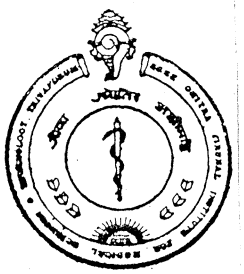

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PROJECT REPORT

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NEURALGIC AMYOTROPHY CLINICAL PROFILE, ELECTROPHYSIOLOGIC FEATURES AND LONG TERM PROGNOSIS

NAME : **Dr. ROBERT MATHEW**

PROGRAMME : **D.M. NEUROLOGY**

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ROBERT MATHEW

CERTIFICATE

I, Dr. ROBERT MATHEW hereby declare that I have actually performed all the procedures listed/ carried out the project under report.

Signature.....

Place : Trivandrum

Date : 11.11.2000

Name in capital letters

ROBERT MATHEW

Forwarded. He has carried out the project under report.

Signature
Head of the Department

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NEURALGIC AMYOTROPHY CLINICAL PROFILE, ELECTROPHYSIOLOGIC FEATURES AND LONG TERM PROGNOSIS

ROBERT MATHEW

CONTENTS

INTRODUCTION

Neuralgic Amyotrophy (NA) is a clinical entity charecterised by acute or sub acute onset of pain either accompanied or followed by weakness and often wasting of various forequarter muscles especially those about the shoulder girdle. This is an uncommon clinical disorder of unknown aetiology. Neuralgic anyotrophy occurs in both familial and non familial forms. The non familial sporadic type has a complex history. A subset was first mentioned in literature as early as the mid 1800, when it was called Serratus palsy. In the late 1800s it was a known complication of various infectious disorders and was called post infectious paralysis. Subsequently after serum therapy and vaccines were introduced it was linked to them as one of the serogenic and vasogenic neuropathies. Soon after 1900 many isolated cases of neuralgic amyotrophy were described in the literature attributed to causes as occupational neuritis, diabetes and should joint arthritis. During world war II when sufficient cases occured among Allied military personal neuralgic amy otrophy became a well defined disorder. This entity has a great number of synonyms (Table1), in part because it was described by several diferent investigators over a relatively brief time each of whom bestowed a diferent name upon it and in part because its pathogenesis and localization along the peripheral neuraxis remained uncertain. Most of the authors prefer the descriptive tittle Neuralgic Amyotrophy because it conveys no assumptions

regardings the etiology or site of the lesion. (1, 20, 21, 25, 32). Neuralgic amyotrophy has an estimated annual incidence of 1.64/100,000 population. Available data on the main features of NA comes from case reports and series drawn from selected referal centers as well as from occasional outbreaks(2,27,35). Information on incidence of the disease in deliniated population is very much limited(35).

The inherited form of Neuralgic Anyotrophy was first reported by dreschfeld in 1887. When familial it is inherited as a dominant trait and affected family members often have recurent attacks. Other features that distinguish this type of NA from sporadic type are equal male: female involvement, younger age of onset extensive shoulder girdle involvement abortive attacks with pain in shoulder or arm, presence of dysmorphic features, involvement of other portions of the PNS such as lumbosacral plexus, lower cranial nerves and various mononeuropathies on other extremities (1). Apart from pain, sensory synptoms and signs are much less prominent than loss of onset of motor functions. Maximal weakness may persist for several weeks or months during which time prominent atrophy and fasciculation may develop. Recovery begins weeks to months after onset of symptoms and proceed usually without interruption over a long period of time. The degree of recovery is usually excellent (1).

Although clinical presentation of amyotrophy is distinctive in general terms specific features can be quite variable(2,6,21,25,32,37). The diversity of the disease can be striking and is often not appreciated.

Because of prominent involvement of shoulder girdle muscles, some authorities consider this entity as brachial plexus neuropathy(2). Indeed a brachial plexus localization usually in an upper trunk distribution usually does sometimes occur(21). However detailed clinical and electromyographic examinations of patients often indicate lesions of induvidiual nerves or nerve branches, many of which are not compatibile with a brachial plexus localization (2,6,21,25,32,37). In this study the clinical features and outcome of 24 patients with Neuralgic Amoytrophy is analysed.

REVIEW OF LITERATURE

History

In Berger's thesis (1873), the 40 previously published reports were collected, and a comprehensive discussion of publications of 1873 up to 1942 was provided by Gathrier and Bruyn (1960). Over the century spacing 1850 - 1950 the clinical effects of serratus anterior palsy on arm abduction and elevation was noticed and during this period serogenic neuropathy also emerged as a distinct entity. Finally the increased prevalence of the disorder among the armed forces during the second world war was noted by Richardson (1942), Burnard and For (1942), Spillane (1943) and Turner (1944). Parsonage and Turner (1948) largely shaped today's still indefinite concept of neuralgic amyotrophy (40). They described 136 cases and in view of the doubts about it's pathology and aetiology, gave it the descriptive name neuralgic amyotrophy. These reports were concerned with service personal in whom long term follow up was impracticable. In 1957, they further described 82 patients seen between 1947 and 1955 at St. Barthelomeow Hospital, with special reference to prognosis(25). In 1972, Peter Tsairis et al analyzed 99 patients with brachial plexus neuropathy and described the outcome of 84 patients (2). The first (and perhaps the only) population based study was conducted by Ettore Beghi et al in 1985. The survey was conducted by reviewing the clinical records of Rochester, Minnesota, residents for the period 1970 through 1981 (35). In 1987, John D. England and Austin . J. Sumner described nine cases of NA (neuralgic amyotrophy) whose

clinical and electrophysiological findings suggested lesions of individual peripheral nerves or peripheral nerve branches occurring singly (mononeuropathy) or in various combination (mononeuropathy multiplex). This study highlighted the increasing diversity of NA (21).

Inherited brachial plexus neuropathy

In 1886, Dreschfeld described a 43 year old women who had suffered 3 descrete episodes of painful upper limb weakness over a 3 year period. He used the term "Rheumatic peripheral neuritis" and also mentioned that the patient's sister had seven similar attacks. The sister's case was described in more detail by Ross and Bury. Since then a number of families have been described in which the clinical and historical hallmarks have been the familial occurrence of recurrent episodes of brachial plexus neuropathy(38).

Clinical Features

The clinical features of NA is so characteristic as to require no strenuous mental acrobatics on the part of the physician for correct diagnosis(40).The study from Rochester, MN by Ettore Beghi et al found an overall annual incidence of 1.64/100,000 population with the age at occurrence ranging form 12-47 years and a male to female ratio of 1.75(35). Large series from hospital based civilian population show a more even distribution of the disorder between the third and seventh decades and a male preponderance with M : F ratio ranging from 1.6 to 10.5 (15). A few reports have described non-familial NA as occurring in epidemic form but the vast majority of cases are sporadic (1). Apparent clusters have been reported from London and Czechoslovakia. Antecedent events

occur in 28-83% of cases. The events are listed in table 2. Various illness that have been described in association with NA is listed in Table 3. The most common single antecedent event appears to be infection in various series.The most common type of antecedent infectious disorder encountered in North America, England and Europe today is probably a non descript upper respiratory infection/ flu like syndrome. Such was noted in 25% of cases in Tsairus series of 99 cases.A coxsackievirus was considered the etiologic agent in the 1953 epidemic of czechoslovakia. In 2 early series,aproximately 50% of Patients became symptomatic while hospitalised.. The hospitalization can be for various medical disorders, for almost any type of surgical orthopedic procedure or for child birth. Unfortunately many physicians are unaware of these links and inappropriately attribute the occurrence of NA in these situations to such iatrogenic factors or improper positioning on the operating / delivery table, to radiation therapy and to neuroradiologic contrast media. This is potentially hazardous practice, considering the litigionus nature of modern society. Also for unclear reasons, many orthopedists, neurosurgeons and sports physicians are unfamiliar with the disorder consequently instances of probable NA can be found in their literature, attributed to traumatic compressive and stretch injuries of various nerves and even to inflaclavicular plexus perforation by blood vessel anomalies(1). Of the various associated illness, diabetes mellitus formed the commonest single illness .A nontramatic brachial plexopathy has also been reported with intravenous addiction to heroin, but it tends to be painless and to have a predeliction for lower plexus(15)

Typically few days to 2 weeks or more pass after the antecedent

event before the onset of the disorder. Pain sets in acutely, often awakening the patient at night or being present on awakening(15). Most commonly the pain is experienced around the shoulder and upper arm, but in some cases, particularly those in which the musculocutaneous and the anterior interosseous nerves are involved, there may be a good deal of pain in the elbow and forearm. The pain may spread to the entire arm (40). At times there is B/L shoulder pain although paralysis may appear only in one arm (25). The pain is often aggravated by movement of the shoulder but not by movements of the cervical spine or coughing or sneezing . There may be considerable tenderness (40). The pain is severe, sharp, deep lancinating boring aching or throbbing (15,40).The pain may prove to be refractory to the usual analgesics. The patient typically immobilises the limb splinting it in adduction and internal rotation(1). Exceptionally the pain may be trivial or even absent (40). Severe pain generally persists for from several hours to approximately 2 weeks, and then abates, frequently being replaced by an emerging ache that can last for months(1). The pain stage lasts less than 2-3 weeks in 90% of cases(40).

Weakness usually is noted as pain lessens, most commonly 1-7days after onset, occasionally in 7-14 days. In 2 large series, weakness was noticed within 2 weeks of pain onset in more than 2/3 of patients (1). Weakness is usually maximal at the onset but can slowly progress over a week or even longer. The specific muscles affected vary strikingly from one patient to another. The deltoid (31-100%) serratus anterior (17-74%) infraspinatus and supraspinatus (52-91%) are more frequently involved, but many different forequarter muscles may be affected either

singly or in a biweldering number of combinations. The muscle involvement variably suggests compromise of branch of nerves to individual muscles; single peripheral nerves; multiple peripheral nerves or portions of the plexus particularly the upper trunk and cervical roots, especially C5 and C6. Occasionally the diaphragm also is affected (1).Weakness confined to a single nerve distribution or multiple nerve distribution also appears common. The long thoracic nerve is most often involved, where as isolated suprascapular and axillary nerve weakness is less common. Rarely the weakness is confined to the distributions of musculocutaneous nerve, anterior interosseous nerve or radial nerve(15). The variability and focality of neuralgic amyotrophy is exemplified further by the occasional affection of nerves such as median, posterior interoseous or lateral antebrachial cutaneous nerve as well as cranial nerves IX, X, XI, XII. On rare occasions, remarkable focality such as isolated denervation of the serratus anterior muscle is seen. Thus many authors now regard NA as a form of mononeuropathy multiplex involving mostly the nerves of the upper extremity but occasionally involving the other nerves also(36). Rarely NA can present with phrenic nerve involvement as the sole manifestation (32). Due to the flaccid paralysis, spontaneous subluxation of humerus may occur(40).Moderate to severe atrophy follows weakness in majority of the patients.

In the series of Tsairis et al U/L diaphragmatic palsy occurred in 6% of the individuals and B/L diaphragmatic palsy in 1% of patients(2). The side of the diaphragmatic weakness may not correlate with the side of severe arm weakness. The physician should think of this immediately when the patient complains of acute shortness of breath.

The most fascinating aspect of neuralgic amyotrophy is the finding that one or more muscles may be spared in the innervational distribution of a single root, trunk or cord . One may for example find atrophy and wasting of infraspinatus muscle, without any abnormality of supraspinatus or paralyzed rhomboids with a normal levator scapula. It is impossible to ascribe involvement of serratus anterior muscle to a lesion of brachial plexus itself. These findings emphazied by O' Brain and Payan 1980(23) and England and Summer (21) confirmed with the electrophysiologic findings of Morten and Craft(1974) point to the hypothesis that we are dealing with Mononeuritis multiplex instead of plexus neuropathy or even radiculopathy.This had already been suggested by Spillane in 1943(40).

The findings on neurologic examination are primarily confined to the motor system. The patient typically immobilizes the limb, splinting it in adduction and internal rotation (1). Waxman(26) emphasized this in the flexion adduction sign, which means that the patient holds his affected arm in flexion at elbow and with adduction and internal rotation of the upper arm (40). Kennedy and Resch noted that many patients maintained a rigid posture at the shoulder because movement caused discomfort and that this sometimes led to a frozen shoulder syndrome. One reasonable explanation for the position assumed by patients with brachial plexus or cervical root lesions is that the flexion-adduction posture reduces mechanical tension on the affected nerve roots or plexus. When the shoulder is abducted and laterally rotated head becomes prominent, displacing the brachial plexus forward and exerting tension on the roots from which the plexus arises. In many patients who exhibit

the flexion-adduction sign, there may be resistance to extension of the elbow when the arm is elaveted at the shoulder (Bikele sign) which as noted by De Jong is similar to the Kernig's sign and is probably due to tension on the irritated nerve roots. The Bikele sign is also occasionally observed in patients with cervical disc disease.

There is no evidence for the alternative hypothesis that the flexion-adduction posture results from a preferential pattern of weakness (26).

The findings on neurologic examination are primarily confined to the motor system (1). The incidence of objective sensory loss has ranged from 33-70% in various series (15) and it tends to be inconspicuous. A numb patch over the outer aspect of shoulder and diminition of sensation in the distribution of the radial nerve are the most common patterns(15). Hyperalgesia in which even a slight touch may provide unbearable buring pain, has been mentioned. Less frequently diminished cutaneous sensation in the distribution of median nerve is found. Radicular pattern of sensory loss seldom or perhaps nerve occur (40). In the series by Tsairus et al, sensory loss was associated with 80 of 133 plexus lesions (66 of 99 patients). Another 16 plexus lesions were associated with subjective cutaneous sensory loss. The objective sensory loss was of the mixed variety (superficial cutaneous and proprioceptive sensation) and did not always parallel the motor deficit. The outer surface of the upper arm (in the distribution of the circumflex nerve) and the radial surface of the forearm were the most commonly affected areas. Proprioceptive sensory loss was present in the most severely affected limbs, the sensory abnormalities were of lesser magnitude than the motor abnormalities (41). Diminshed cutaneous senastion was found

less frequently in the distribution areas of radial or median nerve. Radicular pattern of sensory loss seldom or perhaps never occured(40). To some extend the lack of sensory loss may be related to the frequency with which purely motor nerves such as suprascapular and long thracic are involved in this process(15).

The deep tendon reflexes are variable. They are usually normal but become hypoactive or absent if the appropriate muscles are denervated substantially(1,15).

1/4 to 1/3 of patients may have bilateral signs and symptoms. Usually asymmetrical (2,15). Parsonage and Turner reported a higher number of cases in the right (especially for serratus anterior weakness) but other studies have not confirmed these observation (15,25). In some patients with unilateral involvement, EMG abnormalities are found bilateraly(25). In the series of Tsaris et al, bilateral involvement was recognized clinically in 1/3 of cases but EMG examination uncovered another 11 instances. Bilateral involvement with an occasional interval of some weeks between the two sides of the body has been stressed by Paw-seradel. (40). When bilateral, the involvement is usually sequential rather than simultaneous and is asymmetrical. In many series, the paralysis manifest more frequently on the right than on the left side(53-95%) for unclear reasons (1). Initially this was attributed to right handedness, the weight of the soldiers rifle bearing preferentially on the right side, but these arguements were shown to be fallacious (40).

The CSF examination generally is unremarkable with occasional pleocytosis and /or mild total protein increase when antecedent infection or systemic disease occured.

Routene laboratory studies are usually normal, Cervial spine X-ray and cervical myelograms are either normal or show non-specific abnormalities. Spinal fluid is usually normal although a modest elevation of proteins may occur. A high level of CSF IgG has also been reported(15,27). Solbergs (1976) series of 41 cases is distingushed by a rather high prevalence of CSF abnormalities (5cases) but the series was heterogenous.

Several studies have dealt with the EMG aspect of neuralgic amyotrophy (27,36),and recently its electrodiagnostic evaluation has been reveiwed(33).A plexus lesion can be distinguished from a radicular or more proximal lesion by a reduction in the amplitude of an appropriate peripherally recorded sensory nerve action potential (SNAP). a The lack of abnormalities of paraspinal muscles are sometimes difficult to detect because of difficulty in obtaining adequate relaxation. Also proven radicular lesions may occur without abnormalities appearing in the paraspinal muscle altough this tend to occur in general with chronic lesions (15,39). The SNAP amplitudes are quite sensitive to axonal loss lesion , but the lesions have to involve those parts of the plexus that supply fibre to nerves from which SNAPS can be routinely recorded. In general lower trunk and medial cord lesions cause abnormal ulnar sensory responses, recording from the middle and often the index fingers. Upper trunk lesions are more difficult to evaluate and abnormalities can often be only detected by recording responses from less commonly studies nerves such a lateral antebrachial cutaneous nerve and median nerve recording from thumb ; occassionally the radial and rarely the median (recording from index finger) SNAPs will be of low amplitude as well(15, 33). Lateral cord lesions can be

distinguished from posterior cord lesions by involvement of the median sensory responses in the former and radial sensory responses in the latter. The considerable variation in the distribution of specific root fibers to various digits has been documented by direct recording. It is important to compare amplitudes of SNAP on the two sides to detect subtle lesion(15).

The distinction between lesions of various parts of the brachial plexus as well as between lesions of the brachial plexus and those of more peripheral nerves is made by the distribution of SNAP and EMG abnormality(15).

Over all the available EMG studies suggest that neuralgic amyotrophy is caused by axonal loss lesions involving either the brachial plexus or multiple nerves originating from the brachial plexus. Routene median and ulnar motor nerve conduction studies (NCS), recording from hand muscles are usually normal even with supraclavicular stimulation. However infrequently the lesions affect motor fibers in lower plexus distribution, causing low amplitude median and ulnar motor responses and sometimes mild slowing of median and ulnar nerve conduction velocites. Even when there is clinical involvement of the lower plexus median and ulnar motor nerve conduction study using supraclavicular stimulation may be normal. Motor NCS recording from the proximal muscles such as trapezius supraspinatus, infraspinatus and deltoid, using supracavicular stimulation have been reported to show slightly to markedly prolonged latences(15,27,36).

In contrast to the motor NCS, 1/3 of 16 patients reported by Flogman Kelly had median, ulnar and radial SNAPS of low amplitude

(36). In Newmans study of six patients with involvement in the distribution of the middle or lower plexus, only two had abnormalities of either median or ulnar SNAPs. Thus, although the high incidence of normal SNAPs is often explained by the usual predeliciton for the syndrome to affect the upper plexus this is not always the case. Subramony SH and Welbourn AJ have noted that SNAPs are often intact when muscles sharing their peripherial and segmental innervation are clearly involved. The value of more extensive studies (lateral antebrachial cutaneous nerve,median sensory response recording from the thumb) has not been adequately explored in this entity(15).

Other NCS abnormalities reported include an occasional increase in F wave latency to hand muscles, despite normal perpheral conduction velocity and the lack of sensory response with supraclavicular stimulation in the presence of a normal perpheral sensory study. Paraspinal muscles are in general normal (36). The distribution of the abnormalities reflects the distribution of the weak muscles although clinically unaffected muscles may show abnormalities as well. About 50% of patients with clinical unlateral involvement have bilaterel abnormalities on needle EMG (2,36). Although it is often possible to localize the lesions to predominantly one or the other part of the plexus using the nerve conduction siudies and needle EMG abnormalities often a strict anatomic localization is difficult.

The EMG study in neuralgic amyotrophy often shares abnormalities similar to those seen with other causes of brachial plexus

lesions. However bilateral abnormalities and the ocurence of anatomically difficult to explain distribution of needle EMG and SNAP abnormalities favour neuralgic amyotrophy. With an upper plexus lesion EMG may not always exclude a radiculopathy and radiological studies may be indicated . Somatosensory evoked responses have also proven unreliable for making this distinction.

AIM OF STUDY

To study the aetiology, clinical profile and prognosis of patients with Neuralgic Amyotrophy.

MATERIALS AND METHODS

The study was a retrospective and prospective study in which 25 patients diagnosed as having Neuralgic Amyotrophy attending the Neurology out patient department during the period 1995-1998 were included.

Inclusion criteria

I) Acute or sub-acute onset with non- progressive course of induvidual episodes (Recurrent episodes with residual deficits even though had the profile of a progressive illness was included since the induvidual episodes were non-progressive .)

2) A case was included only when definite weakness due to peripheral nervous system damages could be localized to the arm (on one or both sides) with no demonstrable sign of spinal cord, primary root or symmetric peripheral nerve involvement.

3) Electromyographic evidence of axonal damage or denervation of muscles of brachial plexus.

4) Occurrence of pain at the onset of the disease represented a contributory finding, but absence of pain was not considered a basis for excluding an otherwise typical case.

Exclusion criteria

1) Patients with specific causes of brachial plexus damage such as direct trauma, compression or radiation.

2) Patients with a progressive course.

3) Patients with normal electrophysiologic study

The clinical details and Electrophysiological findings were entered into a prestructured proforma. The patients were called back for a review during which the history of the episode was clarified and detailed examination done for any focal residual neurological deficits. Details regarding any relapse (if present) were collected.

RESULTS

Age and Sex distribution: - The age ranged from 10 years to 54 years. The mean age was 27.4 years. 19 were males and 6 were females, male: female ratio being 3.17:1. In none of the patients was a family history obtained.

Antecedent and Associated illness- Table 4

14 (56%) of patients gave no history of antecedent illness, immunization or exposure to toxic substances within one month of onset of their illness. A history of antecedent fever was given by 6 patients of which one was Gastrointestinal infection and one was Upper respiratory tract infection and 4 patients did not notice any specific systemic symptoms . History of trauma was available in 4 patients.Of these patients 3 had direct trauma to the affected limb .One patient had developed the symptoms within one month of HBs Ag vaccination.None of the patients gave a history of preceeding surgery. One of the patients had consumed alcohol heavily on the day prior to the onset of symptoms. Three patients had diabetes mellitus before onset of illness, and one patient had a sensorimotor peripheral neuropathy whose aetiology was not clear from investigations.

Mode of Onset and initial symptoms

20/25 (80%) patients had pain as the initial symptom.In the remaining 5 patients 2 had no pain at all and 3 had pain subsequent to onset of weakness. The weakness was of acute onset (considerable weaknesses in less than 24 hrs) in 5. The time taken from onset of

weakness to attainment of maximum weakness ranged from as short as 2 days to as long as 30 days. 8 patients had involvement of the right side ,12 had involvement of left side and 5 had bilateral symptoms(symptoms occurring in both sides within one month of onset of illness).

Symptoms:- Pain was the initial symptoms in 20/25 (80%) patients. In 10/25 (40%)the pain was of sudden onset (considerable pain in first 24 hours itself). The pain was aggravated by arm movements in the majority of patients. One patient reported worsening of the pain on coughing and sneezing. The pain was localized to the root of neck in one patent . In others it was localized to the shoulder, arm, forearm or to more than one of the above areas. In all the patients the pain remained confined to the site of weakness.After an initial period of severe pain lasting for 2-3 weeks , most of the patients had improvement in the symptom .In some patients however it persisted at a reduced intensity for up to 3 months continuously or episodically.

Examination findings - All the patients included in the study had weakness at the time of initial examination. The severity of weakness varied considerably. In all the patients, shoulder girdle was involved. All the patients had axillary nerve involvement. In none of the patients was the weakness confined to distribution of one single nerve territory. Most of the diffuse plexus lesions were incomplete in that there was sparing of one or more muscles in the same root distribution. In 5 patients with B/L involvement asymmetry was seen in severity of involvement in 3 patients and in extend of involvement in 2 patients. One patient had diffuse B/L involvement. The deltoid and spinati were most commonly affected.The distribution of weakness is given in table 5. Wasting was seen in 13

patients of which it was mild in 8/13 and profound in 5/13. Only one patient had fasciculations and he did not report any improvement in power even after 3 year of illness. Clinical evidence of diaphragmatic or vocal cord involvement was not seen in any of the patients.

Sensory Loss - Sensory loss was seen in 10 plexus lesions (8/25) patients). Touch as well as pain sensation was impaired in the involved areas. In 6 lesions the area of distribution corresponded to the axiliary nerve teritory, and in 4 lesions in the territory of lateral cutaneous nerve of forearm. In one patient sensory loss was in the medial aspect of arm. The sensory abnormality was always of less magnitude when compared to motor abnormalities in the affected limb. One of the patients had glove and stocking type of sensory loss which he had developed prior to the development of the plexus lesion. There was no obvious cause for the peripheral neuropathy. He however did not have sensory loss corresponding to the plexus lesion.

In most of the patients, deep tendon reflexes in the involved side was either sluggish or absent depending on the extend of lesion.

Electrophysiology

All the patients had underwent nerve conduction studies, and EMG studies when required. The studies were done as early as 15 days to as late as more than 10 years after onset of illness. In all the patients, the findings were complementary to the clinical localization. In none of the patients, electrophysiologic abnormality could be detected in a clinically normal side. 30 Abnormal plexuses were studied . The

commonest abnormality in motor nerve conduction study was in axillary nerve (13 patients). Lateral cutaneous nerve SNAP abnormality was detected in 8 patients.

EMG evidence of denervation was seen most in the deltoid (14 patients). The electrophysiologic study was not done in a strictly standardized way in all patients. The consultant had the freedom to alter the study according to his needs and in some of the patients sampling of all concerned nerves and muscles were not done either because of poor patient co-operation or because the diagnosis was otherwise obvious.

Laterality and Types of Plexus lesion:

At the time of initial examination 5/25 had B/L involvement and one patient was already having significant weakness of C/L limb consequent to brachial plexitis which he developed 10 years prior to the present episode. Of the 20 patients with unilateral lesion, 8 had right sided lesion and 12 had left sided lesion. None of the patients had electrophysiologic abnormality in the uninvolved side.

Taking into consideration the clinical findings as well, all the patients except one had definite evidence of upper plexus involvement. Out of a total of 30 electrophysiologically abnormal plexuses, 11 showed evidence of diffuse involvement. Only in one patient electrophysiologic abnormalities were confined to lower station. This patient on clinical examination had evidence of upper trunk involvement as well - however she was having arthritis of the corresponding shoulder joint and hence proper muscle power testing was not possible.

Lab Studies

MRI scan of the cervical spine was done in only 2 patients which was normal except for incidental intervertebral disc prolapse which was seen in one patient without radicular involvement. CSF study was not done in any of the patients.

Treatment

13/25 patients received steroids. The exact treatment history of many of the patients who had registered quite late into the illness could not be ascertained. Also the dosage of steroids was not standardized and it was given at varying periods into the illness for variable duration. Many of the patients had received treatment at least for a duration of 7 days. Steroids were given only when the patients were seen in the acute phase of illness.

Follow up

Duration of follow up of patients ranged from 0.5 months to 166 months of onset of illness. The duration of follow up was calculated from the time of onset of illness to the period of first review.

The recovery was classified as total recovery, incomplete recovery or no recovery.

2 patients had complete recovery. One of the patient had complete recovery in1year while the other patient took 4 years for complete recovery. The extend of weakness in these 2 patients were more or less the same. The patient who had faster recovery had B/L involvement and was exposed to steroids. 3 patients had no recovery at all.

Patients were divided into 4 broad groups based on duration of follow up. Those who had completed less than 6 months (4) 7-12 months (4), 13-24 months (8), 25-36 months (0) and more than 30 months (6). Proper follow up data as regards further characterization of exact extend of improvement was not available in 5 patients..

In the 13 months - 24 months follow up group (which was the largest available group, all the patients had 50% or more improvement in power and half of them had 75% improvement.

In the >36 months group 2 patients had complete recovery but 3 patients (60%) had 25% or less improvement in power.

DISCUSSION AND CONCLUSIONS

The clinical features of Neuralgic Anyotrophy is very much similar to that described from the western world.

The clinical features in most of the situations are characteristic enough to make a clinical diagnosis of NA.

Associated pain a/c or sub a/c onset and upper trunk or diffuse involvement are strong pointers towards the diagnosis.

The disease appears to have a predilection to involve the upper trunk.

Axillary nerve conduction and EMG of deltoid appears to be most sensitive in picking up the lesion.

The disease is essentially self limiting, majority of the patients improving in first year itself.

No obvious prognostication features could be detected

Recurrence is possible infrequently. The time, side or extend of recurrence is highly variable.

There is no definite evidence to define the use of steroids in treatment or prevention of relapse. A prospective study in the acute phase of NA has to be undertaken with a larger number of patients to define the same.

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TABLE 1

Various names used for Neurologic Amyotrophy
Acute brachial neuropathy
Acute brachial radiculitis
Acute brachial plexitis
Acute multiple neuropathy of the shoulder girdle
Acute scapulohumoral paralysis
Acute shoulder neuritis
Amyotrophic paralysis of the periscapular muscles
Brachial neuritis
Brachial plexus neuropathy
Cryptogenic brachial neuropathy
Idiopathic brachial plexus neuritis
Idiopathic brachial plexitis
Idiopathic brachial plexopathy
Idiopathic brachial plexus neuropathy
Localized neuritis of the shoulder girdle
Localized non traumatic neuropathy
Multiple neuritis of the shoulder girdle
Paralytic brachial neuritis
Parsonage-Turner syndrome
Serratus magnus palsy
Shoulder girdle neuritis
Shoulder girdle syndrome
Paralytic cervicobrachial neuralgia

Reference 1,26, 40

TABLE 2

Antecedent Events
Infections
Upper respiratory infection
Flue-like illness
Infectious mononeucleosis
Hepatitis
A/c Tonsillitis
Glandular fever
Pyelitis
Carbuncle
Viral pneumonia
Anti serum against
Diptheria
Scarlet fever
Streptococus
Phneumocci
Gonococci
Anthrax
Gas gangrene
Tetanus

Vaccines

- Typhoid
- Small pox
- Tetanus
- Pertussis
- Diptheria
- Influenza
- Pregnancy
- Parturition
- Threatened abortion
- Hospitalisation
- Trauma
- Compression
- Radiation

Treatment with

- Interferon
- Botox
- Cytosine Arabinoside

Reference 1,7,11,12,13,14,15,25,29,31,35 ,45

TABLE 3

Associated illnesses
Old Poliomyelitis
Diabetes Mellitus
Allergic history
Rheumatoid arthritis
Polyarteritis Nodosa
Temporal arteritis
Hodgekins Lymphoma
Ehler - Danlos syndrome

Reference 1,7,11,12,13,14,15,25,29,31,35 ,45

TABLE 4

Antecedent illness	No. of patients
Upper respiratory infection	-1
Gastrointestinal tract infection	- 1
Fever without any specific Systemic manifestation	- 4
Trauma	
Involving affected limb	- 3
Unrelated to offeted limb	- 1
Immunization	- 1
Pregnancy	- 1
Surgery	- 1
Intramuscular injection	- 1
Total	14

TABLE 5

Distribution of weakness	
Total number of Plexuses	- 32
Shoulder girdle muscles	- 32
Flexors of elbow	- 7
Extensors of elbow	- 5
Flexors of wrist	- 7
Extensors of wrist	- 6
Small muscles of hands	- 5

Clinical photograph of a patient with bilateral neuralgic amyotrophy, who had only mild improvement even after 3 years