Re-emergence of surgery for dystonia

In the paper by Ford et al (this volume, pp 472–478), the authors describe the results of an open label, retrospective study of a selective denervation for severe cervical dystonia (torticollis) in 16 patients refractory to injections with botulinum toxin type A. Using functional capacity scales they concluded that six (37.5%) patients had “a moderate or complete return of normal neck function”. Despite some improvement in 12 of 14 (85.7%) patients on the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) dystonia rating scale applied to “blinded” ratings of videotaped examinations, the surgery failed to return patients to their occupations. The results of this study are comparable with those of Krauss et al who recently reported the effects of 70 intradural or extradural approaches in 46 patients with severe cervical dystonia. During a mean duration of follow up of 6.5 years, 21 (46%) of the patients reported excellent or marked improvement on a global outcome scale. There was no difference in the distribution of outcome when patients who still responded to botulinum toxin were compared with the non-responders. Using a modified TWSTRS scale, Krauss et al found significant improvements not only in the severity of dystonia, but in occupational and domestic work as well as in various activities of daily living.

The reports by Ford et al and Krauss et al are timely and alternative treatments are being sought. The estimated frequency of secondary unresponsiveness due to the development of immunoresistance is 3% to 10% of patients with cervical dystonia, depending primarily on the individual and cumulative dosages of botulinum toxin, frequency of administrations, and age at onset of dystonia. Once patients develop immunoresistance their therapeutic options are limited. Although drug therapy, such as anticholinergic drugs, baclofen, and muscle relaxants are sometimes helpful, they rarely provide a satisfactory relief. Alternative types of botulinum toxin, such as type B1 and type F, have been found safe and effective in patients who no longer respond to BTX type A, but these forms of BTX are still investigational and not readily available.

The papers by Ford et al and by Krauss et al draw attention to surgical cervical denervation as a re-emerging therapeutic modality in patients with medically intractable cervical dystonia. Various techniques have been used since surgical denervation for cervical dystonia was introduced over a century ago. The approaches used most often utilise intradural and extradural nerve sectioning techniques with the primary aim to selectively weaken the muscles that produce the dystonic posture, with preservation of normal muscle function and avoidance of side effects. Electromyographic guidance is helpful and a staged procedure may be necessary to produce optimal symptomatic and functional benefit. Until a few years ago, extradural sectioning of the upper cervical anterior nerve roots (rhizotomy) and the spinal accessory nerve roots was the most common surgical procedure for cervical dystonia. Extradural denervation procedures, involving sectioning of selected posterior primary divisions of the cervical nerve roots (ramieartomy), have been favoured more recently. The role of myotomies in the treatment of cervical dystonia has not been well defined. No studies have attempted to compare the different procedures and the selection of procedure is often based on the surgeons’ experience and preference.

As well as cervical denervation, other surgical approaches recently reported to benefit patients with dystonia include deactivation of globus pallidus by either stereotactic ablative lesioning or by a high frequency stimulation. We systematically evaluated the efficacy of both unilateral and bilateral stereotactic pallidotomy in eight patients with generalised dystonia. Six (75%) had a marked improvement in dystonic movements and in motor function. The severity of dystonia, as assessed by different dystonia rating scales, decreased by 60%. The findings from this and other pilot studies indicate that pallidotomy is a safe and effective treatment in medically refractory cases of generalised dystonia. Thus surgical denervation and stereotactic procedures are gaining increasing acceptance as treatments for patients with generalised dystonia who continue to be disabled despite optimal drug or botulinum toxin therapy.

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