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DIABETIC AMYOTROPHY

A Report of Two Cases

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Diabetic amyotrophy is a syndrome of bilateral, asymmetrical, proximal muscle weakness particularly in the lower extremities. The syndrome is associated with variable diffuse pain and atrophy and occasionally fasciculations with weight loss and most commonly seen in elderly diabetic males. The first symptom is a gradual muscular weakness and wasting in the legs, the muscles affected being in anatomical proximity (other muscles of the same nervous supply may be spared entirely). Usually the pelvic girdle musculature is affected first but eventually global involvement of the leg may make the patient bedridden. Occasionally, muscles of the trunk and upper limb may be affected. When atrophy is severe, the deep tendon reflexes are usually depressed, but occasionally they persist and are brisk despite marked wasting. Extensor plantar responses may be present. Pain accompanies the wasting and is usually poorly localized and diffuse. Sometimes it is present in the back or abdominal wall, in one of Garland's patients leading to abdominal exploration. Objective sensory findings are usually absent. The spinal fluid may have an elevated protein. Electromyography and biopsy findings are supposedly characteristic.

The syndrome was first described by Bruns in 1890. Although he described three cases of paroxysmal muscle weakness and wasting in elderly diabetic males he considered this a manifestation of diabetic neuropathy. Over the intervening years not too much interest was displayed in this condition although the neuropathies of diabetes were well recognized. In 1953 Garland and Taverner published a series of five cases and by 1955 had collected a series of 20, renewing worldwide interest in this subject. It has been our recent experience to document two more cases.

CASE 1 — The patient was a 68-year-old man who was known to have elevated blood glucose levels at least eight years prior to admission. He had not been following a diabetic diet and had not been treated with anti-diabetic agents. In the year prior to hospital admission he began to notice a gradual weakness and wasting of the muscles of both thighs with difficulty climbing stairs and getting out of low chairs and cars. About the same time he developed an ache in the left popliteal fossa radiating down the posterior aspect of the leg to the sole of the foot. This subsequently appeared in the right leg and did not seem to follow any nerve distribution. His weakness and pain fluctuated in the course of the year.

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In the two months prior to hospitalization he was placed on Orinase and noticed some improvement in leg strength. He also began to follow a diabetic diet and lost 12 pounds.

For about 20 years the patient had had psoriasis and received many forms of therapy. Since April of 1962 he had been taking Decadron, .75 mg. t.i.d. apparently without medical supervision and his appearance was somewhat suggestive of Cushing's syndrome. The psoriasis flared when he tried to discontinue this drug. There was no history of headaches, fits, paralyses or sphincter disturbances. The patient stated that his voice had decreased in volume in the previous two years.

On physical examination he was an alert, cooperative man in no acute distress appearing well nourished. Chest and heart examination were negative. The blood pressure was 180/100. The cranial nerves were intact with no evidence of diabetic retinopathy. He was left handed. Motor examination of the upper extremities was essentially negative. There was moderate wasting of the legs especially the quadriceps, more marked on the left. Weakness, of rather mild degree, was noted in both psoas, hamstrings and quadriceps, again more marked on the left. He experienced some difficulty in stepping up on a high chair due to weakness of the quads. Plantar flexors and extensors were fairly strong. No fasciculations or muscle tenderness were present. Sensory exam showed a slight drop in vibration sense below the ankles. The deep tendon reflexes were present and brisk including those in the lower extremities, the left knee jerk being slightly less active than the right. Babinski responses were absent. Abdominal and cremasteric reflexes were present and symmetrical. The patient’s gait was normal with good heel and toe performance.

Psoriatic plaques were seen in the genital region, and both axillae, and there was a hyperkeratosis of the palms and soles. Chronic radiodermatitis, the result of x-ray therapy for acne, was present on the upper back.

Following admission the patient was gradually weaned from steroids. He received intensive antipsoriasis care (topical steroid creams etc.). His diabetes was brought under better control (placed on a 1200 calorie diabetic diet and Orinase, tabs, one b.i.d.). Electromyography showed scattered areas containing many polyphasic potentials and occasionally some giant potentials with poor interference pattern. However, many areas tested seemed completely normal. A muscle biopsy of the vastus lateralis of the left quadriceps was performed. This showed normal muscle fibers without atrophy. A lumbar puncture showed clear fluid under normal pressure. The spinal fluid protein was elevated (102 mgs. per cent). Initially blood sugar levels were elevated but under more rigorous dietary supervision and Orinase these fell to normal levels. Serum transaminase was normal. ESR was 29. FBI was 6.4 micrograms per cent. Serology was negative. Metastatic survey was negative.

The patient's course in the hospital was uneventful. Two weeks after discharge he was still complaining of pain in the left leg without much change in the muscle strength. His psoriasis was worse.

CASE 2.—The patient was a 72-year-old man admitted from the GI Clinic complaining of constant right lower quadrant abdominal pain. His diabetes had first been diagnosed in 1946. He also complained of a weak left leg and back pain which had been present for two or three months necessitating the use of a cane for walking in the month prior to admission. He had been seen in the Out-patient Department six months earlier at which time he complained of right upper quadrant abdominal pain. At this time chest x-ray, IVP, proctoscopy and cholecystogram were negative and no cause was found for his symptoms. At the time of his hospitalization he was following a 2800 calorie diabetic and taking 32 units of NPH insulin daily.

On physical examination the patient appeared thin and frail. The chest was clear and the cardiac and abdominal examinations were negative. Blood pressure was 150/80. The cranial nerves were intact but a mild degree of diabetic retinopathy was present. Motor exam showed gross weakness and wasting of the quadriceps and hamstring muscles, more marked on the left with weakness of the triceps and biceps bilaterally and the hand grip was weaker on the right. Fasciculations were noted in both upper and lower extremities and trunk. Deep tendon reflexes were hypoactive but symmetrical in the arms. Knee jerks were depressed bilaterally and the ankle jerks were absent. Plantar responses were flexor. Vibration sense was slightly depressed below the ankles but the other sensory modalities were intact. The initial impression was amyotrophic lateral sclerosis. X-rays of the cervical, thoracic and lumbar spines showed degenerative joint disease with some narrowing of disc spaces at C-4-5 and L-5 S-1 with several Schmorl's nodes in the lumbar region. Upper and lower GI series were negative. X-rays of the sacrum in both knees showed minimal degenerative changes. The lumbar puncture showed clear fluid under normal pressure but the spinal fluid protein was elevated (115 mgs.%). Lange curve was normal. Initially blood sugar levels were poorly
Figure 1

Case 1. These views show the atrophy of the quadriceps especially the vastus medialis portion. The calves are involved to a lesser extent.
controlled but these values fell to normal with more intensive antidiabetic therapy. The patient's stools were repeatedly positive for blood. Investigations including proctoscopy showed no cause for this or the abdominal pain and he was discharged.

Over the next six months the patient was followed in the OPD and his weakness was noted to improve to the point where he was able to discard his cane. The fasciculations which were originally diffuse disappeared. In retrospect he was felt to have a diabetic amyotrophic syndrome.

It is apparent from the second case that this diagnosis may be reached only after a considerable period of follow-up. The first patient was probably a mild diabetic since he had not been following any diabetic regimen despite a history of hyperglycemia eight years previously. In addition he was taking Decadron which of itself can cause muscular wasting and occasionally peripheral neuropathy and which can aggravate the diabetes. It is however, unlikely that the steroid caused the myopathy here in view of the asymmetry of the atrophy, the presence of pain and the improvement in his strength noted while still taking the Decadron. The patient noticed improvement in the weakness in the two months prior to admission. Although it may have been fortuitous he had been placed on Orinase and perhaps his diabetes was under better control during this period. A search for occult malignancy and other metabolic causes of myopathy was fruitless.

The second patient's diabetes was more severe and of longer duration. Here again poor control was evident although he had never experienced ketosis or coma. The presence of fasciculations with gross atrophy and weakness lead to the erroneous diagnosis of amyotrophic lateral sclerosis a conclusion proven false by the passage of time and the subsequent remission of the weakness. Again the prominent muscle wasting was proximal in the lower extremities. In neither case was there significant evidence of sensory neuropathy.

In his paper on diabetic neuropathies Sullivan describes two major types, first the bilateral, symmetrical, distal sensory neuropathy and second the asymmetrical motor neuropathy. The latter he postulates could be due to a primary occlusive disease of the vasa nervorum (characterized by abrupt onset), while the former is the result of the long continued disordered metabolism of diabetes (gradual onset). He states that the motor neuropathy is more apt to occur in mild or even unsuspected cases while the symmetrical sensory type is seen in long standing poorly controlled cases as shown by frequent concomitant retinopathy and nephropathy. Amyotrophy is similar to the motor neuropathies in that it may occur in mild or unsuspected cases. It is often confused with the motor neuropathies and it is important to remember that with amyotrophy the muscles affected are in anatomical proximity and do not necessarily have a common nerve supply. The early loss of deep tendon reflexes is in favor of a neuropathy (either sensory or motor) whereas in amyotrophy the loss is delayed until marked wasting occurs. Indeed, in some cases of amyotrophy the muscle wasting can become quite marked without loss of reflexes (as for example our first case).

At present the exact cause of diabetic amyotrophy is unknown but other factors than hyperglycemia must govern its occurrence since it is much rarer than the other
neurological complications of diabetes. According to Locke, Lawrence and Legg the site of the lesion could possibly be the myoneural junction with a selective action on part of the latter by some metabolite. In their paper stressing the pathological features of the disorder they emphasize that the characteristic histological picture is single muscle fiber atrophy in contrast to the motor unit atrophy of neuropathy and myelopathy. In our first case we were unable to corroborate this view since the muscle biopsy showed normal fibers. Garland states that the lesion could be in the spinal cord, in the nerve roots, peripheral nerves or in the muscles themselves. It seems possible that whatever the site of the lesion diabetic vascular changes may play a significant role as in the peripheral and cranial mononeuropathies.

According to Garland the electromyogram always shows an abnormality in diabetic amyotrophy. The motor unit pattern on contraction of the affected muscles show varying degrees of fall-out with the preservation of the appearance of the surviving motor units. In two of his cases synchronization of motor unit activity was demonstrated, a pattern usually denoting cord disease. This, together with the extensor plantar responses occasionally seen, may have been responsible for his earlier view that the lesion was in the cord and his use of the term “diabetic myelopathy”.

The spinal fluid protein was elevated in both of our cases and this is reported in about 50 per cent of the diabetic amyotrophies. It is, of course, elevated in a higher percentage of the neuropathies of diabetes (approximately 80 per cent) and according to Mencer Martin the level corresponds to the severity of the neuropathy. Garland states that an elevated protein level in the spinal fluid of the diabetic amyotrophies has no prognostic significance.

An amyotrophic picture is also known to occur with other disorders of carbohydrate metabolism, for example in hypoglycemia due to adenoma of the pancreas. Treatment here consists of removal of the tumor. Other diseases giving rise to a similar picture include occult carcinoma (especially lung, breast and ovary), thyroid disease, amyloid disease, multiple myeloma and steroid therapy.

Diabetic amyotrophy is usually a self limited disease and some patients give a history of remissions and exacerbations prior to diagnosis. Indeed, one of Bruns original cases described this phenomenon although he was treated by dietary restriction alone. According to Garland the motor disability and strength are the first to return. The tendon reflexes may or may not return and the extensor plantar responses always become flexor. The electromyogram usually remains abnormal but shows improvement.

Treatment consists of rigorous control of the diabetic blood sugar levels and not just abolition of the glycosuria. Rest usually relieves the pain but this should not be overdone and active, not passive, physiotherapy instituted early.
Two cases of diabetic amyotrophy are presented. The similarities and differences between this syndrome and the neuropathies of diabetes are discussed. The motor neuropathy of diabetes and diabetic amyotrophy may occur in mild or unsuspected cases while the sensory neuropathies usually reflect long standing disease. Other conditions giving rise to a similar picture are noted.

REFERENCES