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Unusual Presentations of Leprosy

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A diagnosis of leprosy is usually not long delayed if the patient presents with classic skin lesions, but difficulties in diagnosis may arise if the patient presents with symptoms and signs referable to other systems, particularly if skin lesions are absent, or, as is often the case in lepromatous leprosy, early skin lesions have not been observed by the patient and are therefore not brought to the doctor's attention. Let us now consider some cases of leprosy which have come under my care in London and in which the correct diagnosis was delayed because the symptoms, not being dermal, did not suggest the possibility of leprosy to the doctors who saw them in the first place. After this we shall consider some skin manifestations of leprosy which may puzzle a dermatologist.

PATIENT REFERRED TO NEUROLOGIST

Symptoms such as pain, paresthesiae, numbness, muscle weakness and wasting, although common during the course of leprosy, may be confusing if they are presenting symptoms and may lead to reference of the patient to a neurologist in the first place. An Englishman working in India was investigated in a hospital in Madras and was sent to England with a provisional diagnosis of syringomyelia because of intrinsic muscle wasting in the hands; when he was seen in London there were no skin lesions, but a number of peripheral nerves were thickened, and a nerve biopsy (great auricular nerve) confirmed the diagnosis of polyneuritic leprosy. An English surgeon working in Nigeria was sent to a neurologist in London because of a dropped foot, but the doctor's letter sent with him from Nigeria made no mention of the important fact that the lateral popliteal nerve on that side was twice as thick as the nerve of the unaffected leg. Many investigations were carried out, including lumbar puncture and encephalogram, until the simple test of palpating the lateral popliteal nerve revealed the diagnosis of mononeuritic leprosy. A Cypriot complained of a patch of sensory loss on the anterolateral aspect of one thigh and was considered to be a case of meralgia paresthetica (Bernhardt's syndrome) until, at a later examination, a thickened femoral cutaneous nerve was discovered. An English lady from Kenya was dismissed by a neurologist as a case of hysteria because of patchy sensory loss over feet and lower legs; years later the development of skin lesions led to a correct diagnosis.

PATIENT REFERRED TO SURGEON

An Anglo-Indian (Eurasian) male presented with a cystic swelling in the left popliteal space, and it was in the operating theater that the connection with a thickened lateral popliteal nerve was noted; the caseous material in the cyst was typical of a leprous nerve abscess. A student from Thailand was seen by a surgeon because of a cystic swelling on one side of his neck, but the attachment of the swelling to a thickened great auricular nerve was not observed until the patient was in the operating theater. A West Indian boy was seen in his home country with a sinus in his left upper arm discharging caseous material, and was treated conservatively. Years later, as an immigrant in London, he was sent to me with wasting of the intrinsic muscles of his left hand; the left ulnar nerve was extremely hard and was irregularly thickened, and radiologic examination showed areas of calcification in the nerve. There were no skin lesions, no other nerves were thickened, and the Mitsuda reaction was strongly positive (+++). There is no doubt that his original symptoms were due to a discharging abscess of the ulnar nerve.

A badly blistered leg in an Englishman on leave from Nigeria was treated at a surgical center in London without anyone

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being curious as to why a hot water bottle could cause such a severe burn and as to why the injury was painless. He returned to duty in Nigeria, and 18 months later, shortly before coming home on leave, he developed skin lesions. When I examined him in London he had mid-borderline (BB) leprosy with moderate numbers of acid-fast bacilli in skin smears, and there were a number of thickened nerves which I have no doubt could have been palpated 18 months earlier when he had his painless burn.

A chronic plantar ulcer was the presenting symptom of an Indian student, and after many investigations in a London teaching hospital a diagnosis of a tuberculous ulcer was made. He was given routine antituberculosis treatment, the ulcer healing after every period of bed rest and recurring whenever walking was resumed. By the time he had completed 12 months of treatment there were many lesions on his skin typical of borderline-lepromatous (BL) leprosy and containing large numbers of acid-fast bacilli. Anesthesia of the foot and thickening of the lateral popliteal nerve had not been noted when he first attended.

PATIENT REFERRED TO PHYSICIAN

I have a number of lepromatous patients who were first referred to a physician because of chronic edema of feet and lower legs. One was an Anglo-Guyanan who was accepted by the Post Office in London and was given the task of delivering letters. Three months later he reported to his doctor with edema of his legs, which he had observed at the end of each day's work. He was referred to a physician, who noted certain peculiarities of his skin and asked for my opinion. The patient himself had noted nothing abnormal about his skin, but there was no doubt regarding the diagnosis of lepromatous leprosy.

A Nigerian student reported with multiple skin lesions typical of lepromatous leprosy, and on giving his history told me that he had been investigated in Kaduna the previous year because of edema of his legs. He had been told that his edema was postural, due to sitting for many hours a day at his studies, but I have no doubt that had routine skin smears been carried out at that time the true cause of his edema would have been discovered.

An English lady who had spent all her life in India came to England and consulted a doctor about edema of her legs. She was referred to a physician, who made a provisional diagnosis of myxedema, as she appeared lethargic, her skin seemed a little thickened, her eyebrows were sparse, there was a hoarse quality in her voice, and she had a moderate normocytic normochromic anemia. It was only when tests of thyroid function were found normal that the possibility of lepromatous leprosy was considered, and she was found to have many faintly erythematous macules on her skin which were teeming with acid-fast bacilli. It should be noted that carpal tunnel syndrome may occur in myxedema, together with sensory impairment in the hand, thus simulating leprosy even more closely.

PATIENT REFERRED TO OPHTHALMOLOGIST

A Greek lady resident in London was referred to an ophthalmologist complaining of recurring redness and discomfort in her eyes associated with photophobia, lacrimation and dimness of vision. There was no doubt about a diagnosis of iritis, and it was considered to be syphilitic on account of tiny deposits in each iris, a deformed nose of "saddle" type, and a positive Wassermann reaction. It was only when anti-syphilis treatment was found to be ineffective that her skin was carefully examined and a correct diagnosis of lepromatous leprosy was made. The treponemal immobilization test (T.P.I.) was found to be negative, thus proving that the positive Wassermann reaction was a false positive reaction. The tiny deposits in the irides were leprous—the typical iris 'pearls'.

PATIENT REFERRED TO NOSE AND THROAT SURGEON

An Englishman who had fought during World War II in North Africa, having never previously left England, developed nasal symptoms five years after demobilization. These symptoms consisted of nasal dis-

charge and bleeding, and difficulty in breathing through his nose. He was referred to a specialist, who found septal ulceration and a positive Wassermann reaction. On being sent to the venereal diseases clinic he astonished the venereologist by asserting vehemently that it was impossible for him to have contracted syphilis, and so forcible were the patient's protests that his serum was more fully examined and the treponemal immobilization test was carried out. When a negative result was returned it was clear that an alternative diagnosis had to be found, and it was only then that a possible diagnosis of lepromatous leprosy was entertained. This was confirmed when his skin was examined.

PATIENT REFERRED TO DENTAL SURGEON

A West Indian complaining of pain in his right cheek was referred to a dentist. His teeth were found to be healthy and he was sent back to the family doctor, who treated him with analgesics. A few weeks later an erythematous plaque appeared on the right cheek, with all the characteristics of a tuberculoid lesion, together with facial weakness on that side, and it was then possible to explain his pain on the basis of involvement of the fifth cranial nerve—leprous trigeminal neuralgia.

Another way in which leprosy may present as a dental problem is if one or both upper central incisor teeth become loose, appear discolored and "dead", or actually fall out. It was Professor Møller-Christensen ([3]) who first drew attention to the common occurrence of atrophy of the maxillary alveolar process, together with loss of the upper incisor teeth, in the skulls he discovered in the burial ground of a medieval leprosy hospital in Denmark.

PATIENT REFERRED TO CHEST PHYSICIAN

Although there are no pulmonary changes directly due to leprosy, it may fall to the chest physician to diagnose lepromatous leprosy. This apparent paradox is explained by the fact that not only has the lepromatous patient minimal resistance to *M. leprae* but he has minimal resistance to *M. tuberculosis* also. Because of the extremely slow evolution in lepromatous leprosy, the tubercle bacillus may make its presence felt before the leprosy bacillus, as the following case history illustrates. An Anglo-Indian male came to England to work in a factory, but before long found himself in a tuberculosis sanatorium as a result of attending a mass radiography unit with other factory employees. After a year's treatment the chest physician noticed that the patient was developing an increasingly leonine facies, together with nasal symptoms and hoarseness of voice, and asked my opinion. I found signs of active lepromatous leprosy which clearly had been advancing in spite of treatment with streptomycin, PAS and INH. I have no doubt that this patient had clinical signs of lepromatous leprosy long before he had his chest X-rayed, for the disease was well advanced at the time I examined him. Another way in which leprosy may be diagnosed by the chest physician is if a patient is sent to him with "positive" sputum and a normal chest X-ray, for leprosy bacilli are plentiful in the mucosa of the mouth and upper respiratory tract in lepromatous leprosy and will contaminate the sputum.

PATIENT REFERRED TO DERMATOLOGIST

A retired English ship's captain was sent to a dermatologist because of recurring blistering of the fingers of the right hand. It was only when diminished sensation was discovered in the right hand that I was asked to see him, and, knowing how commonly the anesthetic hand becomes blistered as a result of contact with moderate heat, I handed him a box of matches and asked him to demonstrate how he lit the gas ring on his stove at home. No further blistering occurred after he acquired a gas-lighter. As is so often the case, this patient up to that time had no knowledge of impaired sensation in his hand.

An Indian patient was sent to a dermatologist on account of ichthyosis of his back, arms and legs, and a skin biopsy was found to be typical of lepromatous leprosy. The

patient denied having had any skin trouble prior to the ichthyotic changes.

I have treated a number of patients who first reported to dermatologists with *erythema nodosum leprosum* (ENL). They had caused much diagnostic confusion as most of them had no obvious signs of leprosy on skin examination, for they were secretly taking dapsone (which they had brought with them when they came to England as immigrants, chiefly from Malta and Cyprus). These patients considered their leprosy to be well under control and had no inkling that this new development had anything to do with leprosy; therefore they made no mention of leprosy when asked about previous illnesses. Two patients had actually been given steroid therapy for a year before I was asked to see them. Both had denied any previous antileprosy treatment, and they continue to deny it even though they are now well and the doctor-patient relationship is well established; so we must accept the fact that ENL was the first observed manifestation of their leprosy. There is a similarity between these two cases and the one described by Birrell (1) in 1952 as the first case of Weber-Christian syndrome recorded in Australia; a year later (2) he announced that the diagnosis had been changed to lepromatous leprosy in reaction.

SUMMARY

Doctors working in the tropics and subtropics usually have little difficulty in making a diagnosis of leprosy when a patient presents with classic skin lesions, but difficulties are likely to arise if the patient makes no mention of skin trouble and presents with symptoms and signs referable to other systems. This paper draws attention to these unusual presentations of leprosy so that the doctor will be on his guard.

The patient may present as a neural case, with one or more of the following: pain, paresthesiae, numbness, muscle weakness or wasting. Rarely there is painless thickening of a nerve, such as the great auricular, or a cold abscess may develop in a nerve (particularly the ulnar) in the form of a painless cystic swelling attached to the nerve. Anesthesia often goes unobserved until the patient notices a painless burn or other injury. A chronic plantar ulcer should always be considered leprous until proved to the contrary.

The patient may present with nasal trouble, such as stuffiness, discharge and epistaxis; the presence of septal ulceration may lead to an incorrect diagnosis of syphilis.

The patient may present as an ophthalmic case, with symptoms such as blurring of vision, or with redness of one or both eyes associated with photophobia, lacrimation and pain.

The patient may present as a dental case, with facial pain due to leprosy affecting the trigeminal nerve, or with loosening or loss of the upper central incisor teeth due to atrophy of the maxillary alveolar process.

The patient may present as a case of myxedema, with thickened skin, hoarse voice, edema of the legs, and anemia, but tests for thyroid gland underactivity are negative.

The patient may present as a pulmonary case, as pulmonary tuberculosis is a common complication of lepromatous leprosy, and it is not unusual for the chest physician to fail to observe the underlying leprosy at first.

The patient may present with erythema nodosum, and diagnosis may be difficult if the patient has been taking dapsone secretly and therefore has minimal signs of leprosy. But the true diagnosis can be suspected if the lesions of erythema nodosum are shortlived, and scattered over face, arms and legs, and if the patient gives a history of vaccination against smallpox, or of severe emotional stress, or if there is an intercurrent infection or a history of treatment with sulfonamides or iodides.

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