were re-read and condensed and physical examination was repeated. The story outlined here was typical of the syndrome described by Moersch and Woltman, and physical examination was as unrevealing as it had been in their cases.

All there was to find was a hypertrophy of the muscles of the neck and trunk with board-like rigidity of these muscles, the abdominal muscles, and the muscles of the lower limbs. The legs were contorted into grossly misshapen deformities, one ankle had varus of such degree that the sole of the foot was facing upwards.

The urine occasionally revealed a trace of glucose which 4 out of Moersch and Woltman's 14 cases had shown.

To be able to identify and name the condition gave the patient no benefit whatever. No doubt, had she known Matthew Arnold's lines she might have expressed the same wish as he did:

Nor bring to watch me cease to live
Some doctor, full of phrase and fame,
To shake his sapient head and give
The ill he cannot cure—a name.

With Dr. Abdullah's kind permission the patient was brought over to one of this hospital's weekly clinical meetings at the beginning of January, 1957. When I exposed her body and gently moved one limb to show the muscular rigidity this provoked such dramatic and widespread spasm that she was clearly in great agony, crying out loudly, gripping bystanders till her nails dug into them, and sweating visibly and profusely. It appeared that the one-mile ambulance journey had brought her to a climax of acute spasm, and the nurses who accompanied her begged that she might be given morphine for the journey back. We at the meeting felt guilty that a clinical demonstration had unintentionally produced so much suffering. She returned to Leamington Park under heavy sedation and recovered in a few days. In April a course of "tolseram," 1 g. t.d.s. gradually increasing to 3 g. t.d.s., was tried for one month. It made no difference. Thereafter she remained in her usual rigid bedfast condition for another six months, till her unexpected death on June 18. The house-physician's note makes a suitable epitaph: "Commenced 'hysterical turn' at 10.30 p.m. on June 17, 1957, and settled down at 2 a.m. Found dead at 3 a.m." Her hysteria had at least been awarded the posthumous dignity of inverted commas.

Post-mortem Findings

A complete microscopical examination showed no changes except those attributable to age and atherosclerosis. The only significant changes were in the muscles. These changes, which showed best in the sacrospinalis muscle and hardly at all in the pectoral muscles, were: (a) Abnormal variations in the size and staining reaction of the muscle fibres; the swollen fibres were pale and lacking in striation. (b) Increase of the connective tissues both collagen and elastic fibres, principally the former. (c) Localized areas of atrophy and fibrosis of muscle accompanied by nuclear aggregates and lymphorrhages.

Dr. C. Treip reported that these muscular changes appeared to be primary and not related to any change in the central nervous system. This supports the idea that this illness may be a type of myopathy. Nevertheless it is possible the changes are secondary to spasm or to prolonged bed rest, and the true pathology still remains uncertain.

Comment

The way I have described this case is a little unorthodox, but I thought it would make it easier to read. Reports of single cases of rare diseases are not usually exciting; this was an exciting and rather fearful case, so I have tried deliberately to describe it in a way that might convey this.

Summary

A woman of 50 suffered from progressive, fluctuating muscular rigidity and spasms for over 15 years. The spasms caused a fractured leg and broke the steel fittings

used to repair it. For many years the spasms were labelled hysterical. Six months before death a diagnosis of stiff-man syndrome was made because of the clear resemblance to Moersch and Woltman's cases. No treatment did any good. She died after a severe spasm, and post-mortem examination showed muscular changes, which suggests this illness is some form of myopathy.

I am indebted to Dr. A. Abdullah for his courtesy in allowing me to follow up this patient in his wards after she left my care; to my registrar, Dr. L. E. Lennard-Jones, for his general assistance; and to Dr. C. Treip for the post-mortem report and the extensive microscopy studies.

Medical Memoranda

Two Cases of Bullous Chicken-pox

Though bulla formation is mentioned as an occasional complication of chicken-pox, it is rarely seen. These cases are interesting in that one aroused a transient fear of phenol poisoning and that both made a complete recovery without residual scarring, in spite of the alarming picture which each presented when the disease was at its height.

On August 13, 1956, in a small housing estate in Surrey, where chicken-pox was epidemic, a girl aged 8 years, successfully vaccinated in infancy and with no history of chicken-pox or herpes, was seen at the commencement of what appeared to be a mild attack of chicken-pox. There were about 20 pocks on the trunk and a dozen on the face, scalp, and neck; none on any mucous membranes. Treatment was started with aspirin and local application of 1% phenol in calamine lotion.

When the patient was next seen, three days later (fourth day of illness), there were several large discrete bullae, each overlying the site of a pock, on brow, nuchal region, left axilla, left elbow, upper abdomen, and vulva. These were filled with a turbid fluid, and varied in area from 7 by 5 cm. to 3 by 2 cm. Each was surrounded by a zone of erythema 1 to 2 cm. wide, and the appearance was of a number of localized burns, as from a hot-water bottle. Scattered over the intervening skin were normal pocks in an unexpectedly advanced state of involution.

Though it seemed unlikely, when such a small proportion of treated pocks had proceeded to bulla formation, the possibility was considered that the patient was abnormally sensitive to the low percentage of phenol in the lotion applied or that the percentage had been accidentally increased in the course of dispensing. Local dressing with 0.5% cetrimide cream was substituted and tetracycline administered orally in the hope of reducing septic complications.

The suspicion of phenolic poisoning was renewed on the following day (fifth day of illness), when there was oedema of the eyelids and face sufficient to close the left eye; but there was no oliguria, and the urine was free from protein and normal on microscopy, so the oedema was assumed to have arisen by drainage from tissues surrounding the large bulla in the left frontal region. There was similar oedema of the vulva. By now nearly all the bullae had ruptured and a central pock could be seen in each of the raw areas revealed.

On the seventh day of illness improvement was apparent. Oedema was lessened and the denuded areas were healing without obvious secondary infection. By the end of the second week the patient was ambulant, and at four weeks there remained only two small cicatrices such as may frequently be found after an uncomplicated attack.

On August 26, 13 days after the onset in the first patient, her brother, aged 17 months, developed bullae on both upper eyelids, scalp, thighs, and one finger, with a heavier crop of normal pocks on the trunk than his sister had had. There had been no contact with the first patient from the

onset of her illness, and precautions had been taken against direct transfer of infection.

Owing to scratching, secondary infection of the bullae in the second case was considerable and cardboard arm splints were required. Local treatment with cetrimide cream and systemic tetracycline was maintained, and by the 28th day of his illness he was completely clear. Both he and his sister were afebrile during the period of observation, showed no signs of toxæmia, and had good appetites throughout.

On September 5, 23 days after the onset of the first case and 10 days after the onset of the second case, the mother of these children, aged 28 years, believed not to have had chicken-pox, developed three groups of small soft bullae on the flexor surfaces of the left arm, forearm, and wrist, and one on the right forearm. These bullae were about 1 cm. in diameter except that on the wrist, which was 2 by 3 cm. There were no other bullae or pocks and no systemic disturbance. The lesions responded rapidly to soaks of 1 in 2,000 potassium permanganate, and by September 24, 19 days after their appearance, they had healed completely.

The mother believed that these lesions had arisen from contact with those of the second case, as they occurred on those parts most closely in touch with him when she held him in her arms for the frequent nursing attention required, and the appearance was certainly more that of a direct inoculation than a systemic infection.

Throughout the present epidemic no similar case has been reported from this small housing estate or from a larger one near by.

The second of the cases described was cared for by my colleague, Dr. Phyllis Phipps, during my absence on holiday.

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A Case of Paget's Disease of the Patella

Paget's disease is by no means unknown in the patella, but in almost all cases in which it has been reported in this situation the disease has been widespread throughout the skeleton. This was so in the cases reported by Packard *et al.* (1901), Sugarbaker (1940), and Gutman and Kasabach (1936). The largest series referred to was that of Dickson *et al.* (1945), who found involvement of the patella in only 4 cases out of 367. The present case is of interest in that the disease process was confined to one knee-cap.

CASE HISTORY

The patient, a man aged 46, complained of aching pain in front of the left knee for three and a half months. There was no history of injury, and the pain, of gradual onset and at first intermittent, had become progressively more troublesome.

On examination, the left knee was found to be warmer than the right. There was palpable enlargement of the left patella, but the knee was otherwise strikingly normal. X-ray examination (see illustration) showed a hyperplastic reaction in the patella, and a tentative diagnosis of Paget's disease was made. There were no symptoms related to the other limbs, trunk, or skull, and x-ray films showed no lesion in any other part of the skeleton.

At operation on May 3, 1955, the left knee-joint was opened by a medial parapatellar incision. The naked-eye appearance of the interior of the joint, including the articular surface of the patella, was normal. The patella was excised and the extensor mechanism repaired with mattress sutures of No. 30 linen thread, overlapping the ligamentum patellæ over the quadriceps tendon. The limb was treated in plaster in extension for six weeks, the patient walking in the P.O.P. from two weeks after operation. Exercises were started out of plaster on June 22, and by the end of August the patient had a fully mobile, strong painless knee.

The excised patella was submitted to Dr. J. E. Morison, of the Belfast Central Laboratory, who reports: "There is very active osteoclastic absorption with numerous multi-

nucleate osteoclasts. In other areas there is very active deposition of bone, and osteoblasts are prominent on the bone trabeculae. Ackerman states that Pagetoid mosaic bone can occur in polyostotic fibrous dysplasia. I suppose it could also be expected to occur in monostotic fibrous dysplasia, but I think there can be little or no doubt that the appearances are those of Paget's disease. In no other condition is there such widespread absorption and deposition of bone within localized areas."

The sections have also been examined by Professor R. A. Willis, who writes: "Although the mosaic pattern is not

Lateral view of the affected knee before operation.

very prominent, the lesion is probably Paget's disease. I do not know of any record of this being confined to the patella."

Comment.—I have not been able to find a reference in the English or American literature to a case of Paget's disease confined to the patella. The present case is therefore distinctly unusual even if we concede that other parts of the skeleton in this patient may become affected in the future. Little is known about whether, or how, Paget's disease spreads from one area of the skeleton to another. Is it a disease of multicentric origin? Or does it spread directly from a single original focus? Has the removal of the only evident focus in the present case entirely eradicated the disease in this patient? Prolonged follow-up may answer the last question and provide a single shred of evidence towards answering the others.

In the meantime this case provides a most unusual example of the distribution of Paget's disease with a gratifying local and functional result following surgical treatment.

[Since this paper was submitted for publication the patient has died from an unrelated disease (coronary thrombosis). There was no opportunity for a post-mortem examination or the proposed long-term follow-up.]

I wish to thank Mr. R. J. W. Withers for permission to publish this case, and Dr. J. E. Morison and Professor R. A. Willis for their opinion on the sections.

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