Letters

Coping With Incongruence: Mirror Therapy to Manage the Phantom Limb Phenomenon in Pediatric Amputee Patients

To the Editor

In pediatric patients with cancer-related amputations, the incidence of "phantom limb" pain is close to 90%.[1] Although its physiopathology has not been fully explained, mirror therapy (MT), in conjunction with other treatments, has shown promising results in adults. In children, data are more limited and there are no controlled, prospective studies proving its efficacy, nor is there a standard for when it should be initiated, or its frequency or duration of the sessions.[2,3]

Most articles published about MT in children refer to controlling pain and motor or sensory sensations, but practically none of them evaluated the benefit this technique can provide for diminishing the anxiety and frustration that can be caused by this pain in young patients.

These children have trouble understanding the incongruence they are experiencing (feeling pain in a limb that is gone). They find it frustrating not to be able to explain their symptoms to their parents or doctors, and furthermore, they need to cope with the sudden and dramatic change that losing a limb has at a very young age.

Some papers, such as the one published by Ramsey in this same journal,[4] already intuited that MT could provide benefits beyond the diminishment of pain or the phantom sensations. We believe the case study being presented is a very good example of this.

Case Description

A nine-year-old child was referred to our pediatric oncology unit with telangiectatic osteosarcoma located in the distal third of the left femur. After three cycles of neoadjuvant chemotherapy, the magnetic resonance showed tumor progression. Limb salvage surgery was rejected, and supracondylar amputation was carried out. Gabapentin was initiated two days before surgery and maintained for 12 weeks. He also received nonopioid analgesics and amitriptyline during the postoperative period.

Soon after surgery, he reported pain and unpleasant sensations. He felt cold, burning, and spontaneous movements on the left leg that no longer existed. He described that his foot was stepping on big, hard stones and his ankle and foot wrapped by them. These sensations went from the toes to 10 cm above the ankle. The intensity of these incessant sensations provoked anxiety and catastrophic thoughts. Ten days after surgery, we decided to initiate MT as adjuvant therapy. It was initially applied five days/week, but once symptoms improved, sessions were held on the days he came for chemotherapy. Each session took approximately 15 minutes.

We designed a questionnaire that assessed different aspects of his phantom limb phenomenon, with some figures to help him describe the sensations. Before each session, the patient and the therapist completed the questionnaire.

The patient was asked to sit on the floor with his back against the wall, and his stump aligned with the contralateral limb. A long mirror was placed between both thighs, with the reflecting face toward his intact leg. This way, he could look at the reflection of his leg, creating the illusion that his amputated leg was still there. Once the illusion was created, he was asked to try to move "both lower limbs" at the same time. Before executing these movements, we asked him to "connect" with his left leg. To achieve this connection, he had to keep his mind on the reflection of the intact leg as he tried to move both legs simultaneously. Thereby, both the intention to move the missing leg and the reflection of the intact leg matched.

On the fifth day of therapy, he reported that "something magical was happening." While moving both his legs, he fixed his gaze on the mirror and said that his phantom sensations had momentarily disappeared. From then on, he prepared very thoroughly for each day's MT session. He tried to generate an illusion that was as realistic as possible, placing the mirror carefully or moving away any object that might diminish the realism of the illusion. Once connected to his missing leg, he performed all the movements he was asked

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to do and spent some time practicing free movements. He realized that during MT, his phantom sensations decreased significantly. We also observed that when he connected to his phantom leg, while performing the movements, his hip movements were congruent, although it was the only intact joint of the amputated leg. Somehow, he was practicing some kind of mental imagery.

During the eight-week process, we observed evolution in the pain score and characteristics. During the first two weeks of MT, he gave high scores to his pain. Over the next four weeks, the scores decreased drastically.

Concerning pain distribution, throughout the MT period, he reported the same pain area, regardless of the intensity. It is worth noting that he didn't feel his left leg entirely. "It is as if my ankle just emerged from my stump … as if I didn't have a knee." At the end of the eighth week, he reported pain release and gave very low scores both in the visual and numeric scales in our questionnaire.

Comments

To the best of our knowledge, the use of MT for phantom pain in pediatric cancer patients was first reported by Anghelescu et al.[1] The authors concluded that patients receiving MT had a shorter duration and lower incidence of phantom limb pain in a one-year follow-up. Recently, Ramsey et al. reported the use of MT in a seven-year-old male with osteosarcoma.[4]

The physiopathology of phantom limb pain and the effect of MT are not fully understood yet. MT is associated with instant and long-lasting neuromodulatory effects, involving perceptual, motor, and higher cognitive-related regions.[5] Following our experience with this patient, we concur with the hypothesis that MT creates a vivid illusion of the lost leg's presence, resolving the incongruence between what he visually perceives and what is represented in his motor and somatosensory cortex.[6] The patient feels immediate and temporary pain relief that is thought to contribute to adapting to their new body image.

Our MT protocol was based on those described previously by other authors.[7–9] We realized that the exercise protocol is not as important as connecting with the lost limb through a vivid and long-lasting illusion.

The telescoping phenomenon is infrequent and consists of the altered sensation of the phantom limb's length.[10,11] The patient's drawings allowed us to objectify that he was experimenting the telescoping phenomenon and to give him an explanation, which eased his anxiety.

Our objective with this technique was initially to help to reduce the pain in a noninvasive manner. We immediately saw that the effect of the therapy was very significant to resolve the incongruence between the lack of sensory afference and permanence of the cortical representation of the limb, as well as reducing the high level of anxiety caused by the onset of unexplainable pain due to it being incongruent and unreal.

During therapy, he found a space where the image reflected on the mirror allowed him to understand his own pain and verbalize his sensations. This helped the patient and his family to understand the "phantom limb phenomenon" and to adjust to his new body schema and cope with the loss.

We believe that prospective studies conducted on the effectiveness of MT should try to show the benefits of this technique for pediatric patients in diminishing their feeling of helplessness, frustration, and anxiety associated to phantom pain.

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https://doi.org/10.1016/j.jpainsymman.2018.10.495

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Intractable Neck Pain in a Patient With Newly Diagnosed AML: An Underrecognized Cause of a Treatable Syndrome

To the Editor:

Crowned dens syndrome (CDS) is a variant of pseudogout involving calcium pyrophosphate dihydrate (CPPD) deposition in the atlantoaxial junction. Imaging shows characteristic crown-like calcifications surrounding the dens.[1] Patients present with severe neck pain, rigidity, and high fever. Clinical presentations can be variable, including acute- and chronic-relapsing types, and can mimic many other disease processes. Awareness of this rare, likely underrecognized, and treatable syndrome is crucial because palliative care providers care for patients at high risk for this disease. We present a patient with severe, relapsing neck pain due to CDS that was complicated by a new diagnosis of acute myeloid leukemia (AML) at presentation.

Case Description

A 75-year-old man with a history of prostate cancer, gout, Type 2 diabetes mellitus, and chronic low back pain presented to the emergency department with fever and severe neck pain. He had come to the emergency department several times in the past month for severe neck pain, which improved with nonsteroidal anti-inflammatory drugs (NSAIDs). Blood testing showed pancytopenia, and a subsequent bone marrow biopsy provided confirmation for a diagnosis of AML with 60% blasts. Palliative care providers were consulted for pain management on Day 10 of hospitalization for severe, uncontrolled, cancer-related pain.

On examination, the patient appeared uncomfortable. He had neck rigidity, right shoulder pain with pain radiating to his arm, and neck erythema and warmth. His erythrocyte sedimentation rate was high (120 mm/hour), which initially raised concern for leukemic meningitis. Hydromorphone patient-controlled analgesia was started. His right ankle swelled, and ankle arthrocentesis showed CPPD crystals. After he was given corticosteroids, his neck pain improved, and the possibility of CPPD deposition in the neck became a concern. Review of previous computed tomographic scans of the cervical spine showed calcification of the transverse ligament, consistent with CDS (Fig. 1). He began treatment with a prolonged 30-day course of prednisone with a slow taper, which resulted in complete resolution of his neck pain. He then proceeded with induction chemotherapy for AML without recurrence of CDS symptoms.

Comment

CDS is a rare and likely underrecognized disorder that can cause considerable morbidity and prolonged hospitalization. The incidence is uncertain, but CDS has been reported to occur in 1.9% of outpatients with neck pain and in 35% to 45% of patients with articular chondrocalcinosis.[1] CDS is more common in elderly patients (mean age, 71.4 years) and is equally common in male and female patients.[1] The pathophysiology of CDS involves CPPD or calcium hydroxyapatite crystal deposition in the ligaments surrounding the dens. These crystals induce inflammation, causing production of matrix metalloproteinases and prostaglandins.[2]

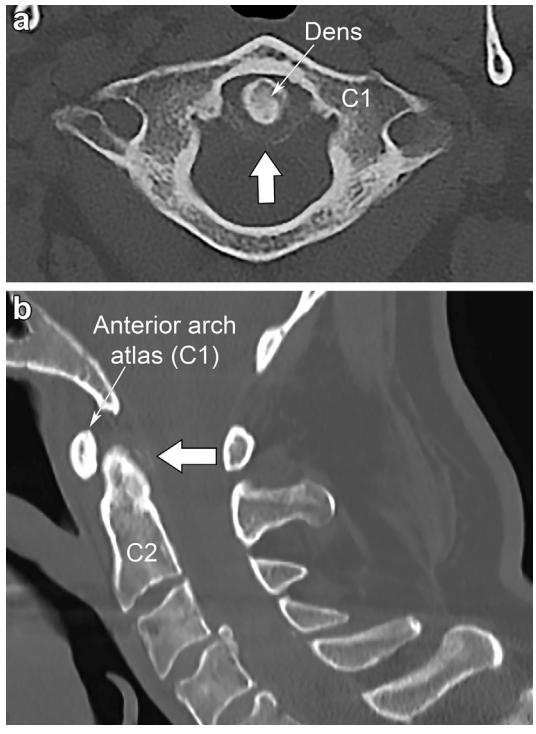

Fig. 1. Computed tomography of the cervical spine. a) Axial view shows linear calcification (arrow) in the transverse ligament of the atlas consistent with crowned dens syndrome. b) Sagittal view shows calcification surrounding the dens (arrow).