SUMMARIO IN INTERLINGUA

Convulsiones Durante Cetosis Diabetic: Reporto de Un Caso

Un femina de quaranta-sex annos de etate con un breve historia de symptomas diabetic disveloppava sever cetosis e deveniva comatose. Ante e durante le tractamento, illa habeva un serie de major accessos epileptic, occurrente in le curso de un periodo de dece-duo horas sed non associabile con ulle causa apparente. In le absentia de altere morbo, il pare que le accessos esseva debite a latente epilepsia idiopathic e que le disturbation metabolic de cetosis diabetic ageva in un maniera o un altere como mechanismo precipitatori.

REFERENCES

[1] Gyorgy, P.: Personal communication, 1955.

[2] Coursin, D. B.: Convulsive seizures in infants with pyridoxine deficient diets. J.A.M.A. *154*:406-08, 1954.

[3] Kruse, H. D., Orent, E. R., and McCollum, E. V.: Studies on magnesium deficiency in animals: chemical changes in blood following magnesium deprivation. J. Biol. Chem. *100*:603-43, 1933.

[4] Gans, B.: Proc. Roy. Soc. Med. *47*:6-7, 1954.

[5] Izzo, J. L., Schuster, D. B., and Engel, G. L.: The electroencephalogram of patients with diabetes. Diabetes *2*:93-99, 1953.

[6] Kety, S. S., Pollis, B. D., Nadler, C. S., and Schmidt, C. F.: Nitrous oxide method for the quantitative determination of cerebral blood flow in man: theory, procedure and normal values. J. Clin. Invest. *27*:476-83, 1948.

[7] Lawrence, R. D., Meyer, A., and Nevin, S.: The pathological changes in the brain in fatal hypoglycemia. Q. J. Med. (N.S.) *11*:181-201, 1942.

EDITORIALS

DIABETIC NEUROPATHY

Although the neurologic manifestations have been recognized in diabetes since 1864, they have not received the detailed analysis usually given to other neurologic conditions. Because of their frequency they are entitled to more attention. Among the twenty million estimated diabetics throughout the world undoubtedly there are probably hundreds of thousands of cases of "diabetic neuropathy." The latter term is generally applied rather loosely, referring to involvement of peripheral nerves only (in the previous sense of "neuritis") in some instances and to more widespread involvement (both motor and sensory) emanating from spinal cord lesions in others. Garland urges that the designation neuropathy be restricted to disorders of the peripheral nerves leaving "myelopathy" for cases in which the cord alone is involved.

In 1953, Garland and Taverner reported three diabetic patients suffering from a purely motor disorder which appeared to them to be a syndrome probably of cord origin. The patients were over fifty years of age and gave a recent history of diabetes of mild degree. The neurologic picture was limited to the legs and included diffuse pain, weakness, atrophy and areflexia, and usually it was asymmetrical. Other features which appeared less constantly were extensor plantar responses, an elevated protein content of the cerebrospinal fluid, and electromyographic changes in the affected muscles. In none was there any sensory loss. The condition was found only in patients who, though usually being treated, had not been strictly regulated. At the same time manifestations were largely or totally reversible.

Subsequently, Garland reported twelve cases suffering from this neurologic entity (*British Medical Journal*, Nov. 26, 1955). The findings were not always limited to the lower extremities, the upper extremities being affected occasionally. Because the manifestations were not always uniform the author selected the term "diabetic amyotrophy" in preference to myelopathy to describe this condition. Although most of the author's cases had been overlooked, probably because it does not coincide with other more readily recognizable neuropathies, the syndrome is by no means rare. Other diagnoses which were tentatively considered by referring physicians in these cases were sciatica, lumbar arthritis or motor neurone disease. The condition is always reversible with the achievement of total diabetic control. Recovery is somewhat hastened by use of exercise, calipers and crutches.

Garland stresses the use of electromyography as a diagnostic aid. Affected muscles showed electromyographic changes in all cases which are subject to wide variations suggesting a cord lesion in some cases but not

so in others. Regardless of the nature of the electromyographic changes the distribution of the weakness, atrophy and areflexia followed a similar pattern. The authors point out that in diabetic neuropathy the sensory fibers of the peripheral nerve are generally affected, but he also envisions the possibility that diabetes can produce purely motor changes in the peripheral nerves.

In this regard, I should like to call attention to a strikingly similar syndrome which I described in 1954 entitled "Femoral Neuropathy in Relation to Diabetes Mellitus: Report of 17 Cases" (DIABETES 3:266-73). These cases were likewise characterized by pain and tenderness along the cutaneous branches of the femoral nerve, weakness and atrophy of the quadriceps muscles and patellar areflexia. Although the cerebrospinal fluid was not examined routinely in these patients a significant elevation of the protein content was found in three cases, a finding which was considered significant. A positive femoral nerve stretch test proved to be a valuable diagnostic aid in many of these patients. Although so-called diabetic tabes had been suggested as a diagnosis in some of these cases and herniated intervertebral disc in others, in our opinion this syndrome represents a peripheral neuropathy or neuronopathy. Aside from the clinical involvement of the femoral nerve, other features which seemed to favor a peripheral neuropathy rather than a spinal cord lesion are the reversibility to normal of the symptoms and signs and the selectivity of various fibers (e.g., motor fibers) within the nerve. Aside from an anterior horn involvement (which is unlikely to be completely reversible) it is difficult to visualize within the spinal cord a lesion of such selectivity as to involve modalities such as the patellar reflex and pain without affecting at the same time other sensory modalities. Furthermore, it is difficult to visualize in those patients who complain of pain and paresthesia in the lower extremities just how these symptoms could be explained by an anterior horn lesion or a purely cord lesion for that matter. One must envision, over and above any cord involvement, that the sensory symptoms probably take their origin in peripheral nerves or sensory roots.

The cases which serve as the basis of Garland's very excellent, thought-provoking report appear to show a striking similarity to those which we encountered with femoral neuropathy. Certainly, the most striking features of his cases—weakness and atrophy of thigh muscles, areflexia and the close association with unregulated diabetes—were present also in ours. The presence of pain and the elevation of the cerebrospinal fluid protein in some cases also occurred in both series. Finally, the reversibility of the manifestations as a result of meticulous diabetic control is another point of total agreement in the two groups. Although Garland has called attention to an interesting, possibly a new syndrome, the question as to whether it is truly a myelopathy in some instances, as this author suggests, or an extensive neuropathy (neuronopathy), extending up into the nerve roots in some cases, still remains to be settled.

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GALACTOSEMIA

Galactosemia is a congenital disorder characterized by an inability to metabolize galactose normally. The earliest cases were reported by von Reuss[1] in 1909 and Goppert[2] in 1917. Mason and Turner[3] described the first case in the American literature in 1935. Since that time several patients have been reported and there is reason to believe that many more well documented cases have not been reported.

Infants with galactosemia usually are not ill during the first several days of life. Then signs of growth failure appear even though the appetite remains good. Enlargement of the liver, often accompanied by jaundice, gradually appears. The jaundice may seem to be prolonged "physiologic" jaundice of the newborn and frequently is transient, subsiding after a variable period of time. Splenomegaly, ascites, low plasma protein, and abnormal liver function tests may be found. Lamellar cataracts develop with a high degree of frequency if the disorder is not recognized and treated early. Mental retardation may be present, sometimes without relation to severity of disease or to onset of treatment. The galactose tolerance curve is prolonged and elevated in all cases. Normocytic anemia and osteoporosis have been reported in some cases. Amino-aciduria, apparently due to diminished tubular reabsorption, has been described.[4]

Reducing substance in the urine is invariably present (although not necessarily in every specimen) and is an important clue in early recognition of this disease. Negative yeast fermentation and the formation of specific osazone crystals with phenylhydrazine will identify the reducing substance as galactose. Recently devised enzyme impregnated test papers,* specific for the detection of glucose, may be used for simple screening to eliminate this sugar as a source of the melituria.

Early diagnosis and treatment are of prime importance to the prognosis for the patient. Complete removal of

* Clinistix, Ames Company, Inc., Elkhart, Indiana. Tes-Tape, Eli Lilly and Company, Indianapolis, Indiana.