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Short report

Congenital ring constrictions with entrapment neuropathies

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SUMMARY An eight year old boy with multiple simple congenital ring constrictions is described with peripheral nerve palsies caused by entrapment in the constrictions. It is suggested that ring constrictions should be considered in the differential diagnosis of entrapment neuropathy.

Congenital ring constrictions (CRC) are uncommon findings in the newborn. Where not associated with skeletal or morphological limb deformities in the distal segment—syndactyly or amputation—histology has shown them to extend deep into the soft tissue.[1] With time these simple CRC tend to "grow out" and become less obvious, though surface markings remain. Sensory and motor changes have been reported distal to the rings.[2 3] but appearance of the secondary problems discussed here has not been reported.

We present an eight year old boy with multiple CRC who sustained over a two year period several peripheral nerve palsies which are believed to have been precipitated by entrapment of the nerves concerned in remnants of these constrictions.

Case reports

This eight year old boy was born at term following a normal pregnancy and delivery with a birth weight of 3·52 kg. At birth he was noted to have multiple deep skin creases producing a "Michelin Man" appearance. These were uncomplicated CRC with no evidence of distal abnormality. A dislocatable left hip was noted and resolved with splintage. His subsequent development was normal. Over the years these CRC have become less obvious and now are only faint surface markings (figs 1 and 2). When aged 4½ years, two hours after a minor fall at home he was noted not to be using his right arm. Radiographs were normal.

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When seen four weeks later he had a flaccid paralysis of the right arm involving especially elbow flexion and wrist extension. Unfortunately, no sensory examination was carried out. Two months after the injury electromyography showed evidence of degeneration within right biceps brachii and right extensor digitorum communis. Assessment of sensory conduction between digits one, three and five respectively, and wrist on both sides showed a slight relative decrease (30%) in amplitude of the sensory action potential during stimulation of right digit one. These features are consistent with a right upper brachial plexus palsy. Subsequent progress was good and full recovery had occurred by one year.

At 5½ years of age the patient's left elbow was hit by a stone and he sustained an impaction injury to the neck of the left radius. The left arm was supported in a sling but one week later the right arm became weak again. Following removal of the sling the right arm recovered over several weeks. Five months after this injury electromyography showed no abnormality in right biceps brachii. However, wasting and weakness of left abductor pollicis brevis was found. The maximal motor conduction velocity in fibres to this muscle was normal between axilla and above-elbow (71 m/s) but slightly reduced between above-elbow and wrist (48 m/s). The distal latency was normal (2·0 ms). A sensory action potential could not be recorded at the wrist during stimulation of left digit one. Sensory conduction between left digit five and wrist remained normal.

At 6 years of age he was first seen at the Ryegate Centre of the Sheffield Children's Hospital, six months after the second injury. Examination confirmed a left median nerve palsy with wasting and weakness of the abductor pollicis brevis but no sensory abnormality. Neurological examination was otherwise normal, in-

cluding that of the right arm. There were many transverse ring constrictions over his arms and legs, although none appeared to extend into the deep tissues. There were also two deep constriction rings in the neck (fig 2) and one in the groin. These were tethered deeply and were more extensive than the limb constrictions. Cervical spine, left arm, and skull radiographs were normal. Blood screening for ESR, fasting lipids, and sugar, lead immunoglobulins and acanthocytes was normal. Urine screening for porphyrins and mucopolysaccharides was normal. Recovery of the median nerve lesion was clinically complete seventeen months later. No further neuropathy has occurred. There is no relevant family history.

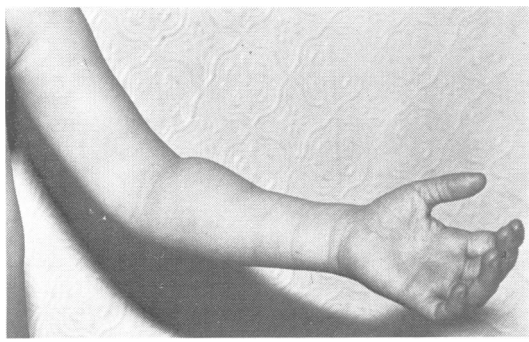

Fig 1 *Surface markings of constriction rings at 6 years.*

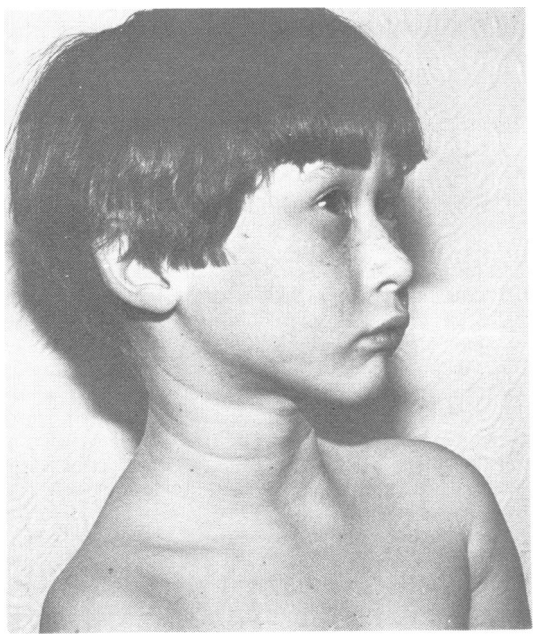

Fig 2 *Surface markings of constriction rings at 6 years.*

Discussion

The incidence of congenital ring constrictions has been quoted at 1 in 10 000[4] to 1 in 15 000.[5] Following the work of Torpin[1] it is now accepted that CRC are caused by bands of amniotic or mesodermal tissue constricting the developing foetus. In this case the postnatal history and findings were typical of simple CRC. Two odd points were the presence of CRC in the neck (which is rare) and the fact that CRC were not grouped distally, as is most common.

Clinically, closed brachial plexus palsies are commonly produced by severe traction or compression injuries.[6] The more superficial upper roots are the most vulnerable. The presence of abnormal anatomy or abnormally sensitive nerves in this region means that a lesser injury may have a more serious outcome. It is reasonably certain that the first brachial plexus palsy was caused by minor trauma and that the second episode of weakness represents a milder palsy caused by compression from the sling. Median nerve lesions are uncommon in undislocated fractures of the radial neck. It was important, therefore, to exclude predisposing anatomical, metabolic or toxic causes.[6 7] There was no evidence to suggest an hereditary form of pressure paresis.[8] The electrophysiological abnormalities correlate with the CRC present in the neck and in the left proximal forearm. Nerves at these sites may be vulnerable to compression or traction. The completeness and rapidity of the clinical recovery would favour compression.

Previous case reports have noted neurological abnormalities, namely, anaesthesia[2] and wrist drop[3] in the segment distal to CRC. In the first case quoted the sensory changes were not noted until after infection of the distal tissue and had recovered in part after six months. It is possible that this too represents an entrapment neuropathy. The experience in our patient raises the question whether in some older children or adults entrapment neuropathies may be due to CRC, no longer apparent to superficial examination.

References

1 Torpin R. *Fetal Malformations caused by Amnion Rupture during Pregnancy.* Springfield: Charles C Thomas, 1968.
2 Barenberg LH, Greenberg B. Intrauterine amputations and constriction bands. *Am J Dis Child* 1942; **42:**87–92.
3 Meyer H, Cummins H. Severe maternal trauma in early pregnancy. *Am J Obstet Gynae* 1941; **42:**150–3.

4 Baker CJ, Rudolf AJ. Congenital ring constrictions and intrauterine amputations. *Am J Dis Child* 1971; **121**:393–400.
5 Patterson TS. Congenital ring constrictions. *B J Plast Surg* 1961; **14**:1–31.
6 Sutherland S. In: *Nerve and Nerve Injuries.* Edinburgh: Churchill Livingstone, 1978, pp 656, 693, 871.
7 Karpati G, Carpenter S, Eisen AA, Wolfe LS, Feindel W. Multiple peripheral nerve entrapments. *Arch Neurol* 1974; **31**:418–22.
8 Earl LJ, Fullerton PM, Wakefield GS, Schutta HS. Hereditary neuropathy with a tendency to pressure paresis. *Q J Med* 1964; **33**:481–98.