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Lesson of the Week

Reflex sympathetic dystrophy in children

Adrian R Lloyd-Thomas, Gillian Lauder

Reflex sympathetic dystrophy also occurs in children, commonly girls; early diagnosis gives the best chance for treatment

Reflex sympathetic dystrophy is well recognised in adults. It occurs in children but is rarely mentioned in paediatric textbooks. Children presenting with painful limbs often have many investigations and the diagnosis may be considerably delayed. This results in prolonged distressing pain for the child, anxiety for parents, and difficulty with treatment. We describe four cases to show the characteristics of the disease in children.

Case reports

CASE 1

An 11 year old athletic girl presented to this hospital with a 10 week history of increasing pain in her right leg. She had first seen her general practitioner with pain over the tibial tubercle of her right knee but no history of trauma. Her local orthopaedic service had diagnosed Osgood-Schlatter's disease and she had been advised to rest and use crepe bandage and elasticated tubular bandage.

Five days later her leg became cold and blue below the knee, with burning pain radiating from the knee to the foot. Deep vein thrombosis was diagnosed and she was treated with anticoagulant drugs. After a week anticoagulant treatment was stopped, but the leg continued to be painful, cold, and blue when in a dependent position. A full length plaster of Paris cast was applied in an attempt to relieve the pain of the Osgood-Schlatter's disease, but within days the symptoms were worse, affecting her whole leg up to mid-thigh. The blueness resolved only when her leg was elevated. The plaster of Paris was removed, but by this stage the pain was so severe that she would not put her foot on the ground and she was confined to a wheelchair.

On first examination at this hospital the leg was noted to be cold and blue and the foot oedematous. She had allodynia (pain provoked by a stimulus that does not normally cause pain) and hyperaesthesia up to her mid-thigh. Calf and quadriceps muscles were wasted, but results from neurological examination were other-

wise normal. Ultrasound scanning of the abdomen, radiography of the legs, venography, and arterial Doppler studies, all performed at the referring hospital, had given normal results; magnetic resonance imaging had shown changes compatible with Osgood-Schlatter's disease. Thermography with the thermal stress test gave results compatible with reflex sympathetic dystrophy.¹

She was given four intravenous regional guanethidine blocks and intensive physiotherapy every week to encourage mobilisation of the leg. At the end of one month's treatment she was walking normally with no pain in the leg, and all the vascular changes and oedema had gone. Moreover, she was able to begin playing sport once more.

CASE 2

A 9 year old athletic girl presented to her general practitioner with a 48 hour history of sudden onset of pain in the calf after playing sport at school. Initially strain was diagnosed and she was advised to rest and wear a support bandage. The pain failed to resolve; it did not respond to analgesics, and after five days the leg became blue when in a dependent position. She was referred to her local hospital, where extensive investigations (radiography, Doppler ultrasonography, magnetic resonance imaging, and blood testing) all gave normal results. The pain worsened until she was unable to put her foot on the ground.

When seen at this hospital she had allodynia and dysaesthesia and paraesthesia up to her mid-thigh; her leg was cold and blue when in a dependent position. She had oedema of the foot and there was some wasting of thigh and calf muscles. Her pulses were normal and there was no neurological deficit. She was prescribed four intravenous regional guanethidine blocks weekly and twice daily physiotherapy. After only two blocks she walked normally and all symptoms were resolved.

CASE 3

A 13 year old athletic girl presented to this hospital

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FIG 1—Case 4: Wasting of calf muscles, oedema of foot, and skin mottling associated with advanced reflex sympathetic dystrophy

FIG 2—Case 4: Osteoporosis seen in radiograph of foot

Symptoms and signs of reflex sympathetic dystrophy

Neuropathic pain

Burning
Dysaesthesia
Paraesthesia
Mechanical allodynia
Hyperalgesia to cold

Autonomic dysfunction

Cyanosis
Skin mottling
Hyperhidrosis
Oedema
Temperature difference between extremities of $>3^{\circ}\text{C}$

with a nine month history of pain in her right hip after a fall. Five days after the fall she had been referred by her general practitioner to her local orthopaedic service with deep seated hip pain. Radiography, ultrasonography, and all blood tests gave normal results. She had previously had an irritable hip, and this diagnosis was repeated. She received nine weeks of bed rest, with traction for four weeks. Although the pain improved slightly, the foot became oedematous and the skin changed colour. Mobilisation caused the burning pain to return with increased severity, and her leg was cold and very blue when in a dependent position. Conventional analgesics were unhelpful. She allowed her heel to touch the ground but refused to put weight on it. She had allodynia and dysaesthesia.

On presentation to this hospital her physical signs were the same but a radiograph of the hip showed osteoporosis on the right, and a bone scan showed increased uptake in the right hip and upper femur. A percutaneous lumbar sympathetic block performed to aid diagnosis relieved the pain and caused vasodilatation in her leg. The lumbar sympathetic chain was catheterised, with repeated local anaesthetic blocks on alternate days for three weeks. This was combined with physiotherapy. Progress was slow and difficult, but after one month the hip pain had gone and she could put weight on her leg but complained of residual pain in her knee. Three intravenous regional guanethidine blocks were given and relieved the knee pain. Over the next six months she returned slowly to normal activity.

CASE 4

Three years before being first seen at this hospital a 14 year old girl was referred by her general practitioner to the local orthopaedic service with pain in her right foot. She had had no history of trauma. Although a radiograph of the foot looked normal, osteochondritis of the metatarsal joint was diagnosed and physiotherapy started. There was little improvement; the pain worsened and typical skin colour changes were seen in the leg. She became unable to bear weight and underwent examination under anaesthesia and manipulation two and a half years after presentation. The pain worsened; she developed florid signs of reflex sympathetic dystrophy and was referred to this hospital.

She had obvious wasting of the calf muscles (fig 1). Foot radiography showed osteoporosis (fig 2), and a bone scan showed an uptake pattern consistent with reflex sympathetic dystrophy. Thermography with the thermal stress test¹ gave results typical of reflex sympathetic dystrophy. Magnetic resonance imaging and other investigations gave normal results. Her condition did not improve with intravenous guanethidine blocks but improved slowly with physiotherapy during percutaneous lumbar sympathetic blocks. Six months after beginning treatment she was barely able to bear weight.

Discussion

Reflex sympathetic dystrophy is "a complex disorder of pain, sensory abnormalities, abnormal blood flow, sweating and trophic changes in superficial and deep tissues."² Its pathophysiology remains unclear, but the initiating event (usually trauma) sensitises small diameter fibres, which generate ongoing activity and therefore react abnormally to mechanical, thermal, and chemical stimulation. Repetitive transmission sensitises and changes the decoding of both nociceptive and non-nociceptive information in the

dorsal horn of the spinal cord, thereby altering the sympathetic outflow to the affected limb.³

The incidence of reflex sympathetic dystrophy in children is not known, but the disease is probably underdiagnosed. Though the symptoms are the same in adults and children, the pattern of presentation is different. In adults it often occurs after a fracture or other trauma and follows a period of immobilisation (Sudeck's atrophy when fully developed). In children it is more common in athletic girls (the ratio in young men and women is 1:1³ but 1:6 in boys and girls); the average age at presentation is 12 years (range 5-17); and in most cases the legs are affected and there is no history of trauma.⁴ The average time from the onset of symptoms to presentation is 10 days, but the average time to correct diagnosis is one year. Diagnosis was delayed in cases 3 and 4. As this condition is not suspected in children, those affected often have many unnecessary investigations (radiography, vascular studies, etc) and may be overtreated—for example, with anticoagulants, as in case 1.

The clinical diagnosis is suggested by the presence of at least two symptoms and signs from each category (box), especially when the severity of pain exceeds that expected from any injury. Dysaesthesia is unpleasant, abnormal sensations occurring spontaneously or when provoked, while allodynia is pain resulting from a stimulus that does not normally cause pain. At presentation the autonomic signs may be subtle but are often present.³ Late signs include muscle wasting and contractures. Investigations should include limb radiography (both sides for comparison), bone scanning, and thermography with thermal stress testing.⁵ Patients should be referred early to a paediatric pain clinic where other diagnostic tests may be performed (sympathetic blocks with local anaesthetic and guanethidine or phentolamine tests). Treatment is directed at (a) the relief of pain (transcutaneous electrical nerve stimulation, amitriptyline 0.5-1.0 mg/kg/day, intravenous regional guanethidine blocks, and central sympathetic blocks); (b) mobilisation (intensive physiotherapy); and (c) psychological support. Physiotherapy is important as immobility exacerbates the symptoms. Psychological support should include relaxation training, learning cognitive-behavioural skills for managing stress, and family therapy.

Early diagnosis increases the chance of a rapid response to treatment, while diagnostic delay makes treatment less effective and much more prolonged. Moreover, the prognosis for full recovery is better in those treated early; patients with prolonged disabling symptoms often have persistent disease despite multiple therapeutic interventions.

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