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TREATING THE CHIEF COMPLAINT: HAND REJUVENATION FOR HIRAYAMA DISEASE

Hirayama disease, also known as monomelic amyotrophy, is a relatively benign motor neuron disease, characterized by insidious muscle wasting and weakness, unilateral or asymmetric restricted to the distal upper limb, followed by a quiescent phase after onset in the second to third decades.¹ Pathophysiology is uncertain and no treatment is available to restore weakness.² Although the degree of disability does not interfere with activities of daily living,^{2,3} the appearance of the hand is of major concern to the patient but is often minimized by clinicians concerned about the cause and the weakness. Here, we describe hand rejuvenation surgery, a procedure that restores youthful appearance to the hand, as a treatment of a patient's chief complaint in Hirayama disease.

Level of evidence is class IV: single observational study without controls.

Case report. Hirayama disease was diagnosed in a 36-year-old man with chief complaints of left hand atrophy and weakness which was noted soon after age 13 and did not progress after age 20. On examination, marked atrophy of left intrinsic hand muscles and forearm flexors was noted. Slight weakness was demonstrated in the C7, C8, and T1 myotomes. EMG disclosed a slightly active and chronic neurogenic pattern in the appropriate spinal segments. MRI revealed asymmetric anterior flattening and atrophy of midlower cervical cord. He consulted several neurologists who recommended occupational therapy with no benefit. Because of his great concern about hand appearance, "rejuvenation" hand surgery was recommended by a plastic surgeon.

Hand rejuvenation procedure. This is a combination of 2 procedures. In the first procedure, infraumbilical fat was harvested using a liposuction cannula. The fat was then washed and refined. A total of 20 mL of fat parcels were serially injected into the first through fourth interosseous subcutaneous spaces with multiple passes at 0.25 mL increments. Fat molding was performed by gentle digital manipulation to achieve a uniform distribution. After structural fat grafting, the patient returned for moulage,

the process of forming the shape and volume of the prosthesis using dental alginate to generate the permanent prosthesis out of the silicone rubber. In the second procedure, an incision was made through the skin and subcutaneous fat, the fascia was opened and separated from the muscle belly, which was severely atrophic. Soft silicone fillers were placed into anatomic positions of the first and fifth interosseous fascial compartments. Pressure dressing was applied after the skin was closed. The patient tolerated the procedures well. Tissue edema was the only postoperative sequela but gradually resolved. The patient immediately recognized a major improvement of his hand appearance (figure). Gradual loss of hand fullness secondary to partial graft resorption is noted 5 years after the procedures; nevertheless, the patient stated that he would have had this procedure performed again.

Discussion. Hand atrophy is a common chief complaint of patients with Hirayama disease once their weakness has reached the quiescent phase.² Because the clinical course of the disease is relatively benign, the concern of hand appearance expressed by young patients is often neglected by neurologists.

Hand rejuvenation is a commonly performed procedure not only in elderly hands,^{4,5} but also in patients with ulnar nerve palsies who have major first web space atrophy.⁶ Among the fillers used, structural fat grafting may be the safest and the most effective filler in rejuvenation surgery.⁵ The procedure utilizes fat from the patient's abdomen or thigh which, after refining of the fat parcels, is placed in the dorsum of the hand. However, the extent and mechanisms of fat graft survival remain unclear.⁷ The result of the procedure is operator dependent and relies on the technique used. The main obstacle has always been fat resorption (the dissolution of adipose tissue) leading to unpredictable long-term results.^{4,6} Since edema is the most consistent postoperative sequela associated with fat grafting, it is sensible to re-evaluate patients at 16 weeks and again at 6 months after surgery to distinguish between tissue edema vs fat resorption.⁵ Only a few minor complications have been reported from large case series. These include an irregularity due to uneven fat distribution,^{4,5} a burn-

(A) The preoperative Hirayama hand with fully abducted left thumb and index finger. Note that the first dorsal interosseous is the most atrophic muscle. (B) Lateral view of the left hand shows significant atrophy of the intrinsic hand and adductor digiti minimi muscles. (C) A close-up view of the normal hand (right) and the Hirayama hand (left). The marker identified the area that was planned for structural fat grafting. (D) Moulage was molded using dental alginate to generate the permanent prosthesis out of the soft silicone rubber. (E, F) Six months after hand rejuvenation surgery, the fullness of the first web space and intrinsic hand muscles in the left hand appears similar to the normal hand (right). Note that the texture of skin and superficial veins remains natural looking.

ing sensation at the operative site,⁶ and skin infection requiring antibiotic treatment.⁴ Patients express uniform satisfaction with the results.^{4,6}

Although Hirayama disease has primarily been reported in Japan and India, the proliferation of cases in Western countries creates a global problem.⁷ Rejuvenation hand surgery with structural fat grafting is safe, natural-appearing, and can be repeated after

partial fat resorption. Such cosmesis of hand atrophy may benefit a symptom of patients with Hirayama disease and other neurologic conditions with hand muscle atrophy.

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1. Hirayama K, Toyokura Y, Tsubaki T. Juvenile muscular atrophy of unilateral upper extremity: a new clinical entity. *Seishin Shinkeigaku Zasshi* 1959;61:2190–2197.
2. Gourie-Devi M, Nalini A. Long-term follow-up of 44 patients with brachial monomelic amyotrophy. *Acta Neurol Scand* 2003;107:215–220.
3. Tashiro K, Kikuchi S, Itoyama Y, et al. Nationwide survey of juvenile muscular atrophy of distal upper extremity (Hirayama disease) in Japan. *Amyotroph Lateral Scler* 2006;7:38–45.
4. Aboudib Junior JH, de Castro CC, Gradel J. Hand rejuvenescence by fat filling. *Ann Plast Surg* 1992;28:559–564.
5. Coleman SR. Hand rejuvenation with structural fat grafting. *Plast Reconstr Surg* 2002;110:1731–1744.
6. Ghobadi F, Zangeneh M, Massoud BJ. Free fat autotransplantation for the cosmetic treatment of first web space atrophy. *Ann Plast Surg* 1995;35:197–200.
7. de Carvalho M, Swash M. Monomelic neurogenic syndromes: a prospective study. *J Neurol Sci* 2007;263:26–34.