

COMPLEX REGIONAL PAIN SYNDROME OF CHILDHOOD AND ASTHMA IN 2 TEENAGERS PRESENTING WITH CHEST PAIN

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Background: Complex regional pain syndrome (CRPS) is a chronic pain syndrome characterized by sensory, motor, autonomic, bone, and skin changes. The onset of CRPS is most typically preceded by a traumatic event. Asthma is a chronic respiratory condition in which bronchial hyperresponsiveness and inflammation of pulmonary airways lead to symptoms of reversible lower airway obstruction, which may lead to musculoskeletal pain.

Case Report: Patient 1 is a teenage girl with poorly controlled asthma presenting with CRPS localized to her chest, upper abdomen, and upper back. Patient 2 is a teenage girl with well-controlled asthma but with a significant historical asthma exacerbation presenting with CRPS in her chest and upper back.

Conclusion: We suggest that a respiratory illness and associated asthma exacerbation served as the traumatic event for these patients' CRPS presentations.

Key words: Complex regional pain syndrome, amplified musculoskeletal pain syndrome, asthma, pediatrics, rehabilitation

BACKGROUND

Complex regional pain syndrome (CRPS) is a chronic pain syndrome characterized by sensory, motor, autonomic, bone, and skin changes (1). The term CRPS is often used interchangeably with other diagnoses, including reflex sympathetic dystrophy, reflex neuropathic dystrophy, and amplified musculoskeletal pain syndrome (2). In contrast to CRPS in adults, CRPS in children often presents with neurologic symptoms that are less pronounced (3). While the causes of the syndrome remain unclear, it is widely accepted that the condition typically arises in association with trauma, with the most common triggers including fracture, blunt traumatic injuries, and surgery (4). However, a significant number of cases arise spontaneously without a memorable event.

Current research suggests an etiology of CRPS related to abnormal inflammatory pathways, vasomotor

dysfunction, and dysfunctional neuroplasticity (5). An autoimmune component has also been theorized to play a role (1). Psychological factors were once thought to contribute, but evidence is contradictory (5). Studies suggest even minor tissue trauma can amplify cytokine signaling in tissue, impacting nerve growth factor and promoting long-term peripheral sensitization. A theory of "hyperalgesic priming" has also been suggested, in which a short-term injury triggers long-term changes in nociceptors, priming them to become hyperresponsive to future minor injuries that would not typically cause pain (5).

Asthma is a chronic respiratory condition in which bronchial hyperresponsiveness and inflammation of pulmonary airways lead to repetitive symptoms of reversible lower airway obstruction (6). Increased respiratory muscle use during asthma attacks and resulting changes in chest wall conformation may lead to mus-

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culoskeletal chronic pain in asthmatic patients (7). An association between CRPS and asthma has previously been suggested in the literature, thought to be due to a shared mediation by neurogenic inflammation (8,9). However, to our knowledge there have been no published reports of patients in which asthma served as a trigger of their CRPS. We present 2 cases of CRPS of childhood presenting as intercostal chest and thoracic pain in patients with a history of asthma.

Patient 1

The patient is a 17-year-old girl with a past medical history of asthma and dysmenorrhea who presented with worsening chest and back pain present for 20 months. There was no obvious antecedent or trauma related to the pain. The pain had been worsening in severity and at time of presentation was 7 of 10 in severity on the Numeric Rating Scale for pain at baseline and “20 out of 10” pain, per patient report, during a flare. She had missed a significant amount of school due to the pain in the previous academic year, and in the current academic year her attendance was so poor that her guidance counselor recommended dropping her advanced placement classes and taking online classes at a local community college instead. The pain had led to 2 emergency room visits in a 2-week timespan, at which time she was prescribed oxycodone-acetaminophen to manage the pain. The patient had also previously tried ibuprofen 800 mg for the pain, which provided no relief. She had seen a pulmonologist for the pain, who determined it was not respiratory in nature and recommended mental health counseling.

The patient’s asthma was diagnosed several years before and was significant enough to interfere with sports participation despite use of a rescue inhaler. She experienced lightheadedness when on inhaled corticosteroids and inhaled corticosteroid plus long-acting beta-agonist combination (ICS/LABA) combinations. She also reported a nonspecific sensation of unease when taking montelukast. Her asthma had been managed with yearly acupuncture, which her mother reported decreased symptoms. She also had a history of significant dysmenorrhea which caused her to miss 2 school days every menstrual cycle.

On exam, the patient was in significant distress but experienced no respiratory distress. She was unable to take deep breaths due to the pain and there was allodynia with palpation of the lower anterior chest and upper abdomen as well as the back in a bandlike

distribution around T6. No color changes or bruising were observed. Magnetic resonance imaging of the back revealed no skeletal abnormalities. Due to allodynia, including a regional distribution as opposed to a specific nerve distribution, and lack of pulmonary or spinal pathology, this patient was diagnosed with CRPS of childhood.

The patient was referred to outpatient physical therapy and psychological counseling and prescribed montelukast for her asthma and pyridoxine for her dysmenorrhea. Oxycodone-acetaminophen was discontinued and naproxen was started for pain management. The patient was able to find a physical therapist who was familiar with CRPS, who recommended a regime of daily activity that incorporated yoga. At that time, her mother also enlisted several complementary therapies to assist with pain, including hypnotherapy, guided meditation, and vitamin C supplementation. At one month’s follow-up, she was able to engage in normal physical activity and began dance again. She noted an increase in pain if she missed one to 2 days of her daily yoga regimen. Over the following months, she experienced occasional flare-ups of her musculoskeletal pain associated with asthma exacerbations. At 5 months after her initial presentation, her CRPS of childhood was well controlled with her yoga regime, and dysmenorrhea was improved with 200 mg pyridoxine. She is being followed by pediatric pulmonology, who have her on inhaled ipratropium and albuterol as needed for asthma exacerbations.

Patient 2

The second patient is a 17-year-old girl with a past medical history of asthma and hypovitaminosis D who initially developed chest wall pain and thoracic back pain, which was ultimately diagnosed as amplified musculoskeletal pain syndrome prior to presentation to our care. The patient described her pain as “burning” in her anterior chest and “dull” in her thoracic chest, with a severity of 4 of 10 at rest and 7 of 10 with normal activities. Over a period of 3 years, the patient had 2 significant pain exacerbations, one of which followed a diagnosis of atypical pneumonia in which a chest computed tomography scan showed a linear streak, possibly consistent with a historically undiagnosed pneumonia. The second exacerbation had no obvious trigger. She was successfully treated with outpatient physical therapy following her initial presentation and after her first exacerbation. However, outpatient physical therapy

was unsuccessful following her second exacerbation. As a result, she began homeschooling and stopped her hobbies including playing cello and participating in sports. She tried ibuprofen, naproxen, and acetaminophen for the pain without relief. Ultimately, her physical therapist recommended inpatient rehabilitation.

On initial examination upon arrival to inpatient rehabilitation, she reported her pain level at 7 of 10 and appeared mildly distressed. Allodynia along the mid-lower paraspinal areas extending to the bilateral flanks and sparing the midline spine, in addition to lower rib tenderness, were elicited on exam. She also had vague left lower-quadrant tenderness that disappeared with reexamination. The patient presented with hypomobility of the vertebral and costovertebral joints throughout her thoracic spine, as well as a pelvic obliquity which was corrected with an orthotic insert. Movement abnormalities were also present at evaluation, including stiffness and guarding with ambulation and activities of daily living. Due to the allodynia and history of previously diagnosed amplified musculoskeletal pain syndrome

and no signs of other pathology, patient 2's diagnosis was reconfirmed as CRPS of childhood.

She began inpatient rehabilitation with intensive occupational and physical therapy, focusing on improved thoracic mobility, general activity tolerance, and education surrounding her diagnosis. Respiratory therapy was also involved to educate her about effective breathing exercises. She began intensive psychological counseling (4-5 days/week) as well. After approximately 2 weeks of inpatient rehabilitation, the patient was able to tolerate activities of daily living, moderately intense exercise programs, and a return to community activities with a significant reduction in pain (4/10). She was discharged home with a home exercise program, continuing to address overall strengthening and thoracic mobility.

In summary, we have identified 2 female patients with symptoms consistent with CRPS of childhood, localized to the thorax, who appear to have pulmonary trauma as the trigger for their pain syndrome. We would recommend that patients with a history of asthma with chest and back allodynia be evaluated for possible CRPS.

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