The Laryngoscope
Lippincott–Raven Publishers, Philadelphia

Reflex Sympathetic Dystrophy of the Face: Current Treatment Recommendations

Richard L. Arden, MD; Samer J. Bahu, MD; Marcos A. Zuazu, MD; Ramon Berguer, MD, PhD

Reflex sympathetic dystrophy (RSD) of the face is an infrequently reported clinical pain syndrome characterized by dysesthesia, hyperalgia, hyperpathia, and allodynia. Treatment strategies, extrapolated from RSD and causalgia of the extremities, remain variable and poorly defined. Sympathetic blockade is generally the diagnostic and therapeutic treatment of choice; however, the frequency, timing, and duration of injections; need for neurolytic blocks; and role of sympathectomy are not well understood. The objectives of this report are to highlight the clinical behavior of facial RSD and contrast its essential differences from extremity RSD in response to standard treatment regimes. The case studies of two patients with this syndrome, following vascular surgery in the neck, are retrospectively reviewed with existent reported cases. Age, gender, etiology, symptoms, onset, triggers, and examination findings; timing, duration, and method of treatment; and outcome are summarized, forming the database for this study. Findings demonstrate an infrequent association of vasomotor and sudomotor changes with facial RSD, and lack of progression to a dystrophic or an atrophic stage, in contrast to extremity RSD. Furthermore, treatment response to sympathetic blockade is durable and less critically dependent on timing. The authors conclude that facial RSD has a favorable prognosis and should be managed conservatively with nonneurolytic stellate ganglion blocks, even when initiated as a delayed and repetitive injection series.

***Laryngoscope*, 108:437–442 1998**

INTRODUCTION

Sympathetically mediated pain syndromes include causalgia and reflex sympathetic dystrophy (RSD), which fundamentally differ in their precipitating event, histopathologic correlate, and potential for clinical progression. Initially described by Mitchell et al.[1] in 1864, causalgia is a syndrome of sustained, burning pain following partial, incomplete nerve damage, typically following a high-velocity missile injury to a major peripheral nerve. The level of injury is characteristically confined to nerve lesions above the elbow and knee (most commonly median and sciatic nerves) and accounts for 2% to 5% of peripheral nerve injury cases.[2] The more severe and proximate the neural lesion to the spinal ganglia, the greater the degree of causalgic response. The burning pain (dysesthesia) commonly appears immediately or soon after the injury and is spontaneous, continuous, and felt superficially in the hand or foot. For most patients (75%) the pain gradually subsides within 1 year, but during recovery they often experience lowered pain thresholds (hyperesthesia), elevated thresholds to touch, and overreactions/aftersensations to stimuli (hyperpathia). Exacerbating factors have been associated with dependent posturing, mechanical or thermal stimuli (allodynia), muscular activity, and disturbances in the sensory neuronal pool (i.e., auditory, visual, somatosensory, emotional stress). Treatment responses to neuronal blockade, truncectomy, or rhizotomy have been poor or unsuccessful in most cases, in contrast to sympathetic block (or sympathectomy), which has provided significant temporary (occasionally permanent) pain relief.

Reflex sympathetic dystrophy is a term first used by Evans in 1947 to describe a pain syndrome following various types of mild injuries in the absence of demonstrable nerve damage.[3] Unlike the rapid and violent neural deformation associated with causalgia, precipitating events in RSD have included fractures (>50%), lacerations, infections, operations, angina/myocardial infarction, peripheral vascular disease, degenerative joint disease, and injuries to muscles, ligaments, or soft tissue. In 10% to 26% of cases, no precipitating factor can be found.[4] Similar to causalgia, RSD pain possesses the components of dysesthesia, hyperesthesia, and hyperpathia, which seem to follow the topography of the sympathetically innervated vascular system rather than a true radicular or dermatomal pattern. In contrast to most vascular or neuropathic pain disorders, symptoms of sympathetic dystrophies may occur with or without precipitating activity (i.e., physical and emotional stimuli). Unlike causalgia, whose symptoms are typically self-limiting over time, RSD is a dynamic, progressive illness that may manifest a wide spectrum of signs and symptoms. To further

From the Departments of Otolaryngology—Head and Neck Surgery (R.L.A., S.J.B.) and Anesthesiology (M.A.Z.), and the Division of Vascular Surgery (R.B.), Harper-Grace Hospitals, Wayne State University School of Medicine, Detroit, Michigan.

Editor's Note: This Manuscript was accepted for publication June 24, 1997.

Send Reprint Requests to Richard L. Arden, MD, Department of Otolaryngology—Head and Neck Surgery, Wayne State University, 4201 St. Antoine, UHC-5E, Detroit, MI 48201, U.S.A.

characterize the clinical behavior of this entity in the facial region, we present the cases of two patients with RSD following distal vertebral artery bypass who were diagnosed and successfully treated with stellate ganglion blocks.

MATERIALS AND METHODS

A MEDLINE search was used to identify all cases of well-documented facial causalgia or RSD reported in the English literature. Eight cases were identified (between 1947 and 1995), to which were added our own two cases of facial RSD, both occurring after vascular intervention in the neck. For comparative purposes, tables were constructed documenting age, gender, symptoms, onset, provocative triggers, examination findings, treatment times, methods employed, and treatment outcomes. Literature review of the characteristics of sympathetically mediated pain syndromes affecting the extremities was used to form comparisons with the tabulated facial RSD series.

CASE REPORTS

Case 1

A 69-year-old man (K.S.) underwent transposition of the right external carotid to the right distal vertebral artery (VA) at the level of C1 for microembolic syndrome affecting the vertebrobasilar territory. A 95% diffuse stenosis of the entire first section of the VA precluded its proximal transposition to the carotid system. Two years earlier, an uneventful left proximal vertebral-carotid bypass had been performed. One month after his distal VA bypass, complaints of ipsilateral facial pain and swelling led to an antibiotic course for presumptive parotitis. A directed history revealed daily perceptions of exquisite sharp pain (visual analog scale [VAS], 8 to 9) diffusely involving the cheek, hemimandible, and ipsilateral tongue. The episodes were triggered by hot and cold liquids and, especially, by citric and sour-tasting foods that resulted in hypersalivatory responses. Light contact with an electric shaver also elicited sharp pains. Analgesic support with codeine derivatives was largely ineffective. Physical findings included marked hyperesthesia over the lower cheek and jaw area with mild loss of light touch. No alteration in skin temperature, edema, rubor, or diaphoresis was found.

With a working diagnosis of RSD, the patient consented to a trial of right-side stellate ganglion blocks with 10 mL of 0.25% bupivacaine using the classic anterior paratracheal approach to the C6 tubercle (Figs. 1 and 2). These were completed without side effects and with good pain relief that extended beyond the anesthetic duration. The VAS decreased approximately 50% after three or four blocks. After the sixth block with bupivacaine, the patient underwent a phenol stellate ganglion injection (2 mL 7.5% phenol in water) with a 40% VAS reduction from the initial complaints. At this point the patient described having no problems shaving and denied tenderness when touching his face. Although the frequency and intensity of painful episodes had decreased, the patient still experienced sharp pains in the same distribution, now with a VAS of 4 or 5. This persuaded the patient to consent to a series of three weekly, left-side stellate ganglion blocks with 10 mL of 0.25% bupivacaine with additional decrease in frequency and severity of attacks. Retesting with additional right-side stellate blocks provided no increased pain relief. Following additional left-side stellate ganglion blocks, the VAS had decreased approximately 70% from his initial pain symptoms 6 months earlier with good control of his excessive salivation problems. The last two left-side stellate ganglion blocks were subsequently given at monthly intervals at the end of which time VAS was reduced to 80% to 85% of initial levels. At this time (3 years of follow-up) the patient reports minimal pain attacks, no pain while shaving, and tolerance for all foods (including citric foods and hot or cold liquids) without any exacerbations of pain.

Case 2

A 69 year-old man (F.M.) underwent a right external carotid to right distal VA transposition at C1 to C2 for symptomatic dissection of the VA unresponsive to anticoagulation. Several weeks following surgery, he complained of right-side facial pain along the jaw from the temporomandibular joint forward, but the pain did not cross the midline. The pain was described as very sharp with a burning hot quality and was triggered by biting and chewing, sour foods, and cold stimuli. Other associated symptoms included mild tongue claudication, excess salivation, and cutaneous hypersensitivity. There was no history of facial flushing, edema, or hyperhidrosis. Physical examination was largely unremarkable except for the facial hyperesthesia.

With a presumptive diagnosis of RSD of the face, the patient underwent a series of right-side stellate ganglion blocks with 10 mL of 0.25% bupivacaine using the classic anterior paratracheal approach to the C6 tubercle. A total of five blocks were given at weekly intervals, resulting in decreased pain (50% to 70% of initial levels) with considerable lessening of the frequency of painful episodes. The patient's problems with excessive salivation were also very much in control. The patient was satisfied with the overall outcome and regarded his remaining symptoms as tolerable. This condition has remained stable at 2½ years of follow-up.

DISCUSSION

The diagnosis of RSD is usually made clinically, since there is no investigative tool that has been proven sensitive and specific enough. A presumptive diagnosis is made when presentation includes burning pain, hyperesthesia, allodynia, and vasomotor abnormalities and is supported by pain relief following sympathetic blockade of the affected area. Considering a complete, spontaneous remission rate of <5%,[3] the establishment of early signs and symptoms to allow for early treatment offers the best chance for long-term control before "sympathetic release." However, the unknown pathophysiology of RSD, lack of clear definition of the syndrome, and variable treatment responses have challenged physicians to better understand this disabling illness.

Observations derived from reported clinical data[5–9] in cases of facial RSD share a common denominator of cutaneous hyperesthesia, hyperpathia, and allodynia that is typically resistant to analgesic support (Tables I and II). While sharing these findings with extremity RSD, the classic associated vasomotor, sudomotor, and trophic changes frequently seen with the latter are less frequently found with facial RSD, often resulting in costly workups and delays in definitive management. The available literature on facial RSD fails to provide evidence to suggest its evolution into a dystrophic or atrophic stage. Clearly, when the aforementioned constellation of symptoms follows (typically within 1 month) surgical dissection, blunt or penetrating trauma, or a dental procedure, diagnostic stellate ganglion block is advocated early. In a study of 71 patients with RSD of the lower extremities, Wang et al.[10] noted sustained improvements in 70% of patients over the 3-year study period when treatment was initiated within 6 months of onset, but the percentage deteriorated to 50% when treatment was delayed 6 to 12 months. Despite these findings, review of the facial sympathetically mediated pain series demonstrates a more favorable prognosis even when definitive treatment is de-

TABLE I.
Reported Facial Causalgia/Reflex Sympathetic Dystrophy Summary.

Report (year)	Age (y)/Sex	Etiology	Symptoms	Onset	Triggers	Physical Findings
Bingham[5] (1947)	28/M	Penetrating shell fragment (R) cheek lodged near foramen oval	Continuous burning pain V_{1-3}	Immediate	Heat, jaw movements	Hyperesthesia, V_{1-3}
	23/M	Mortar wounds (L) cheek	Mild-moderate continuous burning pain V_2	3 Mo after injury	Heat, jaw movements, emotional upset	Hyperesthesia cheek skin
Hanowell and Kennedy[6] (1979)	59/M	Total laryngectomy, glossectomy, (L) radical neck dissection, segmental mandibulectomy for tongue cancer	Continuous burning pain, phantom tongue; intermittent stabbing pain (L) mandibular area	Perioperative	Hot and cold stimuli	Induration, hyperemia (L) face and submaxillary area; hyperesthesia (L) lower face; medial trigger point mandibular remnant
Khoury et al.[7] (1980)	60/M	(L) maxillectomy for CA	Severe burning pain (L) upper eyelid, nose, face, upper lip	1 Mo after surgery	Light touch, cold air or water, mastication	Skin erythema and edema, hyperesthesia supra/infra orbital nerve distribution, coolness, (L) facial skin
Jaeger et al.[8] (1986)	33/F	(L) upper molar extraction	Intense, constant (L) facial burning; "pinching" sensation (L) eyebrow and mandible, photophobia left eye	After extraction	Cold air/liquids, mastication, smiling, light touch	Slight facial swelling, slight increase in skin temperature trismus, cheek hyperesthesia
	31/M	Subtotal resection of frontal sinus osteoma	Constant burning across forehead radiating to orbits and maxilla	Immediate	Not specified	Tender suprabrow scar, marked hyperesthesia forehead; mild loss of pinprick/light touch sensation
Veldman and Jacobs[9] (1994)	47/F	Motor vehicle accident with zygomatic arch impaction and orbital floor fractures	Heavy, dull pain (R) head and face	Several days after surgery	Sound, facial animation	Hyperesthesia (R) face, mild facial paresis; slight swelling, erythema, warmth, hyperhidrosis
Saxen et al. (1995)	32/F	(L) upper molar extractions	Constant burning pain (L) infraorbital skin	1 Mo after extraction	Hot and cold stimuli	2 x 2 Erythematous infraorbital patch; hyperesthesia cheek
Arden et al. (1998)	69/M	(R) external carotid to distal vertebral artery transposition	Diffuse sharp pain (R) cheek, jaw, tongue; excessive salivation	1 Mo after surgery	Hot and cold liquids, light touch, citric substances	Hyperesthesia lower face, mild loss of light touch sensation
	69/M	(R) external carotid to distal vertebral artery transposition	Intense burning pain (R) cheek, jaw; excessive salivation; mild tongue claudication	Several weeks after surgery	Mastication, cold stimuli citric substances	Hyperesthesia lower face

TABLE II.
Treatment and Outcome Summary.

Case #	Report (year)	Treatment Initiated	Method	Outcome
1	Bingham[5] (1947)	13 mo after injury	Single (R) SG block, procaine/alcohol	Recurrence facial/pain tenderness at 3 w
		14 mo after injury	Cervical sympathectomy	Pain-free at 3 mo follow-up
2	Bingham[5] (1947)	11 mo after injury	Single (L) SG block, procaine/alcohol	Recurrence mild pain/hyperesthesia at 2 mo, severe at 9 months
		20 mo after injury	Cervical sympathectomy	Symptom resolution; no follow-up
3	Hanowell and Kennedy[6] (1979)	7 mo after surgery	Diagnostic (L) SG block, bupivicaine Alternate day, 5 block series	60% improvement at 2 d Pain-free at 3 mo follow-up
4	Khoury et al.[7](1980)	7 y after surgery	Diagnostic (L) SG block, bupivicaine	Pain relief for 6 h
			20 block series	75% improvement after last injection
5	Jaeger et al.[8](1986)	1 y after extraction	Diagnostic (L) SG block (local anesthetic unspecified)	Relief beyond anesthetic duration
			15 block series	Pain-free at 15-mo follow-up
6	Jaeger et al.[8] (1986)	3 y after surgery	Diagnostic bilateral SG blocks (local anesthetic unspecified)	Near-complete facial pain relief
			Bilateral morphine sulfate SG blocks (number unspecified)	66% improvement facial pain; persistent dysesthetic scar pain
7	Veldman and Jacobs[9] (1994)	1 y after surgery	*N*-acetylcysteine, 600 mg tid	Partial decrease facial pain; decreased size red, swollen, warm areas
8	Saxen et al. (1995)	10 y after extraction	Diagnostic (L) SG block, bupivicaine therapeutic (L) SG block	Pain relief for 24 h
			Clonidine, 0.1 mg bid	Not specified
			Responded well; follow-up not specified	
9	Arden et al. (1998)	6 w after surgery	Diagnostic (R) SG block, bupivicaine	Relief beyond anesthetic duration
			Weekly, 6 (R) SG block series × 1.5 (R) SG block, phenol	40–50% improvement facial pain
			3 weekly, (L) SG blocks	No change from baseline
			5 monthly, (L) SG blocks	60% improvement in pain
				70% improvement at 6 mo, 80%–85% improvement at 8 mo
10	Arden et al. (1998)	1 mo after surgery	Diagnostic (R) SG block, bupivicaine	Relief beyond anesthetic duration
			Weekly, 5 (R) SG blocks × 1.5 mo	50%–70% reduction in facial pain

SG = stellate ganglion.

layed. Of the six patients in which treatment was initiated 1 year or more after symptom presentation, sympathetic blockade resulted in cure in three (over follow-up period), marked improvement in two, and partial improvement in the one patient treated with *N*-acetylcysteine (Table II). Although the literature would suggest that up to 95% of patients with extremity RSD, and up to 50% of patients with causalgia, respond favorably to sympathetic blockade,[3] this differentiation in the facial region cannot be made at present because of the limited number of patients.

The number of sympathetic blocks that should be given and the timing of surgical sympathectomy remain unclear. A common practice for extremity RSD has been to limit the number of injections to three or four before considering sympathectomy, assuming the patient achieves at least a temporary favorable response.[11,12] It would appear, at least for cases of facial RSD, that an argument can be made for repeated injections over time with the expectations of favorable lasting outcomes. Of the reported cases where this approach was implemented (case reports 3 to 5, 9, and 10), an average of 13 stellate ganglion blocks were performed (range, 6 to 21) that achieved marked improvement or pain-free status over the follow-up period. Recognizing the relative invasiveness of sympathectomy compared with sympathetic blockade, the fact that sympathectomy does not guarantee a solution to the problem, and the reported favorable experience of stellate ganglion injections for facial RSD, the need for superior cervical ganglionectomy is questioned. Although based on an anecdotal case (case report 9), there may be a role for contralateral stellate ganglion blocks when symptomatic improvement with ipsilateral blocks plateaus. Although the mechanism for this idiosyncratic response is unclear, observations of crossed lateral radiation responses (particularly of the hand) favorably influenced by ipsilateral injections have been documented.[13] The need for a neurolytic injection following a positive diagnostic block is also questioned in lieu of progression (case reports 1 and 2) or failure to improve symptoms (case report 9) in the three cases in which it was employed.

In our own two cases of facial RSD following distal VA bypass, it is interesting to note that symptomatic onset, provocative triggers, and examination findings were almost identical. The absence of Horner's syndrome, V_3-distributed hyperpathia, hypersalivation, and hypersensitiv-

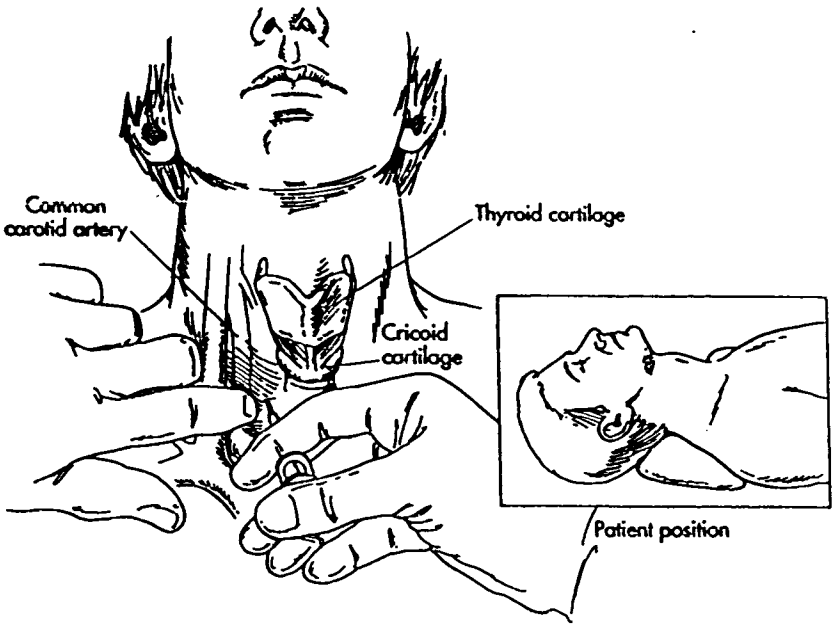

Fig. 1. Classic anterior approach to C6 tubercle for stellate ganglion block. A 23-gauge, 4- to 5-cm needle enters the skin perpendicularly to a depth of 2.0 to 2.5 cm while the carotid artery is retracted laterally. After contacting the tubercle approximately 3.0 cm cephalad to sternoclavicular joint and 1.5 cm lateral to midline), the needle is withdrawn 2 to 5 mm and an initial 1-mL test dose of 0.25% bupivicaine given after a negative aspiration. Subsequently, a 4-mL injection of the anesthetic solution blocks the stellate ganglion. A shoulder roll extends the neck, brings the esophagus midline, and facilitates palpation of the C6 tubercle. (From Raj PP, ed. *Practical Management of Pain.* edn 2. St. Louis: Mosby–Year Book; 1992; with permission.)

ity to thermal and chemical triggers is best explained by injury to postganglionic sympathetic fibers distributed along the external carotid artery plexus. In both cases, to achieve tension-free transposition of the external carotid, it was necessary to ligate its branches and skeletonize the artery above and below the hypoglossal nerve. Since sympathetic innervation to the facial skin, as well as the submandibular ganglion, is derived from the external carotid artery plexus, adventitial trauma or irritation at this location would be consistent with the clinical findings. Unlike preganglionic sympathetic fibers, which do not correspond to a sensory segment, postganglionic fibers are distributed to approximately the same cutaneous areas as that supplied with sensory fibers by the corresponding radicular nerve (i.e., oral tongue-jaw-V_3 dermatome).[14] Considering that 71% of resting salivation is provided by the submandibular glands,[15] with the observation that copious secretions from this gland can be elicited in response to sympathetically stimulated alterations in vasomotor tone,[16] it is plausible to also presume a causal relationship between the patient's complaints of hypersalivation and heightened sympathetic activity in the submandibular ganglion. Interestingly, of the other 22 patients who underwent this specific procedure involving the external carotid artery, no other instances of RSD were identified.

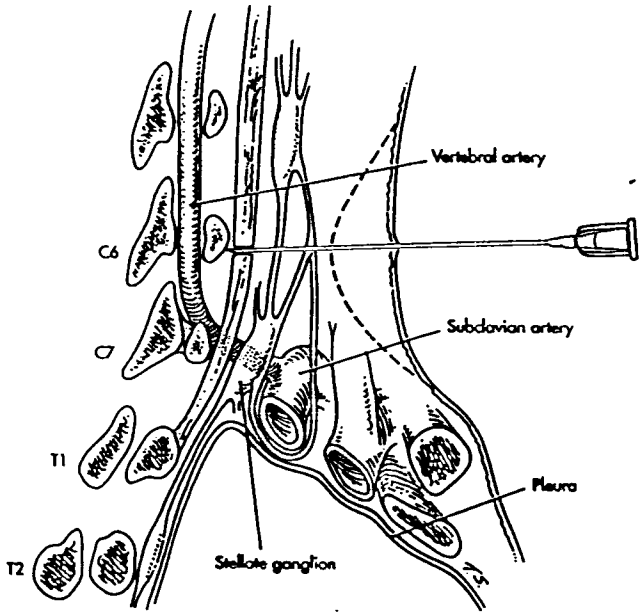

Fig. 2. Sagittal view of the sympathetic chain. Stellate ganglion is positioned directly posterior to the vertebral artery. The longus colli muscle separates the ganglia from the bone at C6 level. The needle is placed superior to the stellate ganglion. (From Raj PP, ed. *Practical Management of Pain.* edn 2. St. Louis: Mosby–Year Book; 1992; with permission.)

CONCLUSION

Facial RSD would appear to have a more favorable prognosis, since sympathetic blockade has consistently yielded favorable responses that appear to be long-lasting. A heightened awareness of this probably underreported disease entity is advocated to allow for early diagnosis and cost-effective treatment. Regardless of cause, stellate ganglion blocks with local anesthetic would appear to be both diagnostic and the treatment of choice, even when initiated as a delayed and repetitive injection series.

BIBLIOGRAPHY

1. Mitchell SW, Morehouse GR, Keen WW. *Gunshot Wounds and Injuries of Nerves.* Philadelphia: JB Lippincott; 1864:77–90.
2. Omer GC, Thomas MS. Treatment of causalgia. *Tex Med* 1971;67:93–6.
3. Rowlingson JC. The sympathetic dystrophies. *Int Anesthesiol Clin* 1983;21:117–29.
4. Veldman PHJM, Reynen HM, Arntz IE, Goris RJA. Signs and symptoms of reflex sympathetic dystrophy: prospective study of 829 patients. *Lancet* 1993;342:1012–6.

5. Bingham JAW. Causalgia of the face: two cases successfully treated by sympathectomy. *BMJ* 1947;1:804–5.
6. Hanowell ST, Kennedy SF. Phantom tongue pain and causalgia: case presentation and treatment. *Anesth Analg* 1979;58:436–8.
7. Khoury R, Kennedy SF, MacNamara TE. Facial causalgia: report of case. *J Oral Surg* 1980;38:782–3.
8. Jaeger B, Singer E, Kroening R. Reflex sympathetic dystrophy of the face: report of two cases and a review of the literature. *Arch Neurol* 1986;43:693–5.
9. Veldman PHJM, Jacobs PBDJ. Reflex sympathetic dystrophy of the head: case report and discussion of diagnostic criteria. *J Trauma* 1994;36:119–21.
10. Wang JK, Johnson KA, Ilstrup DM. Sympathetic blocks for reflex sympathetic dystrophy. *Pain* 1985;23:13–7.
11. Kleinert HE, Cole NM, Wayne L, Harvey R, Kutz JE, Atasoy E. Post-traumatic sympathetic dystrophy. *Orthop Clin North Am* 1973;4:917–27.
12. Lankford LL, Thompson JE. Reflex sympathetic dystrophy, upper and lower extremity: diagnosis and management. In: American Academy of Orthopaedic Surgeons: *Instructional Course Lectures*. St. Louis: CV Mosby; 1977: 163–78.
13. Hannington-Kiff JG. Sympathetic nerve blocks in painful limb disorders. In: Wall PD, Melzack R, eds. *Textbook of Pain*. New York: Churchill Livingstone; 1994:1046.
14. Collins SL. The cervical sympathetic nerves in surgery of the neck. *Otolaryngol Head Neck Surg* 1991;105:544–55.
15. Segal K, Lisnyansky I, Nageris B, Feinmesser R. Parasympathetic innervation of the salivary glands. *Operative Techniques Otolaryngol Head Neck Surg* 1996:333–8.
16. Richins CA, Kuntz A. Role of sympathetic nerves in the regulation of salivary secretion. *Am J Physiol* 1953;173:471–3.