Headache and Diplopia after Rapid Maxillary Expansion: A Clue to Underdiagnosed Pseudotumor Cerebri Syndrome?

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Abstract

Rapid maxillary expansion (RME) is an orthodontic procedure that separates the two maxillary bones at the mid palatine suture level. RME is commonly used in the pediatric age group to reduce and/or eliminate a transverse maxillary deficiency. At our institution we followed up an 11-year-old adolescent who was diagnosed with a class III malocclusion and was treated by RME with the combined use of expansion appliances (i.e., Hyrax) in addition to maxillary protraction devices (i.e., Delaire facemask). Three months after the start of treatment, he complained of headache and double vision, and was admitted to our hospital. A funduscopic examination revealed papilledema. Magnetic resonance imaging of the brain showed intraocular protrusion of the optic nerve head and enlarged perioptic subarachnoid space. Cerebrospinal fluid opening pressure during lumbar puncture was elevated ($+370$ mm H_2O), confirming the diagnosis of the pseudotumor cerebri syndrome (PTCS). Removal of the maxillary expander and facemask led to the complete resolution of clinical symptoms in 1 week. The association of RME and PTCS-related manifestations was first reported by Timms in 1986, but, to the best of our knowledge, further cases have been not described in the medical literature. This overlooks PTCS as a potential RME-associated complication. We suggest that clinicians should carefully consider PTCS in all pediatric patients that complain of headache and/or visual disturbances during a treatment by RME. We also speculate on the possible changes of the cerebral venous circulation during RME, potentially leading to an impaired venous drainage that may cause increased intracranial pressure and PTCS.

Keywords

► rapid maxillary expansion
► idiopathic intracranial hypertension
► pseudotumor cerebri syndrome
► children
► adolescents

Introduction

Pseudotumor cerebri syndrome (PTCS) is defined by the presence of elevated intracranial pressure (ICP) in the setting of normal brain parenchyma and cerebrospinal fluid (CSF) composition. Presenting signs and symptoms are heterogeneous, but usually include headache, visual disturbances (i.e., vision loss or double vision), and papilledema at the

funduscopic examination.[1] Up to 90% of patients typically recover with appropriate medical and/or surgical treatments, although in a minority of cases this condition may lead to permanent visual loss.[2] The awareness of this potentially under-recognized condition is rising among pediatricians.[3]

The literature on childhood (i.e., prepubertal) PTCS has focused on many potential associated comorbidities, including endocrinopathies (e.g., adrenal insufficiency, primary and secondary aldosteronism, Cushing disease) and medical treatments (e.g., recombinant growth hormone therapy, thyroid supplementation, vitamin A intoxication or deficiency).[4–6] Conversely postpubertal PTCS is less heterogeneous in terms of etiologies, being more frequently associated with well-known risk factors such as obesity and female sex.[7,8] The rapid maxillary expansion (RME) is an orthodontic procedure widely used in the pediatric age group to eliminate a transverse maxillary deficiency (i.e., between the dental arches). The treatment induces the widening of the maxilla and leads to (1) the correction of posterior cross bites, (2) the coordination of the maxillary and mandibular dental arches, and (3) the gain in arch perimeter in patients with tooth-size/arch-size discrepancies.[9]

The dentoalveolar (and orthopedic) effects of RME are based on the separation of the mid-palatal and circumaxillary sutures.[10] One of the central pathologic features of class III malocclusion is represented by maxillary bone retraction; in this respect, RME (especially if associated with the orthopedic facial mask) can be regarded as one of the most effective treatments of class III anomalies.[10] Several clinical and histologic studies showed other side effects of RME (e.g., root and/or vestibular cortical reabsorption, pain, excessive tipping of anchorage teeth, transitory ocular diseases, microtrauma, and bite opening) in patients who underwent maxillary expansion.[11]

It has been suggested that this anatomical stress during maxillary expansion and RME on the craniofacial and/or maxillary structures has repercussions in the vascular compartment, consisting of cerebral hemodynamic changes, including increase in the blood volume and flow in the brain.[12] We hereby present the case of an 11-year-old Caucasian boy, referred to us for the treatment of a class III malocclusion with transverse maxillary deficiency, who underwent the application of an RME appliance (Hyrax type) and developed PTCS 3 months after starting the treatment. We now consider the possible role of impaired venous drainage, due to the complex effects of orthodontic procedure, in the rise of ICP.

Case Report

In 2010, an 11-year-old Caucasian boy was referred to undergo an orthodontic examination. His medical history and clinical and radiographic examinations excluded the presence of systemic diseases or congenital craniofacial syndromes. The orthodontic examination revealed the presence of a class III malocclusion with anterior and posterior cross bite, lower midline deviation, and transverse maxillary deficiency. There was neither relevant familial history pertaining to skeletal class III malocclusion nor any pertinent medical history. He presented a late mixed dentition. The

cephalometric analysis showed a sagittal skeletal class III jaw relationship (A point-nasion angle: -0,4).

Vertically he presented a hypodivergent skeletal pattern (Frankfort mandibular-plane angle: 20.6 degrees) and a dental compensation with upper incisors proclination (U1-palatal-plane: 120 degrees). The patient showed a 2-degree stage of cervical vertebral maturation, as assessed on lateral cephalograms (►Fig. 1).[5] Clinical history, moreover, revealed he had suffered from obstructive sleep apnea syndrome since childhood. The treatment objectives were to (1) improve the patient's profile, (2) improve the skeletal jaw relationship, (3) establish a molar class I relationship, (4) increase the maxillary arch width, and (5) correct the cross bite.

We therefore decided to use a maxillary expander, with a Hyrax appliance as a part of his comprehensive orthodontic treatment, banded on permanent teeth. The appliance screw was activated twice a day (i.e., 0.4 mm/day) for 18 days and, after 1 month, a Delaire-type facemask was also applied to support maxillary protraction. The patient was instructed to hold this appliance for 14 hours a day, for a period up to 18 months, using 5/16 in, and 14-oz extraoral elastics. Three month after the application of the Delaire device, the patient started to complain of headache and diplopia on a daily basis. Therefore, he was admitted to our pediatric unit and underwent ophthalmologic examination, which showed a bilateral papilledema. We reviewed his medical history and did not find any recent medical treatment or any drug. His weight and the body mass index were within normal limits. There was no history of infections. Blood clotting tests did not show any anomalies or hypercoagulability. An extensive diagnostic workup excluded any inflammatory and/or rheumatic diseases. The patient underwent magnetic resonance imaging (MRI) that showed intraocular protrusion of optic nerve head and enlarged perioptic subarachnoid space (►Fig. 2). The magnetic resonance angiography was reported as normal and excluded any degree of sinus venous stenosis and/or

Fig. 1 Radiographic examination: class III molar and canine malocclusion.
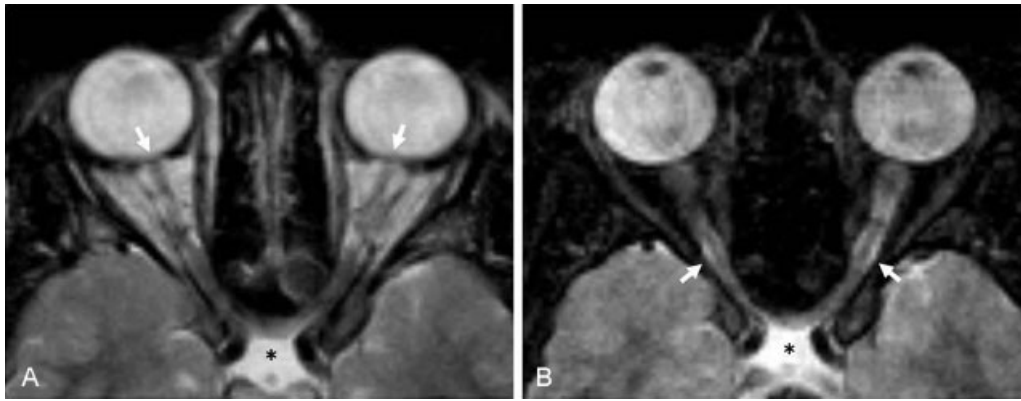
Fig. 2 Magnetic resonance imaging examination; axial T2-weighted fast spin echo images. (A) Intraocular protrusion of optic nerve head (white arrows); (B) enlarged perioptic subarachnoid space with cerebrospinal fluid hyperintensity surrounding the optic nerves (white arrows). The optic nerve sheath widening (A, B) is thought to coincide with papilledema. Partial empty sella has been shown in both images (black asterisks).

thrombosis. CSF pressure during lumbar puncture was elevated ($+370$ mm H_2O).

The boy was put on acetazolamide (20 mg/kg/day) with only partial resolution of headache and visual disturbances. We discussed with the family and orthodontic consultants about interrupting the RME procedure. After 1 week of the removal of the appliance, the headache and double vision completely disappeared and the funduscopic examination yielded a marked improvement of the papilledema. The patient continued the orthodontic procedure exclusively with the *fixed orthodontic appliance* (i.e., *multibracket appliances*) with no further neurologic and/or visual complications.

Discussion

Orthodontic researchers have tried to find the most optimal treatment for class III malocclusion patients. Many authors agree that greater skeletal benefits are achievable with the combined use of the expansion appliances (e.g., Hyrax) in addition to maxillary protraction devices (e.g., Delaire facemask), especially regarding the magnitude of the protraction effects on the maxillary structures.[10] This expansion/facemask therapy is particularly indicated in young class III malocclusion patients with anteroposterior and vertical maxillary deficiency on a background of early mixed dentition.[10] The benefits of the RME on the dimensions of the jaws, pharyngeal structures, and facial structures of patients in the pediatric age group have been studied by orthodontists and otorhinolaryngologists.[9]

RME (with various modifications) is now routinely used as a relatively simple and predictable orthodontic therapy to treat patients with maxillary constriction and posterior cross bites. All the most common side effects of placing maxillary expanders (e.g., pain, discomfort, impaired speech, and chewing and swallowing problems) are usually described as mild and transitory. Because of the various positive side effects, the number of indications for RME in children with constricted maxillary arches and various associated problems (e.g., obstruction sleep apnea syndrome, nocturnal enuresis, conductive hearing loss) is rising.[9] Notably, the maxillary bones are connected to neighboring bones by sutures, and the RME therapy in young patients produces transverse skeletal effects on the maxilla by opening the mid-palatal suture.[11] RME has limited local effects on the mid-palatal suture and the maxillary region. Several studies have focused on the potential RME-related complications on visceral structures and neurocranium.[13]

In fact, many studies based on histologic methods, radiologic imaging, and bone scintigraphy speculated that the forces derived from palatal expansion could spread to deeper anatomical structures. Circumaxillary sutures, as well as other structures not directly joined with the maxillary bones, may be affected by this strong orthopedic force. Different levels of stresses and bone displacement, and several complications following RME, have been recorded in clinical studies and animal models. These include fracture of the cranial base and opening of the sphenoid-occipital synchondrosis.[11,13]

High stresses were recorded in the zygomatic bone and mostly in the sphenoid bone, as well at the base of the pterygoid plates, in the superior orbital fissure, in the round foramen, in the oval foramen, in the spinous foramen, and in the optic foramen.[13] A higher risk of visual defects, including especially convergence defects (e.g., strabismus), as well as myopia and astigmatism, has been noted in children with malocclusions compared with the general pediatric population.[9,11] Although the primary effects of RME forces on craniofacial structures have been largely documented, the secondary effects of RME on the intracranial vascular compartment, likely influencing brain hemodynamics, are still unclear.[11,12]

According to previous studies,[13] the superior orbital fissure, the lacerated foramen, and the carotid sulcus are particularly stressed during RME. Close to these sutures are some important and vulnerable vessels that play essential roles in supplying blood to the brain. The stress focused on several foramina in the cranial base caused by RME may cause dilation of the nearby blood vessels. As a result of the vasodilator effect, manifested as increased cerebral blood volume (CBV) and cerebral blood flow (CBF), the blood and oxygen supply to the brain would also be increased; this provides a feasible explanation of the benefits reported by

enuretic children during RME.[13] In a study on rabbits,[12] perfusion computed tomography (P-CT) showed the total CBV significantly increased after RME. The CBF was increased only in the first phases of the RME treatment and then returned to normal, likely through the autoregulatory capacities of the brain.

Interestingly, the secondary obstruction of intracranial venous drainage likely occurring during early RME treatment, including constriction of the cerebral venous sinus, may impede the processes of CSF drainage and reabsorption.[2] This might possibly lead to raised ICP and PTCS. However, because pretreatment MRI was not performed, the possibility of preexistent congenital and/or acquired anomalies of the cerebral venous sinus (possibly exacerbated by the RME procedure) cannot be excluded. In the hereby reported boy, the removal of the orthodontic device led to the resolution of PTCS-related clinical signs and symptoms. As previously described by Timms,[14] children who underwent RME experienced headache and double vision in some cases. We suggest the careful consideration of PTCS in RME patients and the performance of all pertinent investigations (e.g., funduscopic examination, MRI, lumbar puncture) be given to start early treatment (e.g., diuretics) in order to avoid possible ophthalmologic complications. Lastly, the discontinuation of RME treatment may be sufficient to resolve headache and visual symptoms, as in the present case. Although a single case is insufficient evidence for a strong link between PTCS and RME, this report is meant to encourage further studies to fully ascertain the clinical implications of these observations.

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