I summarize the published reports of NTOH and offer a classification based on anatomic and etiologic factors. Anatomic patterns of NTOH include diffuse intraorbital hemorrhage, "encysted" hemorrhage (hematic cyst), subperiosteal hemorrhage, hemorrhage in relation to extraocular muscles, and hemorrhage in relation to orbital floor implants. Etiologic factors include vascular malformations and lesions, increased venous pressure, bleeding disorders, infection and inflammation, and neoplastic and nonneoplastic orbital lesions. The majority of NTOH patients can be managed conservatively, but some will have visual compromise and may require operative intervention. Some will suffer permanent visual loss, but a large majority have a good visual outcome.

1. Introduction

Nontraumatic orbital hemorrhage (NTOH) is uncommon. There are a large number of single case reports and small case series of NTOH, but no systematic review of the subject. I have summarized the previously reported cases of NTOH and offer a classification system based on the anatomic pattern of hemorrhage and the etiologic factors involved.

2. Terminology

NTOH has been referred to by a number of terms, including spontaneous orbital hemorrhage, idiopathic orbital hemorrhage, orbital hematoma, and orbital hematic cyst. The term *nontraumatic orbital hemorrhage* seems to be the most logical to cover all types of bleeding in the orbit occurring in the absence of trauma. Many of these patients have underlying reasons for

* Corresponding author: Dr. Alan A. McNab, MB, BS, FRANZCO, DMedSc, Suite 216-218, 100 Victoria Parade, East Melbourne, 3002, Victoria, Australia.
E-mail address: amcnab@bigpond.com.
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the development of the hemorrhage, such as a vascular malformation, a sudden rise in cranial and orbital venous pressure, or a bleeding diathesis. Therefore, the term *spontaneous* is not appropriate.

The terminology hematic cyst has been used by a number of authors to describe a range of clinical entities. These include an apparently "encysted" hemorrhage (a localized hemorrhage surrounded by a nonepithelialized fibrous capsule) occurring with or without an underlying vascular malformation, nontraumatic subperiosteal orbital hemorrhage (NTSOH) and orbitofrontal cholesterol granuloma. The term *hematic cyst* should be confined to those intraorbital lesions characterized by a collection of blood and blood breakdown products within a cystlike structure without an epithelial lining and without an underlying vascular malformation. These are quite rare.

3. Historical aspects

In older textbooks, NTOH is described as largely being the result of hemorrhagic diatheses such as hemophilia or scurvy, or "congestion" with venous engorgement caused by thoracic compression, strangulation, violent coughing, or lifting heavy weights.[58]

The example of scurvy provides an interesting introduction to the subject of NTOH. Historically, scurvy has been responsible for the deaths of large numbers of sailors on long voyages from the start of the great Age of Discovery up to the early parts of the 19th century. The preventive role of foods with vitamin C or ascorbic acid became known in the late 18th century; however, scurvy still occurred in large armies on campaign and in groups such as polar explorers. Then, in the latter half of the 19th century in developed countries, infantile scurvy became a common condition as affluent parents fed their infants sterilized infant formula or boiled milk devoid of vitamin C.[160] Around this time, there are numerous references to exophthalmos in infantile scurvy.[51,228] By 1931, when Dunnington reported a case, he could find 22 previously published definite single cases of proptosis in childhood scurvy.[59] The American Pediatric Society conducted a large survey of its members' experience of infantile scurvy and published its report in 1898.[74] They described in detail 379 cases. In 110 of these cases, swelling or protrusion of the eyes was noted as absent, but was present in 49. This suggests that up to 31% (49/159) had orbital hemorrhage as a manifestation of their scurvy. Interestingly, orbital hemorrhage is known to be a rare manifestation of scurvy in adults, even though bleeding elsewhere is a common feature.

In 1905, Snow reported the post-mortem findings in an infant who died from scurvy with an orbital hemorrhage.[185] Dissection of the orbit showed that the bleeding had occurred into the subperiosteal space in the roof of the orbit. As we will see, NTSOH nearly always occurs in the superior orbit.

In reports of NTOH that preceded the advent of computed tomography (CT) or magnetic resonance imaging (MRI), it is often difficult to know what if any underlying cause existed in each case and where anatomically the hemorrhage occurred. Friedenwald, for example, described 2 cases in 1894 of "exophthalmus" owing to orbital hemorrhage.[62] One was traumatic and the other occurred spontaneously in a patient with possible liver disease and a clotting disorder. The anatomic site of the bleeding cannot be determined from the report. In 1912, Gruenig reported a case of "idiopathic hematoma of the orbit" that may have occurred in relation to an underlying vascular malformation, but there is no way of establishing this from the report.[76] Kundert described the case of a newborn infant with bilateral exophthalmos apparently the result of hemorrhage that resolved, but the site of the bleeding cannot be established from the description.[112] In 1951, Law described spontaneous orbital hemorrhage in 2 fit young men, possibly related to mild exertion, that could have been intraorbital or subperiosteal.[117] Both recovered fully.

With the availability of CT and MRI, patients with apparent NTOH can be imaged, and the clinical and imaging features of the majority of these patients allows a confident diagnosis of hemorrhage without the need for operative exploration.

4. Classification

NTOH can be classified on the basis of the anatomic site of the hemorrhage as well as the underlying cause. There are several distinct anatomic patterns of NTOH. In parallel with this, there are a number of underlying clinicopathologic causes of NTOH. Some anatomic variants, such as NTSOH, do not occur in the presence of all types of underlying clinicopathologic causes; similarly, some underlying causes do not lead to hemorrhage in all of the defined anatomic spaces. The classification is useful, however, in thinking about an individual patient presenting with NTOH.

The following anatomic patterns of NTOH are well-described:

1. Diffuse intraorbital
2. Localized intraorbital (hematic cyst)
3. Subperiosteal
4. Related to extraocular muscle (EOM)
5. Related to orbital floor implants.

Underlying clinicopathologic factors in the development of NTOH include:

1. Vascular lesions
2. Increased cranial venous pressure
3. Bleeding disorders
4. Infection
5. Inflammation
6. Neoplasms
7. Other orbital lesions.

In some cases, it may be impossible to assign an underlying cause to the NTOH and these patients can be truly termed idiopathic; however, with modern investigation and imaging, these patients are very rare.

5. Diffuse intraorbital hemorrhage

Diffuse intraorbital hemorrhage, or hemorrhage that is not localized to 1 anatomic site, may occur in several clinical