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Clinical Quiz

Complex regional pain syndrome in a young female

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Case

A 16-year-old female was admitted in the internal medicine ward with severe and sudden onset pain of the right femoropubic region radiating down to the knee. Her pain began 3 days prior to admission and was exacerbated with movement or light touch. At the onset of pain the affected region was cold and cyanotic. There was oedema of the entire right lower limb. The patient was afebrile, and her left lower limb demonstrated no pathology. The patient reported a preceding soft tissue injury of the right lower limb ten days prior to admission.

Preceding the rheumatology consultation, the patient was assessed by internists, neurosurgeons, orthopaedic surgeons, neurologists, vascular surgeons, and psychiatrists. Collectively, they ordered a number of investigations including an MRI of the lumbar spine, MRI and MRA of the thighs and pelvis, radiographs of the affected joints, MRI of the brain, CT of the brain, ultrasound Doppler of lower limb, electromyography, and electroencephalography. None of the above clinical assessments and investigations revealed any pathology indicative of a diagnosis.

Rheumatological assessment revealed severe hyperalgesia in the right lower limb in conjunction with cyanosis and reduced temperature. The left lower limb was well perfused and warm. There were no findings suggestive of arthritis. Upon further questioning, the patient reported having had a similar event two years prior to the current complaint. All investigations and clinical assessments from the previous episode revealed no pathology either and were treated with physiotherapy alone.

A Tc-99m-MDP bone scintigraphy scan was requested by her rheumatologist. The first and second phases of the scan revealed mildly reduced uptake in the right lower limb as compared to the left. The third phase of the scan revealed increased uptake in the area of the right ankle, indicative of increased bone metabolism. These findings, in conjunction with the history, were suggestive of Complex Regional Pain Syndrome (CRPS).

The patient was commenced on intramuscular calcitonin but stopped after a single dose due to side effects including persistent flushing, headaches and nausea. Patient refused further drug therapy. Intensive physiotherapy was initiated with improvement of her symptoms after 2 months. The patient was followed-up monthly for 6 months and the current plan is to review biannually.

Commentary

CRPS in children is a rare entity, it is characterised by limb pain which is out of proportion to the examination and patient history. It is usually associated with sensory and motor disturbances[1]. The nomenclature and description of this syndrome has been under much debate since 1864 when it was first documented. In 1994 the International Association for the Study of Pain came to a consensus with the unifying term Complex Regional Pain Syndrome[2,3].

The pathogenesis of this condition is not fully understood, but it is thought that the syndrome possibly involves genetic, psychological with local and central nervous system factors[1,3]. The incidence of paediatric CRPS cases is <10% of total CRPS cases with observations suggesting that it is rarer in people of non-European ancestry[2].

CPRS in children varies greatly from CRPS in adults. In children, CPRS is frequently seen in adolescents affecting females more than males, typically seen between the ages of 9 and 15. Nevertheless, it can also occur at any age if the child suffers from inherited mitochondrial disease[1,2]. The lower extremities are most predisposed to being affected with an association of a psychological or traumatic event being the trigger[1]. CPRS is classified into two types; type 1 does not involve a clearly defined nerve injury whereas type 2 is associated with a well-defined nerve injury[2].

CRPS can present with a variety of symptoms, with one of its distinguishing symptoms being pain out of proportion to the event. The pain is usually described as throbbing with a burning sensation[4].

The diagnosis is made clinically, based on the physical examination and history, using the Budapest criteria together with

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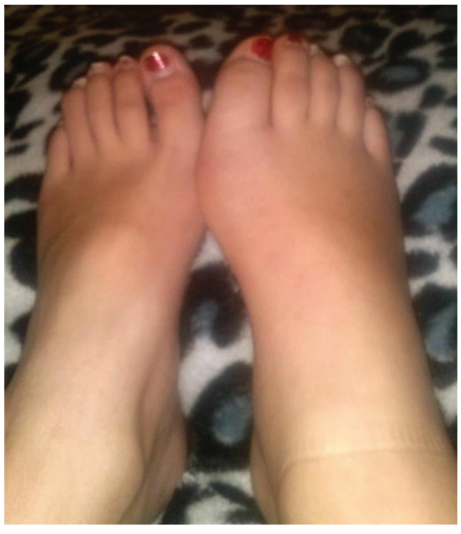

Figure 1. Image showing a stocking distribution of oedema of the right foot.

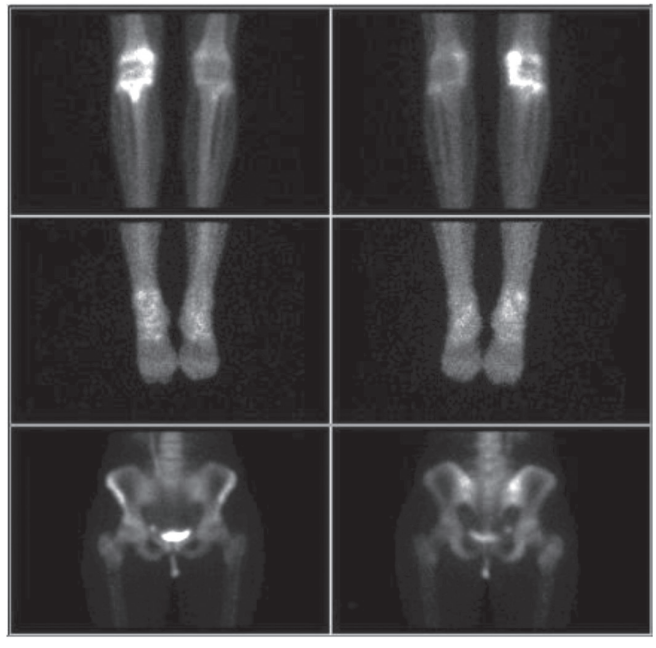

Figure 2. Images of a Tc-99m methylene diphosphonate (MDP) scan of the knee, ankle, and pelvis demonstrating a mild reduction in uptake of the right lower limb in phase one and two with an increase of uptake of the right ankle during the third phase.

signs of autonomic disturbance and allodynia[4]. Laboratory and radiological evaluation do not confirm or exclude CRPS but they help in excluding other pathological processes such as tumours, inflammatory conditions or trauma[4]. Technetium bone scintigraphy has a low specificity and sometimes shows a spotty increased pattern. Magnetic resonance scans may show oedema of the affected bony structure[2,4].

Treatment goals of CRPS in children are to relieve pain, restore function of the affected limb and assist the child in developing skills to improve his activities of daily living. The disease course is seen to vary unpredictably with periods of remission of symptoms. Occupational, physical, and psychological therapy are fundamental in the management of this disorder. Other treatments include pharmacological options such as ketamine, gabapentin, tricyclic antidepressants, topical dimethyl sulfoxide and bisphosphonates and pain reducing procedures. Examination of the sympathetic nervous system with non-invasive electrophysiology techniques can be used to monitor response to treatment[3]. A prospective study showed a 31% recurrence rate within the first 6 months[1]. In recurrent cases more invasive options such as surgery; sympathetic blocks, epidural catheter, regional nerve blocks and transcutaneous electrical nerve stimulation can be used to help provide sufficient pain control[1-4]. The prognosis varies on a case-by-case basis with no obvious predictors of outcome or recurrence[1,4].

References

1. Sherry DD, Wallace CA, Kelly C, Kidder M, Sapp L. Short and long-term outcomes of children with complex regional pain syndrome type I treated with exercise therapy. Clin J Pain 1999;15:218-223.
2. Borchers AT, Gershwin ME. Complex regional pain syndrome: a comprehensive and critical review. Autoimmun Rev 2014;13:242-265.
3. Matsui M, Ito M, Tomoda A, Miike T. Complex regional pain syndrome in children: report of three cases. Brain Dev 2000;22:445-448.
4. Tolba R, Badr R, Guirguis M, Soliman LM. Pediatric Continuous Brachial Plexus Catheter for a Case of Complex Regional Pain Syndrome (CRPS). Open J Anesthesiol 2015;05(01):1-6.

Questions

1. Which of the following is not a possible inciting event for CRPS?
A. Myocardial infarction
B. Cerebrovascular event
C. Urinary tract infection
D. Fractures

Critique

All the previously mentioned injuries in a history have been associated with inciting CRPS, except urinary tract infections. In adults, up to 40% of established inciting events were found to be soft tissue injury, followed by 25% for fractures, 12% for myocardial infarction and 3% for cerebrovascular infarctions. Infections of the urogenital tract were not associated with CRPS. It is important to note that in one report, no precipitating event was identified in 35% of patients.
The correct answer is C.

2. Which of the following imaging modalities is most useful for identifying CRPS?
A. Plain Xray
B. MRI
C. CT Scan
D. Bone Scintigraphy

Critique

Although changes have been observed in multiple imaging modalities, Bone Scintigraphy has been the most useful in identifying CRPS. It was shown to have a higher sensitivity (97%) and specificity (86%) than other modalities and is more able to differentiate it from other pathologies.
The correct answer is D.

3. What is the main difference between Type 1 and Type 2 CRPS?
A. Involvement of obvious nerve damage
B. Skin changes in the affected limb
C. Presence of generalised parasympathetic activity
D. Progression to limb deformity

Critique

Damage of a named nerve is the differentiating factor between type 1 and type 2 CRPS. Type 1, the most common, does not exhibit know nerve damage, while type 2 does. Type 2, often called “causalgia” is often more painful and difficult to control than type 1. Skin changes is a common feature to both types of CRPS.
The correct answer is A.