

Case Report



Chronic Neuropathic Pain of Brachial Plexus Avulsion Mistaken for Amputation Stump Pain for 36 Years: A Case Report

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Conflict of Interest

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ABSTRACT

Following an amputation of the extremities, chronic neuropathic pain and discomfort, such as phantom limb pain (phantom pain), phantom sensation, and stump pain may occur. Clinical patterns of phantom pain, phantom sensation, and pump pain may overlap and these symptoms may also exist in one patient. Serious trauma to the upper limbs can result in brachial plexus avulsion (BPA). If BPA occurs at the same time as severe trauma of the upper extremity and the amputation of the upper limb is performed, chronic neuralgia caused by BPA may be mistaken for chronic amputation pain, such as phantom limb pain or stump pain. No major treatment advances in phantom pain have been made. However, unlike phantom limb pain, chronic neuropathic pain caused by BPA can be effectively treated with dorsal root entry zone lesioning (DREZ)-otomy. We report a patient who suffered for 34 years because the neuralgia caused by BPA was accompanied by an amputation of the arm, and so was thought to be amputation stump pain rather than BPA pain. The patient's chronic BPA pain improved with microsurgical DREZ-otomy.

Keywords: Amputation; Brachial plexus; Brachial plexus neuropathies (avulsion); Dorsal root entry zone (DREZ); Nerve root avulsion; Neuropathic pain

INTRODUCTION

Chronic neuropathic pain occurs in 30–90% of patients following brachial plexus injury.^{1,2,15} In patients with root avulsions that are predominantly preganglionic lesions, severe chronic neuropathic pain occurs in up to 90% of the patients.^{9,11} Neuropathic pain following brachial plexus avulsion (BPA) typically manifests characteristic pattern of continuous background pain described as burning, throbbing, and/or aching sensations, with electrical shooting paroxysm.¹⁵ It is almost consistently unbearable and resistant to all classes of analgesic agents, including opioids, as well as anticonvulsant and antidepressant drugs.¹⁵

Stump pain is defined as pain referred to the amputation stump.¹⁰ Although stump pain is common in the early postamputation period, severe chronic pain has been reported to occur only in 5–10% of the patients with amputations.¹⁰ The nature of stump pain has been described as pressing, stabbing, burning, squeezing, or stabbing.⁶ If a patient with trauma

to the shoulder and upper extremity requiring amputation has a BPA at the same time, the pain caused by the BPA may be mistaken for amputation stump pain. We report a patient who suffered for 34 years because the neuralgia caused by BPA was accompanied by an arm amputation, and so was thought to be amputation stump pain rather than BPA pain. The patient's chronic BPA pain improved with microsurgical dorsal root entry zone (DREZ)-otomy.

CASE REPORT

A 65-year-old right-handed male patient presented with an unremitting pain in his right shoulder over a period of 34 years (**FIGURE 1A**). He was involved in an accident in 1983 when his right arm was pulled into a press machine while working in a factory. Severe crushing injury in the right arm led to the amputation of the entire arm at the shoulder joint (shoulder disarticulation). About a year later, a tingling pain developed in the phantom hand and fingers of the amputee. The tingling pain persisted throughout his phantom hand and fingers, and intermittent electrical shock-like pain superimposed every few seconds. His phantom hand and finger pain was very severe and lasted day and night and was not controlled by anticonvulsants and painkillers. The pain was so uncontrollable that he was admitted to a university hospital twice and examined, but no specific treatment was advised.

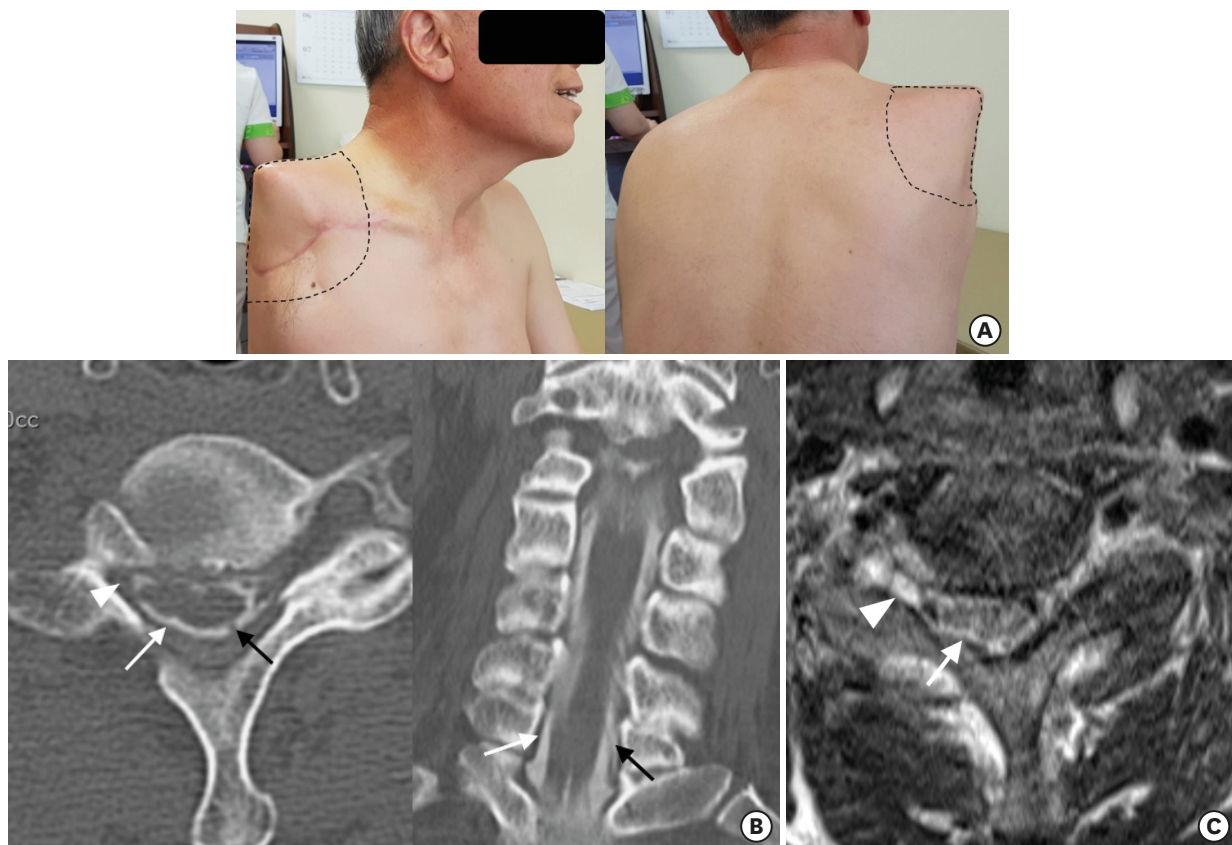


FIGURE 1. Clinical photographs and radiological images showing the amputation location and the area of pain. (A) Photograph showing the amputation stump on the right shoulder. The stump was well-padded and healthy. Chronic pain was located in the area inside the dotted line. (B) Axial (left) and coronal (right) myelographic computed tomographic images show the absence of the dorsal root (white arrowhead) and atrophy of the right-sided hemicord (white arrow) at the C6/7 level. The left-sided C7 root (black arrow) is normal. (C) The axial T2-weighted magnetic resonance imaging image shows the loss of the dorsal root (white arrow) and atrophy of the right hemicord at the C6/7 level. In the right C6/7 foramen, widening of the subarachnoid space, suspicious of pseudomeningocele (white arrow head), suggesting brachial plexus avulsion is identified.

As the decade passed, the right phantom limb disappeared as it gradually shrank in the severed shoulder area (telescoping phenomenon). Although the right phantom limb disappeared as it became smaller, but the continuous and paroxysmal pain did not disappear and continued in the shoulder area of the amputation. Opiates did not work and have severe side effects, so he lived with pain for 20 years while taking 600 mg per day of pregabalin.

Upon examination, there was no right arm in the right shoulder due to shoulder disarticulation caused by the accident in 1983 (**FIGURE 1A**). There was no mass in the stump of the right shoulder and no tenderness was observed. The skin over the stump looked healthy. The sensation over the right shoulder stump was normal and movement of the right scapula was possible. He denied any phantom sensation and phantom pain. The pain pattern was the same as when it first occurred. With the presence of continuous tingling pain, additional pain such as electric shock occurred once every few seconds. His pain was mainly distributed in the area of the amputation stump. It was persistent and did not worsen in any particular instance, such as emotional or physical activity. However, if he stayed in one position for a long time during sleep, he would wake up from a sudden attack of shoulder pain and could not sleep until the pain is reduced for 2 or 3 hours. He rated his pain as 7 out of a range from 0 to 10 on the NRS-11 scale.

Because his pain, along with typical continuous tingling pain, included intermittent electric shocks, a cause of BPA was suspected. Myelographic computed tomography (CT) and magnetic resonance imaging (MRI) of the cervical spine with myelographic MRI were requested. On the myelographic CT scan, ventral and dorsal roots were absent from C7 to the T1 level (**FIGURE 1B**). Pseudomeningocele was not observed. In the MRI, the cervical spinal cord was distorted and a slightly high signal was observed along the dorsal horn of the avulsed segments of the cervical spinal cord (**FIGURE 1C**). Some widening and elongation of the dorsal root sleeve of the right C7/T1 dural sleeve were noted.

After confirming a diagnosis of chronic neuropathic pain of BPA, microsurgical coagulation of the DREZ (microsurgical DREZ-otomy) was performed. After performing right-sided hemilaminectomy from C5 to T2, the dura was opened. Intraoperative neurophysiologic monitoring of the motor evoked potential (MEP) and the somatosensory evoked potential (SEP) of ipsilateral and contralateral arms and legs were performed.¹⁸⁾ The avulsed posterolateral sulcus (C7–T1) was identified under microscopic vision. After opening the pia mater of the avulsed sulcus, coagulation of grayish-yellow-colored gliotic dorsal horn was performed with fine-tipped, bipolar coagulation forceps specially designed for the application (**FIGURE 2A & B**).^{15,17,18)} During the coagulation, the SEP and the MEP of right lower extremity and left extremities were stable (**FIGURE 2C**).

Within 2 days after the microsurgical DREZ-otomy, the patient's recurrent paroxysmal pain disappeared completely and his continuous tingling pain decreased significantly. There were no neurologic complications postoperatively. His pain attacks did not recur until a year after the operation. He assessed that pain was 80% less than before surgery and was tolerable. His medication was 150 mg of pregabalin a day.

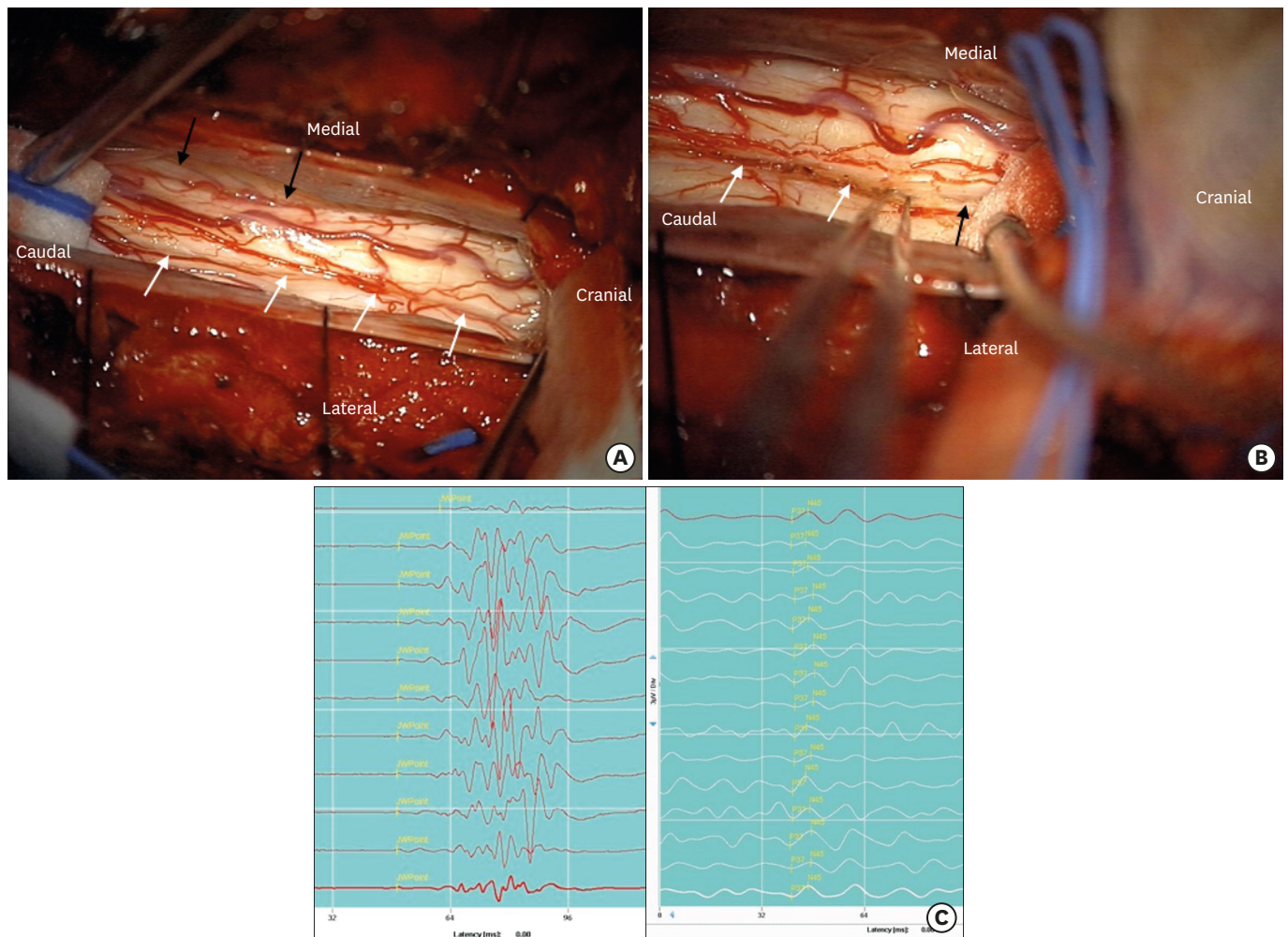


FIGURE 2. Intraoperative photographs during DREZ-otomy and intraoperative monitoring for brachial plexus avulsion pain. (A) Intraoperative photograph showing avulsion of the right-sided dorsal roots. There is no dorsal root in the right dorsolateral sulcus (white arrows) and grayish-yellow discoloration is observed along the avulsed dorsolateral sulcus. Normal dorsal roots (black arrows) are observed in the left dorsolateral sulcus. (B) Intraoperative photograph showing DREZ-otomy with fine-tipped bipolar coagulation. The lower level dorsolateral sulcus (white arrows) on the left side of the photograph is in the coagulation state, and the sulcus on the right side (black arrow) in the picture is still in the state before coagulation. (C) The motor-evoked potentials and somatosensory-evoked potentials in the right leg did not change significantly during surgery. DREZ: dorsal root entry zone.

DISCUSSION

Pain following amputation

Amputation is carried out for diseased or injured limbs when attempts at salvage and reconstructions are unsuccessful.^{19,21} Traumatic injury is the most common indication for upper limb amputation, whereas peripheral vascular disease consistently remains a more frequent cause of lower limb amputations.^{19,21} Amputation is followed by phantom sensations, painful or not, in almost all patients.^{3,10,22} In some patients, the missing limb becomes the site of severe pain.³ Phantom limb pain (phantom pain) refers to painful sensations referred to the missing limb and phantom sensation refers to any sensation of the missing limb except pain.¹⁰ Stump pain (residual limb pain³) is defined as pain referred to the amputation stump. Spontaneous movement of the stump that ranges from small jerks to visible contractions is referred to as stump contractions.¹⁰ There is overlap among these

elements, and in the same individual, phantom pain, phantom sensations, and stump pain often co-exist.¹⁰⁾ Although much research has been done on the subject, no major treatment advances in phantom pain have been made.¹⁰⁾

The causes of post-amputation stump pain vary. It can be classified as having intrinsic and extrinsic causes.¹⁹⁾ Aggressive bone edges, heterotopic new bone formation, painful neuroma, osteomyelitis, phantom pain, and tumor recurrence are reported as intrinsic factors.¹⁹⁾ Sump bursitis, bone bruises, and stress reactions and cutaneous lesions are known as extrinsic factors.¹⁹⁾

Pain of brachial plexus avulsion

The majority of brachial plexus traumas occur in motor vehicle and motorcycle accidents.²⁰⁾ However, a sudden force to the head or shoulder can stretch the brachial plexus, as is not uncommonly seen in sports injuries or industrial or occupational accidents from a falling object, and also result in brachial plexus injury.^{8,20)} The mechanisms of brachial plexus injury are variable but traction and compression injuries are most commonly involved.²⁰⁾ In traction injuries, downward displacement of the upper extremity causes a greater degree of injury at the level of the upper roots and trunks. Alternatively, upward displacement of the upper extremity forcefully increases the scapulohumeral angle and is more likely to injure the lower plexus.²⁰⁾ Fixed attachments of the cervical roots to the vertebrae and prevertebral fascia limit the mobility of the brachial plexus. With severe forces, avulsion of the nerve or root may occur.²⁰⁾

The level of the traumatic lesion relative to the dorsal root ganglion (DRG), which contains the sensory cell bodies, affects the clinical presentation and the management.⁷⁾ The location of the lesion relative to the DRG provides information to the potential feasibility of surgical reconstruction. Injury distal to the DRG (postganglionic lesion) indicates a more distal lesion and suggests that health nerve stump is more likely to be present for grafting.⁷⁾ Injury proximal to the DRG (preganglionic lesion) is much less likely to preserve a proximal nerve stump for grafting.⁷⁾ BPA is a typical example of a preganglionic lesion.

In the current case, BPA occurred in an industrial accident that resulted in severe crushing injuries to the upper extremity and severe traction injury to the brachial plexus. It seems that a thorough history by a focused physical examination, which could facilitate the recognition of potential brachial plexus injury for concomitant severe crushing injury to his right upper extremity, had been missed. For occupational or industrial injuries, information about the involved machinery, the force and vector of the pull that caused the injury, and any attempts to pull the patient's arm from the machine by coworkers would help to identify an occurrence of BPA.²⁰⁾

Delayed diagnosis of BPA pain

A patient with a brachial plexus injury is often seen initially with a multisystem trauma in the emergency department and should be managed by the standard trauma resuscitation protocol.²⁰⁾ Because the patient is usually the first to detect brachial plexus palsy, the diagnosis may be delayed in an unconscious patient or if sedation or anesthesia is required.²⁰⁾ In the current case, the diagnosis of BPA was typically delayed due to amputation of the right upper extremity. The pain from BPA was mistaken for that caused by stump pain. Because the patient had no right upper extremity, weakness and sensory change in the right upper extremity could not be detected. In addition, amputation of the right upper extremity prevented not only the physical examination but also the electrophysiology tests such as electromyography and nerve conduction tests.²⁰⁾ However, even if there is no arm due to

amputation early after the accident, EMG signals from the paravertebral muscles may help diagnose the avulsion of the brachial plexus.⁴⁾ Radiological examinations were helpful in identifying the presence of BPA in the current case. Myelographic CT and MRI of the cervical spinal cord are important in identifying BPA.^{15,20)} The absence of a root at a particular level is diagnostic of a root avulsion.^{15,20)} Pseudomeningocele is the visualization of healed radicular dural sleeves by the formation of scar tissue following a tearing of the surrounding dural sheath.²⁰⁾ Pseudomeningocele is consistent with root avulsion but is not diagnostic, as the dural sheath remains intact despite root avulsion, or a dural tear may occur without root avulsion.^{15,20)} In the present case, the absence of roots at the C7–T1 level was confirmed, but pseudomenisocels were not identified.

Treatment of medically refractory BPA pain

Treatment of post-amputation pain is challenging owing to the diversity in etiologies, which can be attributed to an underlying disease. In the past, surgery on amputation neuromas and more extensive amputation played important roles in the treatment of stump and phantom pain.¹⁰⁾ Today, stump revision is probably performed only in cases of obvious stump pathology. In properly healed stumps, there is no indication for proximal extension of the amputation because of pain.¹⁰⁾ The pain of BPA that is a preganglionic injury to the brachial plexus is not relieved with amputation.¹⁰⁾ In a series of 750 patients with brachial plexus injuries, elective amputation was performed in 13 patients at the patient's request and as a possible element of rehabilitation.^{6,10)} However, the pain of preganglionic injury (BPA) was not relieved at all by amputation.^{6,10)}

Transcutaneous electrical stimulation and spinal cord stimulation are not particularly effective in the treatment of BPA pain because after BPA, most of the fibers targeted by stimulation undergo degeneration up to the brainstem.¹⁶⁾ Deep brain stimulation has not demonstrated long-lasting relief in patients with pain after BPA.¹⁵⁾ Other neuroablative operations, cervical anterolateral cordotomy and mesencephalic tractotomy have not demonstrated long-term efficacy and are not exempt from disabling side effect.¹⁵⁾ In contrast, lesioning in the DREZ has been shown to be significantly effective in relieving pain after BPA.^{5,9,12,15)} The aim of DREZ lesioning is the treatment of neuropathic pain associated with dysfunction of the gating circuitry of the dorsal horn and the pain generated by dorsal horn hyperactive neurons.¹⁵⁾ Indeed, abnormal spontaneous hyperactivity in the avulsed dorsal horn was demonstrated with microelectrode recording.⁵⁾ Lesioning in the DREZ is undeniably effective.¹⁵⁾ In a series of 55 patients with BPA pain, 66% showed excellent (total relief without medications) or good (total relief with medication) pain relief and 71% experienced an improvement in activity levels.¹⁵⁾

In general, paroxysmal pain responded better to DREZ lesioning than continuous pain.¹⁵⁾ According to Sindou et al.,¹⁵⁾ only the paroxysmal pain component was statistically associated with a worthwhile long-term result ($p=0.04$). However, the presence of continuous background pain did not constitute a negative prognostic factor ($p=0.26$).¹⁵⁾ Failures and recurrences of pain following DREZ lesioning were not found to be statistically correlated with the time elapsed between the injury and the onset of pain or even with the duration of pain prior to DREZ lesioning.^{12,15)}

Because DREZ lesioning involves ablation of the Lissaur tract and the dorsal horn within the intraparenchymal spinal cord, it inevitably carries a small but significant risk of neurological injuries, such as ataxia with tactile hypesthesia, weakness in the ipsilateral lower extremity,

and genitourinary disturbances.^{15,17)} The dorsolateral sulcus and the underlying dorsal horn, which are surgical target for DREZ lesioning, are anatomically located between the dorsal column and the corticospinal tract. These long tracts are potentially vulnerable structures during DREZ lesioning.^{17,18)} Therefore, intraoperative monitoring using somatosensory-evoked potentials and muscle MEPs (mMEPs) has been introduced in DREZ lesioning.^{17,18)} In the current case, there was no major alarming event in SSEP and mMEPs during lesioning and there were no neurologic sequelae after the operation.

CONCLUSION

Severe trauma to the upper limb sometimes requires amputation. At the same time, BPA occurs in severe upper limb trauma. If amputation is performed in the upper limbs where BPA has occurred, the chronic pain caused by BPA may be mistaken for stump pain that occurs after amputation. If severe chronic pain occurs in the amputated arm, careful and radiographic examinations are needed to see if BPA has caused the pain.

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