

ART. X.—*Case of Scleriosis with Observations.* By JOHN K. BARTON, M.D., F.R.C.S.I.; Surgeon to the Adelaide Hospital; Lecturer in Surgery in the Ledwich School of Medicine, &c.

THE disease, which was originally described by Thirial under the name of "*Sclereme des adults*," has by modern German authorities been called *skleroma* or *sklerodermia*. Virchow recommends *skleriosis*, and this name has been adopted by Dr. Fagge, in an elaborate article upon this and some allied affections in the 13th Vol. of *Guy's Hospital Reports*, and may, I think, be advantageously retained, expressing as it does the prominent and characteristic feature of the disease, viz.:—a peculiar *hardness* of the integument, and involving no theory of its nature or cause. This disease is of sufficient rarity to make it desirable to record any well-marked example of it, and its true pathology, progress, and proper treatment, are as yet so obscure as to render it a subject of much interest.

CASE.—Mary Jane Campbell, a girl of twenty-three years of age, an assistant in a printing establishment, applied to me for admission to hospital, in April, 1869, on account of a hoarseness and feeling of pain and constriction in her throat, which had then lasted about three weeks, and seemed to be increasing in intensity. Upon examining the throat externally, the neck was found to be the seat of a peculiar kind of diffused induration, which extended across the larynx in front, and laterally as far back as about the edge of the trapezius muscle, the back of the neck feeling natural; the larynx seemed to be the centre of this induration, and felt larger than natural, while at the same time its prominent cartilages could scarcely be distinguished. When pressed the patient complained of pain, but the swelling at all other parts of the neck was painless; the affected skin was pale, and of natural temperature. Upon further examination the induration was found to extend down over the skin of the chest as far as the mamma, the upper part of which was hard, but the skin over the lower part of these glands was supple and healthy. Upon turning to the face to see if the disease had involved it at all, it became apparent that the skin over the entire face had a smooth, tightened, peculiarly drawn look, which made it almost impossible to pinch up the skin, and gave a remarkable expression to the countenance. When told to frown the patient's utmost efforts could only produce a slight wrinkling of the skin of the forehead, while smiling produced creases around her mouth, such as might be the result of folding parchment.

The only history the patient could give was, that about four months previously she had began to find her strength and appetite failing; she was at times sick after meals, sometimes vomiting; she suffered at the same time pains in the shoulders and back, which were worse at night. Three weeks before applying for advice she had felt some difficulty in swallowing, and at the same time began to loose her voice. This increased slowly until she was not able to raise her voice above a whisper; she became at the same time troubled with a constant cough; she was admitted to the hospital upon the 29th of April. Within a week after her admission the peculiar hard swollen state of the integument had extended round from the anterior and lateral region of the neck to the back of the neck and shoulders—where its characters were very marked—the affected integument felt brawney, like the skin of swine; it could not be pinched up. At this time the colour was perfectly natural, but it subsequently became of a yellowish tinge; no pressure would make it pit. The treatment adopted upon the patient's admission to hospital consisted of small doses of Grey and Dover's powder internally, and the *Lin. Hyd. Camph.* externally, rubbed over the whole of the affected skin once a day. Within a week the voice had become nearly natural, and the patient declared herself much better. A note taken upon the 8th of May, states that "an improvement has taken place in all the symptoms; the hoarseness has nearly altogether disappeared, and the peculiar drawn expression of the countenance does not exist to the same degree; the patient no longer complains of tightness of the throat." The skin over front and sides of the neck which had been chiefly affected, gradually became softer and more natural, and could be pinched up, but at the same time the back of the neck which, when the patient was admitted, was free, remained markedly affected, and the whole skin over both shoulders and scapulæ, and over the deltoid region on both sides, the induration gradually decreased down the arm, until at the elbow the integument felt natural, but upon the back of the right forearm, a patch of skin about the size of a crown piece was indurated in the same way as the rest of the diseased integument. When the arm on either side is moved up and down the movements of the scapulæ are not seen as usual beneath the skin; they are, as it were, concealed by the thick integument, and when the hand is laid on the part and the scapulæ moved this became very evident to the touch. No improvement took place in the state of the skin over the back and shoulders before the patient

left hospital, but the face became less drawn in its expression, and at the same time got a furfuraceous look, the cuticle coming off in small heavy scales. She left hospital upon the 29th of May, and returned to her employment of book-folding. There was not at any time any inflammatory or feverish symptoms whatever. The sensibility of the affected skin was tested upon different occasions; it was diminished, but not very much so in any part. The lower part of the body remained quite free from the disease. The urine was free from albumen, and the menstrual functions were healthy.

The great majority of recorded cases of this disease have been observed by French and German authors; however, Dr. Robert M'Donnell, of this city, has related two similar cases in the *Dublin Hospital Gazette*, Vols. for 1855 and 1856. The patients were in the great majority of cases young women. Of seventeen, collected by Dr. Fagge, fifteen were women, and two men, their ages varying from four to fifty-six, but the majority being between twenty and thirty.

The *cause* of the disease was in most of these cases, as in the one now related, obscure; but in some of the best marked instances it was plainly produced by arrested perspiration. Thus Henke relates the case of a girl aged twenty-four, who, one hot summer's day when heated with field work, lay down and slept upon some damp grass in a cool cellar, on her back, her shoulders being bare. When she awoke she felt her neck stiff, so that its movements were difficult and painful; in a few days an induration of the skin of the whole upper half of the body was established. And in another case recorded by Bouchut, and quoted by Dr. Fagge, a man, twenty-three years of age, was exposed to a draught of cold air for several hours while covered with sweat, and upon the next day the symptoms of scleriasis were established. The disease begins most frequently in the *neck*, and thence extends, as in my case, to the upper half of the body, but seldom affects the lower limbs; sometimes a few patches of induration will be found isolated, as it were, from the rest of the affected integument. The symptoms which characterize this disease are sufficiently striking to prevent its being mistaken for ordinary œdema; a peculiar thickening of the whole tissue of the skin, which renders it tense and brawney to the touch, and makes it impossible to pinch it up, without any redness or discolouration at first, but subsequently a faded yellow colour may be noticed over the affected part; sensation is dulled over the part, but not lost. The patient's own sensations are simply a feeling of

constriction or tightness, accompanied by general aching pains in the shoulders or arms, but no acute pain in the part affected, which may be handled freely without complaint.

The natural course of this disease appears to be, slowly to extend for some months from the point where it begins (usually the neck), and slowly to subside and disappear. The subsidence begins, as we might expect, in the part first affected, and the improvement extends very gradually to the parts subsequently affected. In several of the continental cases, the patients have been kept under observation for years, and a gradual improvement, and finally a disappearance of the disease took place without any treatment beyond that adopted at first, and which was not very active. The rapidity with which the symptoms have appeared and progressed varies considerably; in most cases two or three weeks elapsed before the disease was decidedly established, and then as many months before any subsidence took place; in some, years elapsed without any decided change; in one of Dr. M'Donnell's cases the disease had existed a year, and the patient remained under treatment for more than a month without any improvement whatever. In one case only has an opportunity of examining the pathological anatomy of this affection been afforded. Forster, who made the examination, thus sums up his observations—"The scleroma of this case was characterized by a chronic process of proliferation in the connective tissue of the corion, and particularly of the subcutaneous tissue, unattended with fever or local inflammatory symptoms;"<sup>a</sup> the fat usually present beneath the skin had completely vanished, being replaced by the hypertrophied connective tissue. The treatment hitherto adopted, although including a great variety of local applications and internal remedies, has not seemed to make any change whatever in the disease. In my case the laryngeal complication induced me to give for several days alterative doses of Grey and Dover's powder, which was quickly followed by a subsidence of all laryngeal distress and hoarseness, subsequently Lin. Hyd. Camph. was rubbed daily over the affected skin for about a fortnight; no decided effort appeared to be made by this; but the patient, as already stated, left hospital, relieved to a great degree of pain and the feeling of constriction, and able to return to her employment, but still exhibiting the peculiar characteristic induration over the shoulders, neck, and face; warm baths, and cod-liver oil rubbed over the affected integument has appeared in some cases to be of use.

<sup>a</sup> Guy's Hosp. Rep., Vol. xiii., p. 313.