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Neuro-ophthalmology Update

Intracranial pressure and skull remodeling

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Abstract

In this article we review bony changes resulting from alterations in intracranial pressure (ICP) and the implications for ophthalmologists and the patients for whom we care. Before addressing ophthalmic implications, we will begin with a brief overview of bone remodeling. Bony changes seen with chronic intracranial hypotension and hypertension will be discussed. The primary objective of this review was to bring attention to bony changes seen with chronic intracranial hypotension. Intracranial hypotension skull remodeling can result in enophthalmos. In advanced disease enophthalmos develops to a degree that is truly disfiguring. The most common finding for which subjects are referred is ocular surface disease, related to loss of contact between the eyelids and the cornea. Other abnormalities seen include abnormal ocular motility and optic atrophy. Recognition of such changes is important to allow for diagnosis and treatment prior to advanced clinical deterioration. Routine radiographic assessment of bony changes may allow for the identification of patient with abnormal ICP prior to the development of clinically significant disease.

Keywords: Intracranial pressure, Hypotension, Hypertension, Bony changes

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Introduction

In this article we review bony changes resulting from alterations in intracranial pressure (ICP) and the implications for ophthalmologists. Before addressing ophthalmic implications, we will begin with a brief overview of bone remodeling. Two specific situations will then be addressed: (1) ophthalmic implications of bony changes seen with intracranial hypotension and (2) ophthalmic implications of bony changes seen with intracranial hypertension.

Dynamic bone

The maintenance of bone involves a dynamic process mediated by continual absorption by osteoclasts and creation of new bone by osteoblasts. (1) The balance of bone absorption and formation is mediated in part by mechanical loading or stress on the bone. The strain-sensitive cells are thought to be osteocytes. While this process is more active during childhood, even mature bone has the potential of remodeling. This can be loosely classified into three categories: (1) primary disease, (2) hormonally regulated changes in bone, and (3) alteration in stress or loading pressure. We are all familiar with more common primary diseases of bone, such as fibrous dysplasia, Paget's disease, and osteogenesis imperfect. With the exception of trauma, osteoporosis is the most common abnormality of bone. The cause of osteoporosis is multifactorial and likely is in part connected to age related hormonal changes (i.e. menopause). These examples only serve to illustrate the plasticity of bone.

We will now focus on the reshaping of bones in response to mechanical forces. Bony changes in response to alterations in ICP fall in this group. We are all familiar with bone

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remodeling in response to tumors. Bone may be infiltrated or destroyed by malignant neoplasm. Changes in bone due to malignant tumor infiltration are more complex than simple changes in mechanical loading. However, bony changes in response to benign neoplastic disease, molding and thinning of adjacent bone, are likely pressure related. Fig. 1 illustrates bony remodeling secondary to long standing pressure exerted by an expanding benign hemangioma. The loss of stress of daily activity (loss of loading) with simple disuse results in bony changes. For example, prolonged bed-rest has been shown to result in bone remodeling with a reduction in density.[2] In addition, previous work has shown that during prolonged bed rest, the skull will increase in mass, which is believed to result from a net bone formation from the increased ICP from a chronic rostral shift in fluid.[1] Astronauts experience a similar effect with extended periods of zero-gravity.[3–5] Long bones may resorb due to the lack of mechanical loading required to maintain bone density. Before specifically addressing the bony effects of altered ICP, we should focus on two related entities: silent sinus syndrome and pneumosinus dilatans.

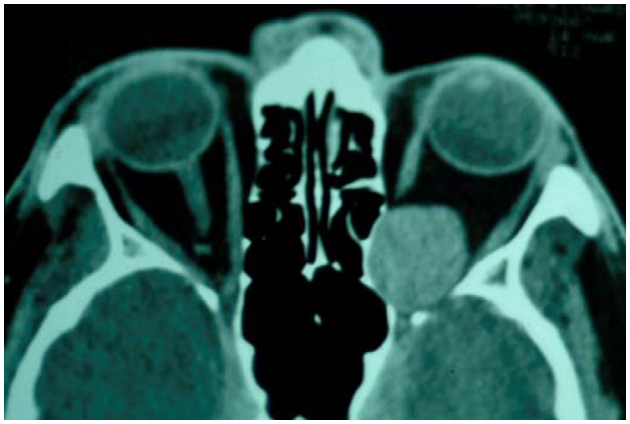

Figure 1. Axial computed tomography demonstrating bony remodeling secondary to a slowly enlarging benign orbital neoplasm.

Silent sinus syndrome

''Silent sinus syndrome'', first described in 1994 by Soparkar et al.[6] is characterized by spontaneous maxillary sinus atelectasis with orbital floor resorption, resulting in ipsilateral enophthalmos and hypoglobus (Fig. 2). Due to an absence of sinus disease symptoms, globe displacement is frequently the presenting sign, hence the name ''silent'' sinus syndrome. In rare cases ocular motility may also be affected.[7–9] Although patients of all ages can be affected, onset has been most commonly reported to occur in the fourth decade of life.[1,10–15] The mechanism of atelectasis in silent sinus syndrome has been postulated to be negative sinus pressure created by prolonged sinus hypoventilation due to outflow obstruction, and/or chronic inflammation with contraction of fibrous bands, resulting in distortion of the antral wall. Additionally, bone resorption occurs, further contributing to orbital floor displacement and altered globe position. The initial cause of outflow obstruction is not always clear; however, numerous etiologies have been implicated including a mucocele or polyp, lateralized middle turbinate, inspissated mucus, and intra-orbital ethmoidal (Haller) cells.[16,17] Although silent sinus syndrome has no direct relationship with ICP, it is relevant because the mechanism of an alteration in pressure gradient affecting an orbital wall with resulting enophthalmos is analogous to the process and consequences seen with intracranial hypotension.

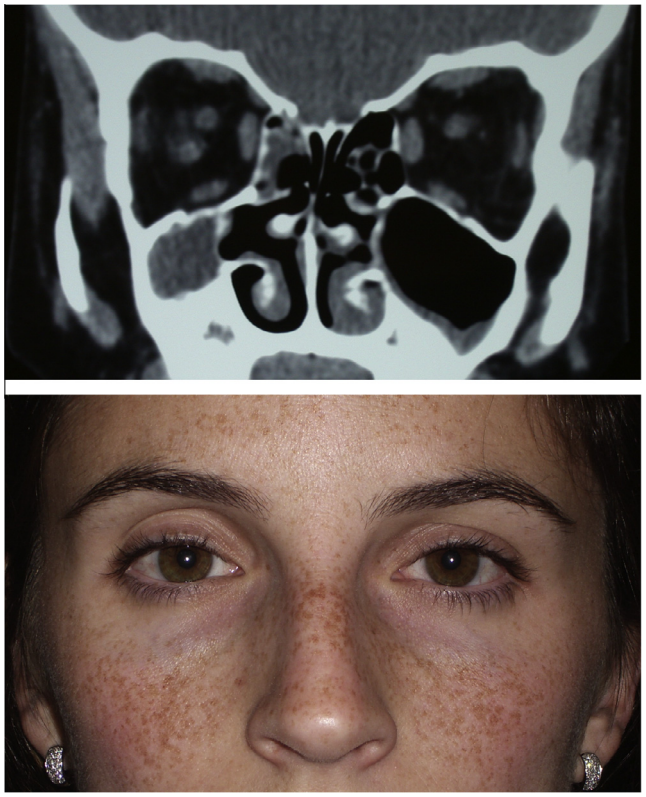

Figure 2. Silent sinus syndrome. Atelectasis of the maxillary sinus (top) results in downward bowing of the orbital floor resulting in enophthalmos (bottom).

Pneumosinus dilatans

Pneumosinus dilatans is a somewhat uncommonly used term that describes enlargement of the paranasal air sinuses. Sinus expansion has been described to occur without an identifiable cause, or in association with other abnormalities including fibro-osseous disease and meningiomas.[18,19] Presenting symptoms have included headache and ocular misalignment. Decreased visual acuity and field loss have been attributed to presumed optic nerve compression by an enlarged sphenoid sinus.[20] Many patients have been described as presenting with proptosis, whereas in others no mention of globe position was provided.[18–21] Of particular interest, in 1992 Schayck and Niedeggen described two patients with cerebral hemiatrophy who developed enlargement of the sphenoid, ethmoid and frontal sinuses.[21] They termed this pneumosinus dilatans, but noted that sinus enlargement occurred after cerebrospinal fluid (CSF) shunting. It seems likely that these cases represented intracranial hypotension related bone remodeling. Perhaps other cases labeled as pneumosinus dilatans also occurred secondary to unrecognized intracranial hypotension.