A CASE OF NEURALGIC AMYOTROPHY OR SHOULDER-GIRDLE SYNDROME

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A syndrome comprising pain and flaccid paralysis of the muscles around the shoulder girdle and arm occurred fairly often during the war years 1941-45, though previously it had been rare. Parsonage and Turner reported 136 cases in 1948 among army personnel in the United Kingdom and in the India Command during the Second World War. This condition would seem to be rare in Singapore.

The essential clinical picture is simple. It starts with pain of sudden onset around the shoulder region. The pain may radiate down the outer side of the upper arm or into the neck. This pain lasts from a few hours to a fortnight or more, and then a flaccid paralysis of some of the muscles of the shoulder girdle and often of the arm develops; in some cases there is a patch of sensory loss over the outer side of the upper arm. When the paralysis appears, the severe pain usually stops, but a dull ache may persist considerably longer. There is usually no constitutional symptoms.

Of the 136 cases reported by Parsonage and Turner, the majority were in the age group between 20-40. 98 of the 136 cases had had either an infection or operation or trauma shortly before the onset of the disease.

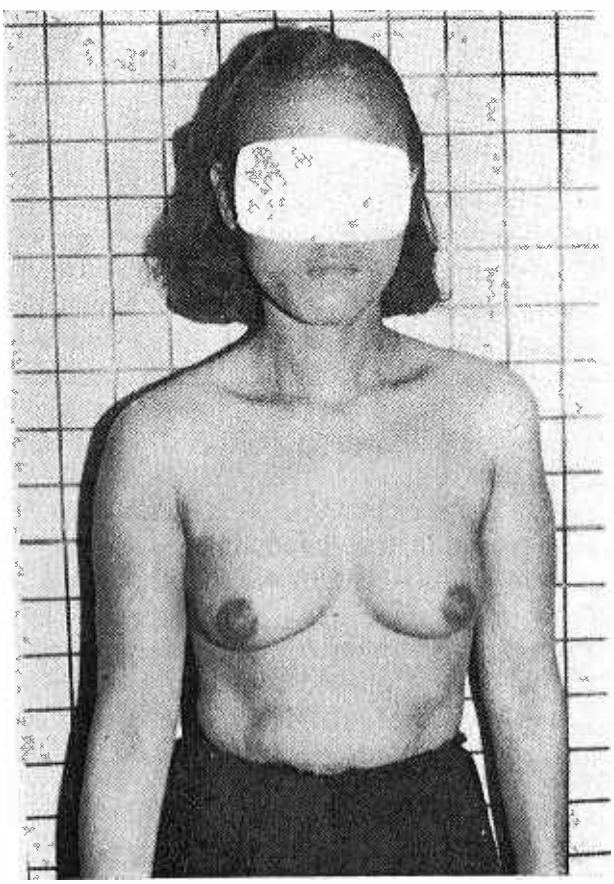
One or more nerves may be involved; involvement may be unilateral or bilateral. In single nerve involvement, the commonest nerve affected is the long thoracic leading to paralysis of the Serratus Magnus and winging of the scapula. Next in frequency are the circumflex nerve and the suprascapular nerve. In multiple nerve involvement the commonest combination is paralysis of the spinati and deltoid, usually with sensory impairment in the distribution of the circumflex nerve. In Parsonage and Turner's series, 57 cases had this combination. In 21 of their cases it was impossible to explain the muscle involvement in terms of a peripheral nerve lesion; there was a patchy muscle wasting and weakness which did not correspond to the distribution of any combination of peripheral nerves or nerve-roots. These cases would seem to indicate a cord lesion. The present case illustrates this.

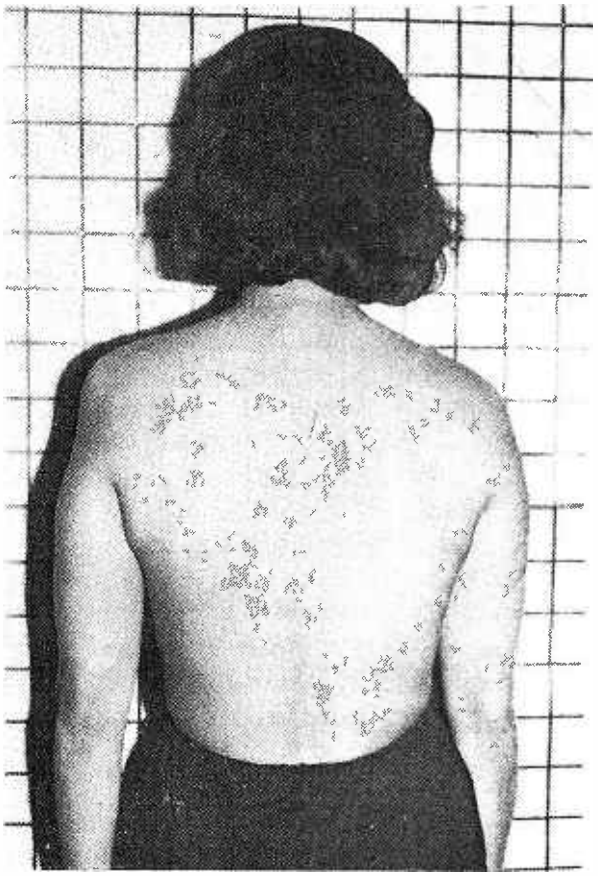
CASE REPORT

A Chinese female age 30, presented on 15.11.65 with a severe pain radiating down her left shoulder to the elbow of one month's duration. The pain started two weeks after the delivery of her first child. Five days after the onset of pain she noticed weakness of her left arm. There was no constitutional symptoms. On examination, she was afebrile. The left deltoid was wasted and there was fairly extensive paralysis of certain groups of muscles of her left upper limb.

MUSCLE CHART

Left Arm	**19-11-65**	**10-2-66**	**12-4-66**	**30-3-67**
Serratus Magnus	5	5	5	5
Pectoralis Major	4	5	5	5
Latis, Dorsi	4	4	4	5
Spinati	2	3	3	5
Deltoid	2	4	4	5
Biceps	4	4	4	5
Brachialis	4	4	4	5
Pronator Teres	3	3	3	5
Ext. Carpi Radialis	0	2	3	5
Ext. Carpi Ulnaris	0	2	2	5
Ext. Comm. Dig.	0	0	2	5
Ext. Pollis Longus	0	0	0	5
Flexor Prof. Dig.	3	3	3	5
Flexor Sub. Dig.	0	0	3	5
Flexor Pollicis Longus	0	0	3	5
Abductor Pollicis Longus	0	0	0	5





Figs. 1 & 2 show severe wasting of the left Deltoid

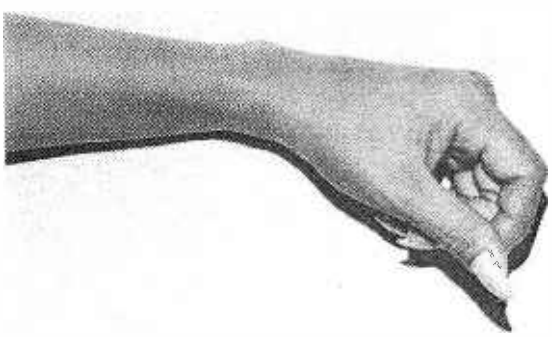


Fig. 3 shows weakness of finger flexors.

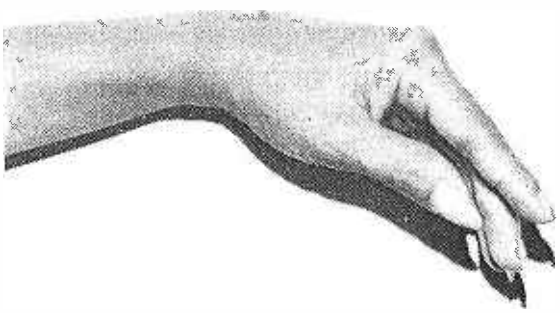


Fig. 4 shows weakness of wrist & finger extensors.

The muscle chart showed a patchy pattern of muscle paralysis consistent with a cord lesion rather than a peripheral nerve involvement.

There was a zone of impaired sensation to pin prick and cotton-wool on the outer aspect of the deltoid region and upper arm. Her lumbar puncture showed no C.S.F. abnormality. The Kahn test was negative. A diagnosis of shoulder girdle syndrome of Neuralgic Amyotrophy was finally made after consultation with Professor Gordon Ransome.

The pain was reasonably well controlled by disprin but did not disappear till January 1966. By 3.2.66 the sensory loss had disappeared. Her motor power improved very gradually but it was not till the end of February 1967 that she regained normal strength in all her muscles.

DIFFERENTIAL DIAGNOSIS

1. *Anterior Poliomyelitis:* In the early stages of the shoulder girdle syndrome constitutional symptoms are absent, the C.S.F. is normal and in many there are mild sensory changes. Cases seen after the acute phase may well be mistaken for old cases of poliomyelitis.
2. *Prolapsed Cervical Intervertebral Disc:* Cervical disc protrusion usually affects only one root—usually C6 or C7. The essential feature is pain though slight weakness and wasting of the muscles innervated by these roots may be present. However, the profound weakness and atrophy of the shoulder girdle syndrome do not occur.
3. *Brachial Neuritis:* This diagnosis used to include several conditions. As in sciatica neuritis most of these turned out to be due to cervical disc syndrome or spondylosis. True brachial neuritis must be rare; used in its strict sense it implies a diffuse affection of the brachial plexus, leading to pain in the arm, slight generalised weakness, diminution of tendon jerks and diffuse sensory changes.

ETIOLOGY

This remains obscure. Viral infection has been suggested but there are features unusual with virus infections of the nervous system, particularly the absence of constitutional symptoms and the normal C.S.F.

A similar condition has been known for many years as an occasional complications of serum injection. This usually develops from 7 to 10 days after the serum has been given. It starts with severe pain across the shoulder and upper arm, which lasts several days and is replaced by an atrophic palsy, normally affecting muscles supplied by C5-C6 roots. The pathology of these cases is believed to be a perineural oedema of the affected roots or nerves, comparable to urticaria of serum sickness.

There is such close clinical similarity between the serum cases and those of the Shoulder Girdle Syndrome that it is reasonable to assume that they are of similar etiology.

PROGNOSIS

Parsonage and Turner stated that muscles not completely paralysed during the acute attack, or muscles showing some return of voluntary power during the first month will usually recover completely in 6 months or less. They also said that when severe wasting occurs early, the prospect of useful power returning is poor. Dixon and Read (1945) followed up 16 cases for two years. By the end of this period all but 2 of these cases had shown very marked improvement.

The present case under discussion had rapid and severe wasting but recovered completely although she took nearly one and a half years to do so.

It would therefore appear that the prognosis for recovery even in completely paralysed muscles is not necessarily as poor as stated by Parsonage and Turner.

SUMMARY

A case of Neuralgic Amyotrophy or Shoulder Girdle Syndrome was presented. A brief review of the literature was given and the salient clinical features of this condition discussed.

REFERENCES

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