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Surgical care burden in orbito-temporal neurofibromatosis: Multiple procedures and surgical care duration analysis in 47 consecutive adult patients

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ABSTRACT

Patients with orbito-temporal neurofibromatosis (OTNF) bear a heavy burden of surgical care. We studied 47 consecutive patients with OTNF from the French Neurofibromatosis 1 Referral Center cohort (n > 900), over a 15-year period to determine the clinical features most likely to predict repeat surgery and longer duration of surgical care. Forty-seven patients (5.2% of the NF1 patients' cohort) underwent 79 procedures with a 4.8 years average follow-up. Soft-tissue surgery had a high revision rate (19/45 patients), skeletal surgery did not (2/13 patients). Transosseous wire canthopexy and facial aesthetic unit remodeling were associated with stable outcome. Ptosis repair carried an unfavorable outcome, particularly in the presence of sphenoid dysplasia. Stable skeletal remodeling was achieved with polyethylene implants and/or cementoplasty. Multiple procedures were undertaken in 70% of patients and were predicted by the NF volume, canthopexy, skeletal dysplasia, or a Jackson's classification 2 and/or 3; but not by declining visual acuity. A classification based upon predictive risk of repeated procedures is proposed: Group 1: Isolated soft tissue infiltration not requiring levator palpebrae or canthal surgery; Group 2: Soft tissue involvement requiring ptosis repair or canthopexy, or NF great axis over 4.5 cm; Group 3: Presence of sphenoid dysplasia with pulsatile proptosis, regardless of visual acuity.

1. Introduction

Neurofibromatosis 1 (NF1) has an incidence of 1 in 3500 live births (Riccardi and Eichner, 1986). Orbito-temporal neurofibromas (OTNF) appear in infancy and affect 1% of NF1 patients (Moore, 1931; LeWald, 1933; Jackson et al., 1983; Poole, 1989), with composite skeletal and soft tissue factors causing unfavorable functional and aesthetic effect (Riccardi and Eichner, 1986; Huson et al., 1988). Primary dysplasia of the sphenoid wing (LeWald, 1933) is typical and occurs in 3–7% of NF1 patients (Zimmerman et al., 1983; Riccardi and Eichner, 1986), most severely presenting as a pulsatile proptosis precipitated by intra-orbital herniation of the temporal lobe. The cause of progressive orbital dysplasia is multifactorial (Binet et al., 1969; Friedrich et al., 2003) and NF developed from the trigeminal nerve (Riccardi and Eichner, 1986) in 5% of NF1 patients may contribute (Jacquemin et al., 2002; Friedrich et al., 2010, 2013). Transcranial repair of the OTNF orbit (Dandy, 1929) is facilitated by contemporary anesthesia, neurosurgical access, and implantology. Despite improvements in resectional planning of OTNF (Hivelin et al., 2010), the long-term results of soft-tissue restoration are often unstable (Lee et al., 2004; Wise et al.,

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2005; Erb et al., 2007) in the face of disease progression and its effect on the rheology of the skin and soft tissue (Mimoun et al., 2006).

Despite multidisciplinary coordinated care through NF1 reference centers, multiple procedures and hospitalizations increase the burdens and reduce patients' quality of life. Classification grading the severity of OTNF (Jackson et al., 1983), and quality of life scores related to skin disorders (Chren et al., 1996; Wolkenstein et al., 2001) have been described, however, the specific clinic features of OTNF which predict multiple interventions are not identified in the many published series (Van der Meulen et al., 1982; Jackson et al., 1983; Marchac, 1984; Poole, 1989; Jackson et al., 1993; Krastinova-Lolov and Hamza, 1996; Lee et al., 2004; Marchac and Britto, 2005; Altan-Yaycioglu and Hintschich, 2010; Rilliet et al., 2010; Chaudhry et al., 2012; Singhal et al., 2013). Anticipating the clinical stability of operative outcome for these features, and the resulting duration of surgical care will further inform patient counseling in OTNF surgery.

We analyzed the clinical features and operative sequence of a consecutive series of 47 OTNF patients operated over a 15-year period from the French NF1 Reference Center, to identify the predictors of repeated procedures and longer surgical care.

2. Materials and methods

2.1. Patients' data collection

A retrospective case series analysis was performed including all patients older than 16 years with OTNF operated in our department from 1995 to 2010, all part of the over 900 NF1 patients enrolled consecutively on the same period of time at the Neurofibromatosis French Referral Center of our university hospital. Patients presenting malignant tumor, isolated cutaneous NF involving the orbito-temporal region, and two patients transplanted (Lantieri et al., 2011) were excluded from the analysis. Clinical data, tumor characteristics with the great axis of the resected part of the neurofibroma (pathology reports, corresponding to the longest single dimension of the NF tumour as measured on the skin), type and number of surgical procedures, outcomes, and chronology of global management were retrieved from two prospectively held databases: Fusion® (4D, Clichy, France) for clinical, pathology and radiology reports and Ipop® (Cristalnet, CRIH, Grenoble, Frances) for preoperative data. We detailed the peri-orbital soft-tissue lesions as: eyelid infiltration, palpebral ptosis, canthal malposition; and the involvement of the bones as: greater wing of the sphenoid, 'orbital frame' or fronto-temporal dysplasia, and additionally the visual acuity in order to classify patients following Jackson (Jackson et al., 1993). 'Exophthalmos' and 'proptosis' are often used indifferently in the literature. Proptosis implies normal or abnormal orbit, with an intraorbital content anteriorly displacing the globe such as temporal lobe, or NF tissue (Clauser et al., 1998). 'Pulsating exophthalmos' is assimilated to a 'piston' movement. In 'exorbitism' the orbit is small and the normal contents are pushed forward (craniosynostosis) (Nada et al., 2010). In 'exophthalmos' the orbit is normal, but the contents of the orbit push out the globe. This last is usually use for thyroid eye disease (Clauser et al., 2012a,b).

2.2. Surgical details of OTNF management

After intra-tumoral resections of the orbito-temporal NF by facial aesthetic unit remodeling procedures (Hivelin et al., 2010), the surgical reconstruction potentially involved any or all of sphenoid wing repair and encephalo-meningocele correction, orbital floor reconstruction, canthopexies, and ptosis correction. The reduction of temporal lobe herniation, suppression of arachnoidal cyst and sphenoid wing repair with a Medpor® sheet were performed through a unilateral frontal craniotomy (Dandy, 1929; LeWald, 1933; Friedrich et al., 2003; Wu et al., 2008, Pessis, 2012; Niddam et al., 2014), and the orbital floor or walls were reconstructed through a trans-palpebral subciliary approach. Orbital reconstructions were undertaken with screwed HDPP (high-density porous polyethylene) implants (Medpor®, Stryker Craniomaxillofacial, Portage, Michigan, USA) (Pessis, 2012). The temporal 'hard' and 'soft' tissue components, that affect 88% NF1 patients with lateral upper lid infiltration (Friedrich et al., 2010), were treated with HDPP implant or methylmetacrylate (MMC) cement (Jackson et al., 1993) placed under the temporalis muscle or the resected/debulked NF. Superior orbital rim defects were repaired with screwed HDPP. Medial canthal tendon fixation was undertaken with non-absorbable suture to the periosteum or transnasal steel wiring (Poole, 1989) with a local approach (no coronal approach required (Marchac and Britto, 2005)). Lateral canthal reattachment was performed by transosseous non-absorbable suture or steel wiring. Tarsal strip, periosteal flap (Chaudhry et al., 2012; Singhal et al., 2013) or Montandon's canthoplasties (Montandon, 1978) were also used as required. We mainly used the levator aponeurosis plication technique for ptosis corrections (Clauser et al., 2006); otherwise, levators aponeurosis resection-suture (Marchac and Britto, 2005). Constant extensive postoperative swelling after ptosis surgery prevented any estimation of surgical result before 3 months, and was not considered as a complication.

2.3. Selected clinical cases

The cases presented in Figs. 1–4 illustrate the surgical care for various presentations of OTNF. The first clinical case (Fig. 1) illustrates the reconstruction of orbital walls and correction of meningo-encephalocele and exophthalmos with HDPP implant on a 34-year-old previously multiply operated male patient. He suffered a complete absence of the greater sphenoid wing and of the orbital floor, an associated meningo-encephalocele and exophthalmia with right eye blindness (Jackson grade 3). The postero-superior orbital roof and floor was reconstructed with HDPP implants (Medpor®) via a craniofacial approach. Medial and lateral canthopexies and ptosis correction using a trans-palpebral approach (levator en-bloc plication technique as levator advancement with sparing of the Müller muscle is unrealistic in such infiltrated upper eyelid) were performed during the same procedure. Revision was undertaken at 5 month to adjust the orbit floor level (removal of the polyethylene sheet), revise the transnasal canthopexy, and for levator plication (additional resection of NF). A third procedure, 7 month after consisted in a facial aesthetic unit remodeling. No recurrence of exophtalmia and skin sagging occurred at 4.5 years follow up. The second clinical case (Fig. 2) illustrates medial and lateral canthopexies combined to the filling of a temporal bone defect with cementoplasty. A 17-year-old male patient who previously underwent facial aesthetic remodeling for a facial plexiform NF presented with greater sphenoid wing absence and fronto-temporal dysplasia. The temporal bone defect was filled with methylmetacrylate and medial and lateral transosseous canthopexies were performed concomitantly. The results on the orbito-temporal area were stable at a four years follow-up while the sagging worsened on both upper and lower infiltrated eyelids. The blindness and strabismus on the right eye prevented the indication to treat further the palpebral ptosis. The third clinical case (Fig. 3) illustrates the reconstruction of superior orbital rim defect with HDPP implant (Medpor®) on a 27-year-old NF1 female patient presented a right temporo-orbital plexiform NF invading the upper eyelid, leading to a ptosis and canthal malposition, and