well as progressive muscular atrophy and degenerative lesions of the cervical spine, had to be considered.

Whereas the acute form of polymyositis presents with constitutional symptoms, muscle pain, and tenderness, these are all strikingly minimal or absent in the subacute and chronic forms of the disease (Eaton, 1954). Others have observed that, though striated muscle anywhere in the body could be involved, characteristically the proximal limb muscles and trunk muscles are chiefly affected (Biernard, 1948; Walton and Adams, 1958). It is this distribution of muscular wasting which has led to confusion with muscular dystrophy, so that muscle biopsy is essential for the diagnosis of most cases of subacute and chronic polymyositis. As the disease involves necrosis of muscle, creatinuria has been found to be a constant feature of it (O'Leary and Waisman, 1940; Jager and Grossman, 1944; Cumings, 1953), and persists as long as the disease remains active.

With regard to the prognosis of polymyositis and dermatomyositis, it is now well recognized that up to 50% of the florid cases terminate fatally, usually from pharyngeal or respiratory paralysis or from associated bronchopneumonia. In many of the others the disease becomes arrested, with varying amounts of residual disability (Oppenheim, 1903; Brock, 1934; O'Leary and Waisman, 1940; Sheard, 1951).

In conclusion, it is felt that rare and fatal conditions which have previously been reported from European communities in the temperate zones can also occur in tropical countries among the indigenous population, and that more and more of these conditions will doubtless be discovered as medical services, with adequate laboratory facilities to establish accurate diagnosis, become more readily available.

Summary

Three cases of polymyositis occurring in West Africans are described and the pleomorphic character of the clinical picture is discussed.

The literature is reviewed. Constitutional symptoms, it is generally agreed, are absent in the subacute and chronic forms of polymyositis and the presence of skin lesions is variable. Spontaneous remissions often occur.

The existence of the disease in West Africa is shown.

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A CASE OF PROGRESSIVE HEMIATROPHY PRESENTING WITH SPONTANEOUS FRACTURES OF THE LOWER JAW

BY

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Hemiatrophy of the body is a rare recessive hereditary degeneration, which can affect the skin, connective tissue, muscle, bone, and peripheral nerves, and involve many organs and systems. It presents to specialists of widely separated interests, so that none may have a true picture of the disorder. To the neurologist it is progressive facial hemiatrophy, its usual textbook designation, and a rare cause of trigeminal neuralgia, epilepsy, and hemiplegia. To the dermatologist it is morphoea, scleroderma en coup de sabre, and various minor paracentral abnormalities of pigmentation of the hair and skin. To the dental surgeon it is a cause of unilateral malocclusion and dental deformity. To otolaryngology it contributes unilateral deafness and malformation of the sinuses. For gastro-enterologists it may induce dysphagia from unilateral atrophy of the oesophagus. It has also caused hypertension from unilateral kidney disease and unilateral diaphragmatic paralysis. To those interested in medicine as a whole its widespread modes of presentation will be of interest. To these is added another, of interest to surgeons, particularly those specializing in facio-maxillary work. Spontaneous fractures of the lower jaw are usually caused by weakening of the bone from infection or neoplasia. The following case is believed to be the first recorded one where the cause of the fractures was progressive facial hemiatrophy.

Case History

The patient is a widow aged 53, with one son. There has been no left-handedness, tic douloureux, or anything else of note in the family history. There were 11 siblings, three of whom have died of unknown causes.

At the age of 13 she had skin changes which began below the right breast and which slowly spread. Since this time the right arm and breast have always been smaller than the left.

At 33 exophthalmic goitre and pre-eclamptic toxæmia occurred, from which she recovered with termination of the pregnancy. She has had no injury, miscarriages, or blood transfusions. At 43 about four attacks of unconsciousness occurred without warning, in which she fell to the ground. She has bruised herself, but not been incontinent or bitten her tongue in these attacks. There was no observer. About this time Raynaud's phenomenon began, and persisted for some years, but she does not get this now.

At the age of 49, frontal and vertical headaches began, yielding to aspirin and usually associated with nausea and vomiting. These occurred several times a week, usually on waking. They have been less severe and without nausea for about a year. A few months after the headaches began, momentary jabs of severe pain in the right infra-auricular region developed. These radiated up into the temporal scalp

and would occur 50 to 60 times a day, every day. They lasted two months. About the same time similar pain lasting two to three seconds began in the right cheek and occurred 6 to 150 times a day. Talking and drinking brought on the pain, but there were no trigger points, associated salivation, lacrimation, or swelling of the face, though it occasionally flushed. Shortly after this she had a similar pain in the right lower jaw and would get spasms in which the jaw clenched on the right spontaneously, sometimes with the pain and sometimes not. At other times she would clench the jaw voluntarily with the pain, which was severe and not relieved by aspirin.

On October 12, 1953, after one bad spasm, the pain persisted and her face swelled. A spontaneous fracture of the neck of the right condyle of the mandible was found; it was treated by intermaxillary fixation and healed well. Both jaws were edentulous. She cannot remember whether a spontaneous contracture without pain initiated this fracture. No facial asymmetry was then observed. About this time numbness of the right lower lip and chin began, and has persisted. Koilonychia of the third and fourth fingers of the right hand developed an unknown time ago.

On June 12, 1957, while resting in a deck-chair, she had a severe muscular spasm and the right angle of her mandible was found to be fractured. A biopsy of bone taken from the fracture site was normal. The fracture was treated as before and united satisfactorily. On both occasions that the mandible fractured she was wearing full upper and lower dentures.

She was quiet and a good witness; she looked older than her age and had white hair. The right eye was indrawn and the right side of the face was smaller than the left in the upper part (Fig. 1). The muscles of the right arm, shoulder-girdle, and hand were all wasted (Fig. 2), but the muscular power was fair. All the tendon and cutaneous reflexes were present and equal and the plantar responses flexor. There was no evidence of spasticity on either side. There was an inconstant fasciculation of the muscles over the right side of the chin near the midline.

There were patchy sclerodermatous changes (morphoea) in the upper half of the body, all over the right shoulder, the arm, and forearm, where the skin was particularly thick and hard. A patch extended around the left side of the body, at the level of T 9. On the right temple, just in front of and above the ear, there was an oval patch without hair, slightly violaceous in the centre, in which the skin was thickened and atrophied. The legs were unaffected and equal in size. The right breast was smaller than the left. The remainder of the physical examination, including the fundi, showed nothing abnormal. Her blood-pressure varied between 100/80 and 175/95. The urine contained no

albumin or sugar. X-ray examination showed: skull and mandibles, no abnormality; humeri, no lesion, equal in size; hands, several phalanges showed an abnormal increase in density on the right side; cervical spine, generalized spondylosis C 4 to 6, abundant calcifications seen in the thyroid. I.V.P. was normal. Barium swallow showed no abnormality of oesophagus. Haemoglobin, 91%; W.B.C. 7,000; E.S.R. 7 mm. per hour (Westergren); normal differential and film appearances. Plasma cholesterol 297 mg./100 ml. W.R. and Kahn, negative. Serum alkaline phosphatase 5.5 and 2.9 K.-A. units per ml. Serum calcium, 8.4 mg./100 ml. Serum inorganic phosphate, 3.4 mg./100 ml. E.E.G. (Dr. N. S. Alcock): "The resting record is normal. In one of her spasms only muscle activity was seen. There is a flat channel over the scarred area on the right side. There is no evidence of neurogenic activity as a basis for her spasms."

Myography (Squadron-Leader J. L. Milligan).—"I performed intensity duration curves on three muscles on each side—abductor digiti minimi, triceps, and tibialis anterior. The curves for the right side were all normal, but the curves for the left side showed a more sluggish response and a kinked curve most noticeable in the abductor digiti minimi. This could indicate a partial denervation. Electromyography was performed on all the above muscles. In only one muscle, left triceps, was there any evidence of spontaneous activity, and this I do not think was truly spontaneous, as it could be eliminated by contracting the antagonist. There was no evidence of fibrillation to support the intensity duration curve. On volition, the left side showed normal recruitment of motor units, the amplitude and duration of which were normal. The interference pattern was complete. On the right side, on volition, the motor units were much larger in amplitude, some being as large as 8 millivolts and 10–15 milliseconds in duration. A full interference pattern was produced on maximal volition, but of a much higher amplitude than on the left side. There was no evidence of an increased number of polyphasic potentials as compared with normal. The picture over all, then, is normal on the left except for some small deviation in intensity duration curves, which could stand being repeated at a later date. On the right side, the large motor unit responses may mean that there is a lesion in the cord affecting the small motor unit cells as in poliomyelitis, but one would then expect to find an abnormal intensity duration curve and the presence of fibrillation."

Local anaesthesia of the mental nerve relieved the spasms of pain, and peripheral neurectomy was performed. This gave relief for only a week. Infraorbital nerve blocks gave no relief.

She was therefore referred to Mr. G. Alexander for consideration of section of the Gasserian sensory roots.

Temporary block of this ganglion relieved her pain, and on March 20, 1958, Mr. Alexander avulsed slightly more than the outer half of the root from the posterior limb of the ganglion. The motor root was preserved. This resulted in analgesia of the face to the level of the lower eyelid and retention of corneal reflex. The patient's constant pain was abolished and also the trigeminal neuralgia.

Subsequently she remained well, but still had painless attacks in which the muscles of the right side of the face bunched up. These have not been observed.

In June, 1958, pain developed in the back of the neck. Grating of the cervical spine was noted, and x-ray films showed spondylosis. Traction of the neck in hospital has not relieved this pain. In December, 1958, she reported intermittent sweats all over the body and a slight increase of the weakness of the right side, but there

FIG. 1

FIG. 2

was no objective change. She remained pleased to have had the partial Gasserian neurectomy.

Discussion

Hemiatrophy of the body is usually facial, and not always progressive and unilateral. Like other hereditary degenerations, most cases begin near adolescence. Wartenberg (1945) reviewed the subject thoroughly. The present case has the typical features of onset at puberty, scleroderma, fits, trigeminal neuralgia, muscular spasms, sweats, and slow progression.

There is no evidence of involvement of the internal organs in the hemiatrophy. The E.E.G. showed no evidence of epilepsy. The findings on myography are not clear. Possibly there may be a cord lesion on the right, and there is no evidence of an upper motor neurone lesion affecting the muscles of the left side. Cases in which this occurs have been described by Brain (1955), Reider and Player (1946), and Wartenberg (1945), who quoted several other instances. The operative results suggest that the cause of the trigeminal neuralgia was peripheral.

The segment of scleroderma on the opposite side of the body suggests that in this case the factor controlling the process is not confined to the right. It is assumed that the scleroderma and the hemiatrophy have a common origin on the evidence collected by Wartenberg (1945). Urechia and Retezeanu (1936) have reported a case of right hemi-facial atrophy with atrophy of the opposite arm, and Thoma (1954) mentions others in which the hemiatrophy was patchy and bilateral.

The spontaneous fractures of the jaw, with which this case presented, are believed not to have been described before in a case of hemiatrophy of the body. The cause was probably intense spasm of the right side of the face due to the trigeminal neuralgia, often found in this condition, acting on a jaw weakened by the hemiatrophy.

Summary

A case of progressive hemiatrophy is described, in which the patient twice suffered a spontaneous fracture of the edentulous mandible on the affected side, attributed to facial spasm associated with trigeminal neuralgia.

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Since the discovery of the curative powers of the sulphones, interest in leprosy has increased, campaigns have been launched bringing as many as possible of the estimated 10 to 12 million people affected throughout the world under treatment, and more doctors are engaged in research into the unsolved problems of the disease. W.H.O. has played a leading part in spreading knowledge of advances in treatment and in helping organize leprosy campaigns in countries where the disease is an important public-health problem, and its work is described in the booklet *International Work in Leprosy, 1948-59*, and is set against the background of a short history of leprosy, its incidence, its treatment, and the research and other problems it raises. (Available from H.M.S.O., P.O. Box 569, London, S.E.1.)

THE INHERITANCE OF ESSENTIAL PENTOSURIA

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Essential pentosuria is a rare condition characterized by the constant excretion of xyloketose in the urine. Included by Garrod (1923) among the inborn errors of metabolism, the condition probably results from a block in the metabolism of xyloketose, one of the normal products of glucuronic acid metabolism (Enklewitz and Lasker, 1935; Flynn, 1955).

This report describes the finding of xyloketosuria in a mother and her two children.

Methods.—The following methods were used to investigate the reducing substance in the urine: (1) qualitative Benedict's test for urine reducing substance; (2) Bial's test for pentoses; (3) the Lasker-Enklewitz test for xyloketose (Lasker and Enklewitz, 1933); (4) osazone preparation; (5) paper chromatography (Horrocks and Manning 1949) using 3:2:1.5 *n*-butanol-pyridine-water solvent (Hough *et al.*, 1950), demonstrating the sugar spots with aniline hydrogen oxalate; (6) quantitative estimation of the urine xyloketose (Lasker and Enklewitz, 1933), using the factor of Greenwald (1930) for the calculation.

Cases

The propositus, a Jewish woman aged 40 (E.R.), was admitted to the National Hospital, Queen Square, complaining of unsteadiness of gait, giddiness, and nausea for two years. The symptoms dated from a hysterectomy for fibroids two years previously, and no organic cause for them was found. During investigation this patient was found to have a reducing substance in the urine; it transpired that this had been first noticed in 1939 and on a number of occasions since, resulting in three normal glucose-tolerance curves and the warning that she was "knocking on the door of diabetes."

On the present admission the urine showed a constant yellowish-green precipitate with Benedict's solution; a glucose-tolerance curve was normal (fasting blood-sugar 98 mg./100 ml.; half-hour, 155 mg.; 1 hour, 138 mg.; 1½ hours, 115 mg.; 2 hours, 112 mg.; 3 hours, 80 mg.), with no alteration of the amount of reducing substance in the urine during the test. The urine gave a positive Bial test for pentose and a positive Lasker-Enklewitz test for xyloketose. An osazone was prepared, showing fine needles similar to glucosazone, but smaller. Glucose-oxidase test paper ("clinitix"), specific for glucose, was negative. Paper chromatography with suitable markers showed a pentose

FIG. 1.—Results of paper chromatography.