Reflex neurovascular dystrophy

A case report

E. B. SCHRAADER

Summary

A child suffering from reflex neurovascular dystrophy (RND) who presented with a bizarre neurological picture is reported. RND is an extremely painful and disabling condition readily amenable to treatment. A striking feature is the localisation of pain and hyperaesthesia which do not follow a somatic dermatome distribution.

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Reflex neurovascular dystrophy (RND)[1] or reflex sympathetic dystrophy[2] is well documented in children.[1-3] Caused by sympathetic dysfunction, probably secondary to an underlying psychological disturbance,[1,4] it responds to sympathetic blockade.[2] It is characterised by severe burning pain, hyperaesthesia, vasomotor and sudomotor changes, oedema and muscle spasm,[1-3] and permanent dystrophic changes result without treatment.[3,5]

Case report

An 11-year-old girl had a minor injury to the right knee and later complained of pain and stiffness in the right leg, foot, arm and hand. The hand adopted a claw position with loss of function. Her father had recently sustained a head injury and subsequently had personality changes with severe fits of temper.

On examination the skin over the right hand, forearm, leg and foot was oedematous and cold with marked hyperaesthesia. The hand was discoloured blue with sweating in the palm, and was in a fixed claw position with joints partially flexed. There was muscle guarding with limitation of movement in the knee and elbow joints, more severe in the wrist and fingers and rigid plantar flexion in the ankle. No joint swelling was noted but all movements were extremely painful.

Urinalysis and full blood count were normal. Antistreptolysin-O titre, rheumatoid factor and screening for collagen diseases were negative. Radiographs of the chest and the bones of hands and feet were normal. An electro-encephalogram and an ECG were normal.

After physiotherapy the oedema, coldness and hyperaesthesia disappeared, with recovery of function after 4 days. A psychiatric evaluation concluded that the father's behaviour after his accident might have been a stress factor. The parents were fully informed and instructed how to respond to a recurrence.

Four months later a relapse occurred. Both the patient's legs and feet were affected and she was bedridden. Muscle wasting was present in both legs. The additional physical findings in both legs were identical to those initially present in the right leg. The father had begun abusing alcohol and maltreated his children. The patient again responded to physiotherapy, which was prolonged because of shortening of the right Achilles tendon. Psychotherapy was continued at home. She remained well at follow-up.

Department of Paediatrics and Child Health, University of Stellenbosch and Tygerberg Hospital, Parowvallei, CP
E. B. SCHRAADER, M.B. CH.B., M.MED. (PAED.)

Discussion

RND is an extremely painful disabling condition lasting from days to years;[1] there is a risk of permanent damage.[3,5] These patients present on crutches, in wheelchairs and with arms in slings. The diagnosis is unsuspected in the majority,[1] and many cases with minor manifestations may improve spontaneously.

RND is more common in girls and mean age at onset is 12,4 years.[1] Remissions may be followed by relapses.[1-3] The main presenting complaint is constant burning pain in the involved extremity and hyperaesthesia,[1-3] which may follow a stocking distribution.[4] Vasomotor instability is always present and oedema of the skin and sudomotor changes are found as manifestations of autonomic dysfunction.[1-4] Reflex spasm of small or large muscle groups occurs,[1] resulting at times in unusual posturing without apparent effort.[3,5] The patient may become completely immobilised by pain.[1-3] Late dystrophic skin changes are followed by atrophy of subcutaneous tissue and muscle, osteopenic changes in the underlying bone, and shortening of limbs.[1,3,5] Subcutaneous fibrosis or fixed joint contracture rarely develop in children.[1,3,5] Dystrophic changes are reversible if treated early.[1,3,5] The initiating factor in RND is unknown. Less than 50% of patients have a history of injury (often minor) in the preceding year.[1,3] A predisposing personality trait and psychosocial background with a history of parental conflict, stress or a critical incident may be found.[1,5]

The clinical diagnosis should be suspected in patients who present with unusual neurological symptoms and signs. The only useful diagnostic investigation is infrared thermography.[4] Treatment includes reassurance, physiotherapy and psychotherapy.[1,5] Treatment with aspirin, indomethacin or corticosteroids or sympathetic block is not recommended.[1]

Conclusion

RND is probably underdiagnosed. Greater awareness of the condition will result in earlier diagnosis, the elimination of unnecessary invasive investigations and improvement in the response to treatment, thus avoiding permanent damage. A description of RND should be included in standard textbooks on paediatrics.

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REFERENCES

1. Bernstein BH, Singsen BH, Kent JT *et al.* Reflex neurovascular dystrophy in childhood. *J Pediatr* 1978; **93:** 211-215.
2. Doolan LA, Brown TCK. Reflex sympathetic dystrophy in a child. *Anaesth Intensive Care* 1984; **12:** 70-72.
3. Rush PJ, Wilmot D, Saunders N, Gladman D, Shore A. Severe reflex neurovascular dystrophy in childhood. *Arthritis Rheum* 1985; **28:** 952-956.
4. Gandhavadi B, Rosen JS, Addison RG. Autonomic pain. *Postgrad Med* 1982; **71:** 85-90.
5. Gold S. Diagnosis and management of hysterical contracture in children. *Br Med J* 1965; **1:** 21-23.