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TECHNICAL NOTES

Sphenoid dysplasia in neurofibromatosis type 1: a new technique for repair

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Abstract
Purpose Sphenoid bone dysplasia in neurofibromatosis type 1 is characterized by progressive exophthalmos and facial disfiguration secondary to herniation of meningeal and cerebral structures. We describe a technique for reconstruction of the sphenoid defect apt at preventing or correcting the ocular globe dislocation.
Methods After placement of spinal cerebrospinal fluid drainage to reduce intracranial pressure, the temporal pole is posteriorly dislocated extradurally. The greater sphenoid wing defect is identified. A titanium mesh covered by lyophilized dura, modeled in a curved fashion, is interposed between the bone defect and the cerebro-meningeal structures with its convex surface over the retracted temporal pole.
Results The particular configuration of the titanium mesh allows a self-maintaining position due to the pressure exerted by the brain over its convex central part with its lateral margins consequently pushed and self-anchored against the medial and lateral walls of the temporal fossa. Screw fixation is not needed. The technique utilized in four cases proved to be reliable at the long-term clinical and neuroradiological controls (6 to 19 years).
Conclusion Sphenoid bone dysplasia in NF1, resulting in proptosis and exophthalmos, is usually progressive. It can be surgically repaired using a curved titanium mesh with the convexity faced to the temporal pole that is in the opposite fashion from all the techniques previously introduced. When utilized early in life, the technique can prevent the occurrence of the orbital and facial disfiguration.

Keywords Dysplasia · Neurofibromatosis · Technique

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Introduction

Sphenoid bone dysplasia occurs in 3–11% [1] of the subjects with neurofibromatosis type 1 NF1 patients. This malformation, a distinctive diagnostic feature for NF1, is characterized by progressive proptosis and facial disfiguration secondary to herniation of meningeal and cerebral structures. Unilateral in most of the cases, sphenoid bone dysplasia associated with plexiform neurofibroma in the territory of the trigeminal nerve in several cases and pooling of cerebrospinal fluid (CSF) in all cases often described in the past as an arachnoid cyst.

Surgery with reconstruction of the bone is the treatment of choice, and different methods have been proposed [4, 10–12]. We describe a technique for reconstruction of the sphenoid defect in pediatric age apt at preventing or correcting the ocular globe dislocation.

Surgical technique

For the operation, the patient is placed in a supine position with the head turned 45° controlaterally to the affected size. A lumbar CSF drainage is placed in order to decrease intracranial pressure, reduce volume of the CSF pooling at the temporal pole, and dampen CSF pulsation. The lumbar drainage is left in place for 7 days postoperatively. A curvilinear skin incision beyond the hairline is done to perform a small frontotemporal craniotomy in order to expose the posterior orbital wall and the dura of the temporal lobe. The temporal pole is then dislocated posteriorly through an extradural

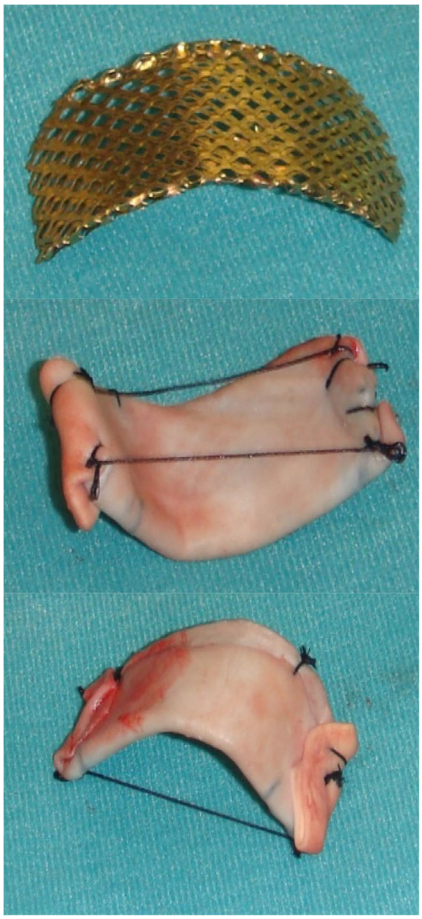

Fig. 1 Titanium mesh covered by lyophilized dura and modeled in a curved fashion. It is intraoperatively positioned between the bone defect and the cerebro-meningeal structures with its convex surface over the temporal pole

approach with particular attention paid during the detachment of the dura, which may be excessively thin as a result of its chronic distention. The defect in the sphenoid bone is then identified and reconstructed using a titanium mesh covered by lyophilized dura and modeled in a curved fashion (Fig. 1). This material is interposed between the bone defect and the cerebro-meningeal structures with its convex surface over the retracted temporal pole. The particular "reverse" configuration of the titanium mesh allows a self-maintaining position of the implant without screw fixation. This is due to the pressure exerted by the brain over its convex central part of the mesh with its lateral margins consequently pushed and self-anchored against the medial and lateral walls of the temporal fossa. After closure of the craniotomy, the scalp is sutured in layers.

The technique has been successfully utilized in four cases and proved to be reliable at the long-term clinical and neuroradiological controls (6 to 19 years) in preventing the progressive enlargement of the bone defect and the proptosis as well as in repairing an already established massive ocular globe dislocation (Figs. 2, 3, 4, and 5).

Discussion

Three main hypotheses have been taken into account to explain the malformation and its progression: a congenital malformation, an acquired bone defect, and the combination of multiple concurrent pathogenetic factors [1]. According to the congenital hypothesis, the sphenoid bone dysplasia would result from a mesodermal disorder of the sphenoid bone and associated maldevelopment of its ossification centers [2, 5]. Actually, subjects affected by NF1 are supposed to have an intrinsic bone tissue abnormality due to the downregulation of osteoblastic activity caused by the loss of neurofibromin [13]. Moreover, decreased bone density and low levels of 25-hydroxyvitamin D have been found in a large percentage of patients with NF1. However, the characteristic of the bone defect in this malformation is remarkably constant and does not coincide with the known ossification pattern of the sphenoid wing. Actually, the CT studies do not show new and various bone defects in the greater sphenoid wing but an enlargement of the superior orbital fissure [6]. In favor of the congenital hypothesis are, however, the invariably unilateral occurrence of the bone defect and the predominant involvement of the left sphenoid bone. The "acquired" theory ex-

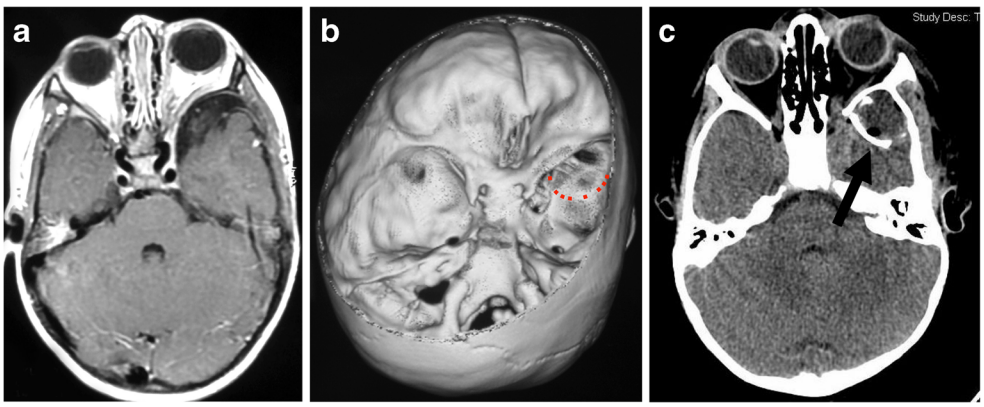

Fig. 2 Preoperative axial T1-weighted MRI with contrast (**a**) and CT three-dimensional reconstruction (**b**) of a child affected by sphenoid dysplasia on the *left side* with CSF pooling and proptosis. Postoperative CT scan (**c**) showing the "reverse" positioning of the titanium mesh. *MRI* magnetic resonance imaging, *CT* computed tomography, *CSF* cerebrospinal fluid

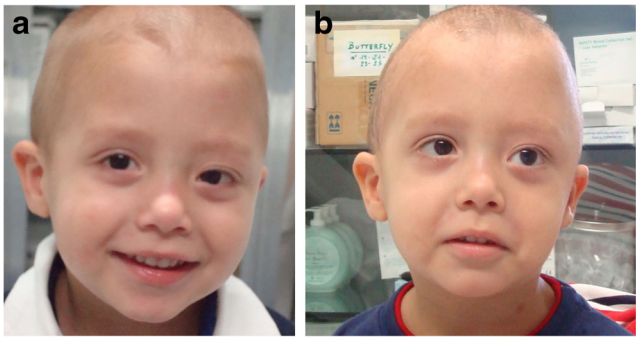

Fig. 3 Preoperative (**a**) and postoperative (**b**) picture of the patient treated for sphenoid dysplasia on the *left side* (radiological images in Fig. 2). The improvement of preoperative proptosis can be seen

plains the bone defect as the result of the erosion by neurofibromatous tissue that, at the orbit, may be very vascular fed by a rich network via the superior orbital fissure [9]. This theory finds its limit in the absence of neurofibromatous tissue in the orbit at the neurosurgical exploration in many cases. The multifactorial theory still takes into account a congenital defect of the sphenoid wing, resulting in an enlarged superior orbital fissure that postulates its progression as due to the chronic application of mechanical stress (CSF pulsation, secondary meningocele, CSF pooling, frequent association with plexiform neurofibroma, and dural ectasia in the territory of the trigeminal nerve). It is worth to note that the combination of dural mechanics changes, and CSF dynamics alteration has misled several authors to describe the occurrence of an anterior temporal arachnoid cyst as a significant component of the lesion [7, 8]. The goals of the surgical treatment vary according to the age of the surgical repair. The prevention of the progressive bone dysplasia and associated herniation of the cerebral structures as well as facial dislocation is the main aim of an "early" surgical repair. Goals of a "late" surgical correction, once the condition is fully expressed, are still not only to arrest the progression of the bone defect and further herniation of cerebral structures but also to correct the cosmetic deformity and to improve or reverse the functional impairment, possibly related to the stretching of the optic nerve. In

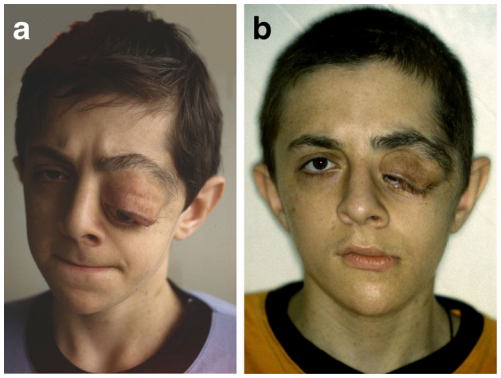

Fig. 5 Preoperative (**a**) and postoperative (**b**) picture of the patient treated for sphenoid dysplasia on the *left side* (radiological images in Fig. 4). The ocular globe and optic nerve functions have been preserved. The patient underwent successively plastic surgery treatment

such regard, it should be emphasized that, in almost all the cases, an adequate surgical correction may avoid the ocular globe enucleation and the sacrifice of the optic nerve.

The evidence of an evolving bone defect, as demonstrated by seriate neuroimaging studies, is an important reason to perform the treatment of sphenoid dysplasia in young age. It is obvious that an early treatment requires dealing with a smaller bone defect and with a minor distortion and stretching of the neural structures such as temporal lobe and optic nerve.

The goal of the surgery should be to reposition temporal lobe dura with careful dissection of the neighboring orbital structures, reconstruction of the defective orbital wall creating a diaphragm to avoid dislocation of the ocular globe, and transmission of cerebrospinal pulse transmission.

Various surgical techniques have been proposed for this pathology using bone graft, bone reinforced with titanium mesh, or alloplastic material alone.

Autologous bone in children, usually harvested from the calvarium split or rib, has the advantage to be biologically inert materials. On the other hand, complication at donor site, increased operative time, and most important reabsorption of the bone [3] are the disadvantages of this technique. A mixed implant (titanium mesh and iliac spongiosa) through a lateral

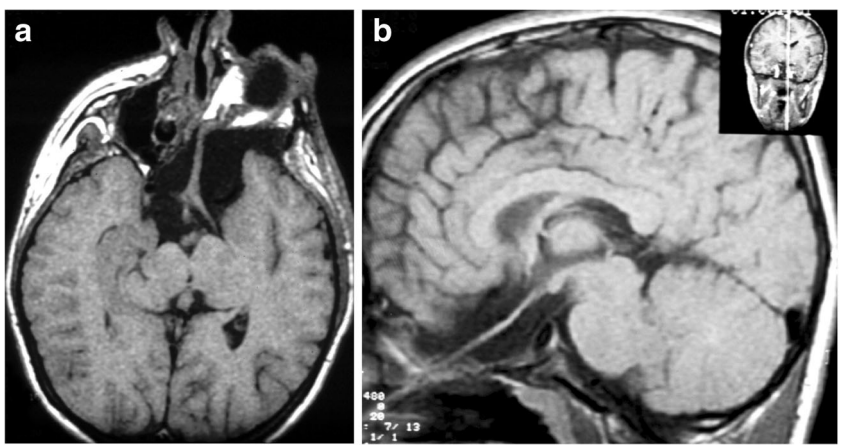

Fig. 4 Axial (**a**) and sagittal (**b**) T1-weighted MRI showing a case of left sphenoid dysplasia. The severe displacement of the ocular globe and stretching of the optic nerve is clearly depicted. *MRI* magnetic resonance imaging

orbitotomy has been also proposed, but it showed reabsorption of the bone after 6 months [4]. Recent reports show experiences using alloplastic materials such as titanium, demineralized bone, and polyethylene [10]: These are more ductile, avoid donor site morbidity, and reduce surgical time.

Our technique involves the use of a titanium mesh covered by lyophilized dura in order to avoid adherence between tissue and mesh or the passage of meningoencephalocele through its holes [10]. The unique feature is the orientation of the curved mesh with its convex surface over the retracted temporal pole with consequent self-anchorage of the lateral margins on the temporal fossa due to the pressure exerted by the brain over its convex central part. This avoid screw fixation and dislocation of the implant as observed in previous literature [4].

Conclusion

Sphenoid bone dysplasia in NF1, resulting in proptosis and exophthalmos, is usually progressive. It can be surgically repaired using a curved titanium mesh with the convexity faced to the temporal pole that is in the opposite fashion from all the techniques previously introduced. When utilized early in life, the technique can prevent the occurrence of the orbital and facial disfiguration.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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