MORPHEA ASSOCIATED WITH HEMIATROPHY OF THE FACE

EARL D. OSBORNE, M.D.

Fellow in Dermatology and Syphilology, the Mayo Foundation, and First
Assistant in Section on Dermatology and Syphilology, Mayo Clinic

ROCHESTER, MINN.

Linear morphea or circumscribed scleroderma following closely a nerve distribution is of relatively common occurrence, but morphea associated with facial hemiatrophy is distinctly a rarity, if one may judge from the literature. Recently, two such cases have been observed in the Section on Dermatology and Syphilology of the Mayo Clinic. Barrs,¹ in 1891, reported a case of this type. Since then, three other cases have been recorded, all from the British Isles. Barrs' patient was a woman, aged 27, whose face was markedly asymmetrical, and the skin and subcutaneous tissue on the right side, from the upper border of the orbit to the level of the teeth in the lower jaw, were atrophied. The eyeball was noticeably sunken. Definite diminution in sensation over the area supplied by the superior maxillary nerve was detectable. Alopecia was not present. The patient had had neuralgia of the right side of the face for many years. In the right inguinal region was a typical patch of morphea.

In 1903, Savill² presented a case of morphea with well marked facial hemiatrophy and called attention to the relation between morphea and hemiatrophy, which he believed to be identical. The patient, a man aged 50, had struck the top of his head while running upstairs twenty years before. Two or three years later, he had noticed that he was losing his hair and eyebrows, and that the left side of his face was becoming sunken and painful. The atrophy was progressive and involved the subcutaneous tissue as well as the skin. Scattered patches of morphea were noted on other parts of the body. A depressed scar was found at the point of injury. At the time of presentation, an affection of the eye had supervened which Savill's colleague, Dodd, declared to be trophic in character.

In 1904, Galloway³ reported the case of a woman, aged 37, with a typical patch of morphea involving the distribution of the first two divisions of the right fifth cranial nerve. The lesion commenced above the orbit and spread back approximately to the lambdoidal suture. There

1. Barrs, A. G.: Cases of Atrophia Cutis and Morphea, *Brit. J. Dermat.* **3**:152-155, 1891.

2. Savill, T. D.: *Brit. J. Dermat.* **15**:106-107, 1903.

3. Galloway, J.: *Brit. J. Dermat.* **16**:20-21, 1904.

was also a small round patch on the upper portion of the right nasolabial fold. Distinct shrinking of the right nostril and ala nasi had occurred, with general atrophy of tissues of the right side of the face, especially around the orbit. The nasal mucous membrane on the right side was dry and atrophied. The eyeball had retreated within the orbit so that the eyes were noticeably asymmetric. The eyelids were unaffected. Bilateral deafness due to otosclerosis was also present.

In 1886, Steven⁴ observed a remarkable case of scleroderma in a girl, aged 14 years, with pronounced hemiatrophy of the face, body, and extremities of the right side. When the patient was first examined, she complained of a moderately well advanced scleroderma of the right side of the body, of about two months' duration. The condition was first noticed as a hard patch of skin in the right groin; shortly afterward, the right hand became swollen. These lesions gradually extended, and new lesions were noticed on the upper right arm, extending over the shoulder and scapula. Seven months later, the condition was found to be much worse. The whole of the right arm was involved except for a small patch at the lower end of the radius. The right hand could neither be opened nor closed perfectly, and the arm could not be straightened at the elbow. The affected skin was more or less deeply pigmented, being brownish. On the abdomen, the condition had extended from the level of the umbilicus to Poupart's ligament, and was sharply demarcated at the middle line. The whole of the skin on the anterior surface of the right leg, from the groin to the toe, was extensively involved and presented all the characteristics of typical scleroderma. Besides this, there were a few small patches developing on the left arm, around the wrist, between the thumb and forefinger, around a vaccination scar, and in the axilla. There were also several small patches on the left side of the abdomen. The patient was in excellent general health. Sensation over the affected areas was normal. A month later, a patch was observed on the right temple and a very small cicatrix on the left. In 1897, Steven⁵ made an extensive report on this case, the chief characteristic at that time being marked atrophy of the right side of the body, more particularly affecting the face, arm and leg, and very extensive and advanced scleroderma, which was not so strictly limited to the right side of the body. Atrophy of the

4. Steven, J. L.: Case of Scleroderma Adultorum, *Glasgow M. J.* 26:280-283, 1886.

5. Steven, J. L.: A Case of Scleroderma, Leading to Pronounced Hemiatrophy of the Face, Body and Extremities, with Deformity and Fibrous Ankylosis of the Joints, After a Lengthened Period of Superficial Ulceration, *Internat. Clin., S. 7*, 2:195-202, 1897; Case of Scleroderma with Pronounced Hemiatrophy of the Face, Body and Extremities, Death from Ovarian Tumor; Account of the Postmortem Examination; a Sequel, *Glasgow M. J.* 1:401-408, 1898.

right side of the face was pronounced, being most marked on the chin and forehead. The right side of the lower jaw was markedly diminished in length, and a depression of the surface over the right temple and side of the frontal bone was distinctly visible. The depression of the forehead, however, began a little to the right of the middle line. The cutaneous sensibility and the hair were unaffected. The right arm and leg were practically universally involved in the sclerodermatous process associated with pronounced atrophy of the subcutaneous tissues of the fingers and toes. There were, besides, several old patches of scleroderma on the left side, evidently the patches seen at the time of the first examination eleven years before. No new patches were observed on this side.

The histories of both the parents and the patient were essentially negative, except that three years before the onset the patient had accidentally drunk vitriol, with resulting difficulty in swallowing any but soft foods.

Twelve years after the first examination, the patient died, following an operation for ovarian tumor. When the brain was removed at necropsy, the cortex of the motor area was found to be slightly thinner in the left hemisphere (opposite the affected side), than in the right.⁶ Sections from various levels of the cord were examined. The findings were as follows:

In each, there was a slight, though undoubted, diminution in size of the right anterior cornu (the affected side). The ganglion cells in this horn, besides being fewer in number than normal, presented signs of degeneration, in that their nuclei and plasma granules were not nearly so well defined as in the ganglion cells of the left horn. The neuroglia, too, in the right horn seemed denser than in the left. Changes corresponding to those just mentioned could not be made out with any certainty in either the medulla or the pons. Throughout the cord, medulla and pons, the arteries, especially those in the gray matter, were surrounded by certain spaces, sometimes containing a homogeneous structureless material, and in some cases free from any such contents. On transverse section, these spaces were round or oval in shape and most of them had sharply defined margins. The appearance they presented suggested dilated lymph spaces rather than a softening of the gray matter. They were best seen around the anastomotic arteries and, on the whole, were larger and more apparent on the right side of the cord than on the left. The nerve fibers from the cervical and lumbar plexuses showed a well marked parenchymatous degeneration. This change was evidently recent, and almost certainly had no connection with the more chronic changes in the cord.

Steven, in summarizing his observations on this case, concludes:

1. Scleroderma with hemiatrophy, as regards its essential pathology, is a trophoneurosis due to changes occurring in the trophic cells of the central nervous system, certainly in those of the spinal cord, and probably also in those

6. Unfortunately, a histologic study of the cortex was not made.

of the brain. As a result of this change in the nerve cells, the blood vessels, both of the cord and of the periphery controlled by the diseased nerve cells, are first of all affected.

2. Following upon the derangement of the blood supply caused by the central nervous disease, the atrophic changes in the skin and subjacent tissues are slowly developed.

REPORT OF CASES

CASE 1 (A347956).—A schoolboy, aged 9, was brought to the Mayo Clinic because of a depression over the left eye (Fig. 1). At birth, he was delivered by forceps; and he developed a large hematoma over the vertex which it was necessary to open. Measles and pertussis were his only illnesses until three years before, when a small whitish scar with a blue border was noticed below the left eye. The father referred to it as a "black eye." No history of trauma.

Fig. 1 (Case 1).—Morphea and hemiatrophy of the face of a boy, aged 9 years.

other than at birth, was obtainable. Four months after the appearance of the white scar, the left eyebrow began to thin and a depression which appeared over the eye gradually extended onto the forehead.

Examination revealed a typical patch of morphea on the cheek below the left eye and extending around the inner canthus onto the supra-orbital ridge. The skin was smooth, firm, and slightly elevated, with a definite violaceous border, especially marked below the eyes. There were no other patches on the face or body. The skin and subcutaneous tissue of the left side of the face were atrophied and the left ala nasi was retracted. The left eyeball was considerably sunken in the orbit. A partial alopecia involved the left side of the scalp anteriorly. The whole picture was one of definite asymmetry of the face. No changes were seen in the mucous membrane of the nose or throat. A careful neurologic examination did not reveal sensory changes in the area involved. There was, however, a suggestion of an extensor type of reflex in the left foot;

that is, an occasional Babinski, Oppenheim and Gordon reflex. Because of these findings, the consulting neurologist suggested a possible old inflammatory process in the brain.

CASE 2 (A321327).—A girl, aged 7 years, was brought to the Mayo Clinic on account of an old fracture of the left humerus with limitation of motion of the elbow on that side (Fig. 2). Ten months before, she had fallen down a short flight of stairs, with consequent injury to the left elbow, and had since been unable to move her left hand. Sense of touch was absent. For some weeks, the arm was kept in a cast and later in a sling. Six months before examination, her parents had noticed slight thickening of the skin over the left side of the face and neck; this had gradually extended, but had not caused symptoms, except slight stiffness on movement of the head.

Fig. 2 (Case 2).—Morphea and hemiatrophy of the face in a girl, aged 7 years.

Examination revealed that the child was healthy and of normal weight and height. A diagnosis of ischemic paralysis of the left forearm and wrist had been made in the orthopedic section of the Clinic. A roentgenogram of the left elbow was negative. There were several discrete, firm, smooth, irregular patches of morphea on the skin behind and in front of the left ear and on the skin of the neck on that side. These patches were slightly elevated and had a very faint violaceous border. The hair was thin in front of the left ear. The chin was definitely asymmetric, and there was early atrophy of the skin and subcutaneous tissue of the entire left side of the face; but change could not be demonstrated in the bones of the skull. The mucous membranes of the nose and throat were normal. The cutaneous sensibility appeared normal. The basal metabolic rate was +6 per cent. The Wassermann reaction on the blood was negative.

Because in these two cases morphea began definitely at the same time or shortly before hemiatrophy and the conditions existed together, it was thought that a review of the literature might reveal a common pathologic background and throw some light on the hitherto unknown etiology of both conditions. The cases of Barrs, Savill, Galloway, and Steven are the only ones in which a positive diagnosis could be made from the authors' descriptions. There are numerous cases reported of facial morphea, especially of the frontonasal type, in which coexisting hemiatrophy may be suspected from the description, but in which a positive diagnosis is not made by the reporter.

Some dermatologists question whether facial morphea and facial hemiatrophy are distinct entities. Sequeira,⁷ who believes in a ganglionic neurotrophic origin of morphea says that progressive hemiatrophy of the face, involving bone, muscles and skin, sometimes occurs in conjunction with morphea. Hyde⁸ classifies hemiatrophy of the face under scleroderma, but does not commit himself as to the etiology. Highman writes⁹: "Hemiatrophy facialis is scleroderma of one side of the face, which involves the deeper tissues. It is rare and usually accompanied by other evidences of scleroderma." On the other hand, neurologists are in accord in considering the two conditions as separate entities. Practically every textbook on neurology describes skin changes as one of the first symptoms of facial hemiatrophy. A white spot appears on the side of the face and gradually becomes larger, or, if there is more than one, they coalesce and become yellow or yellowish-white. The skin over these patches sinks and forms pits or trough-like depressions, owing to the disappearance of the subcutaneous fat. Pigmentation may occur not only in the areas where atrophy is marked, but on other parts of the face. This pigmentation is in small spots or patches, and varies in color from a grayish-yellow to brown or blue. The hair of the head, the beard, and the eyelashes may lose color and fall out in patches. The sebaceous glands are atrophied, and their secretion diminished and eventually checked. The secretion of perspiration is often increased.

The foregoing description of the skin changes in facial hemiatrophy is taken from one of the well recognized reference books on neurology.¹⁰ The close similarity between the skin changes of facial hemiatrophy and morphea is apparent. It seems highly probable that the skin changes in patients who later develop hemiatrophy are of the morpheic variety.

7. Sequeira, J. H.: *Diseases of the Skin*, London, Churchill, 1915, 658 pp.

8. Hyde, J. N.: *A Practical Treatise on Diseases of the Skin for the Use of Students and Practitioners*, Philadelphia, Lea & Febiger, 1909, 1126 pp.

9. Highman, W. J.: *Dermatology. The Essentials of Cutaneous Medicine*, New York, The Macmillan Company, 1921, 482 pp.

10. Dercum, F. X., Ed.: *A Text-Book on Nervous Diseases by American Authors*, Philadelphia, Lea Brothers, 1895, 1056 pp.

In this connection, attention is directed to a case recently observed of typical hemiatrophy of the face of twelve years' duration (Fig. 3, Case A298444). The skin changes could very well have been the remains of an old involuted patch of morphea. The only diagnostic point lacking was the violaceous border typical in active cases. This patient also had had frequent typical epileptic attacks for eight years. The two other patients (Figs. 1 and 2) were observed in childhood, during the early years of the disease. In both cases, the morphea was typical and well advanced, whereas the hemiatrophy was just beginning. There are on record between 200 and 300 cases of hemiatrophy of the

Fig. 3.—Hemiatrophy of the face in a man, aged 27. The scar of scalp and forehead is indistinguishable from that of an involuted patch of morphea.

face. It is impossible to estimate how many of these cases presented patches of morphea at some time. It is probable, however, that many of them would have been diagnosed by an experienced dermatologist as morphea associated with hemiatrophy of the face. Besides the similarity in symptoms between the two conditions, the recorded etiologic factors are practically the same, although the exact cause of each is unknown. The accompanying tabulation presents the supposed etiologic factors of both conditions according to standard works in dermatology and neurology.

SUPPOSED ETIOLOGIC FACTORS IN MORPHEA AND HEMIATROPHY FACIALIS

Morphea

1. Both sexes, females predominating.
2. Occurs at all ages, especially from 15 to 45 years.
3. Exposure to cold and wet and prolonged exposure to the sun are predisposing causes.
4. Often follows infectious diseases, such as rheumatism and erysipelas.
5. Often accompanies emotional and nervous states.
6. Often follows exhaustion from any source.
7. Local irritation or injury is supposed to play a part.
8. Thyroid diseases are as yet factors little understood.

Hemiatrophy Facialis

1. Both sexes, females predominating.
2. Occurs at all ages, especially from 10 to 40 years.
3. Exposure to cold and extreme temperatures are predisposing causes.
4. Often follows infections, such as erysipelas, scarlatina, pneumonia, malaria, tuberculosis and rheumatism.
5. Often accompanied with psychic disturbances, such as grief, anxiety and worry.
6. Often follows anemias and scrofulosis.
7. Traumatism sometimes precedes.

The etiologic factors are practically the same in both conditions except for the possible rôle of the thyroid in the production of morphea.

In summarizing the six cases reviewed, the evidence points strongly to involvement of the central nervous system. The history of trauma from four months to six years preceding the onset of symptoms in four of the cases, the necropsy findings of Steven, and the neurologic findings suggesting an old inflammatory process in the brain of one of the patients in the Mayo Clinic, are all incriminating evidence. The close relationship between facial hemiatrophy and morphea is emphasized by the common etiologic factors involved and in the similarity of skin changes seen in the two conditions. That the early skin changes seen in cases of facial hemiatrophy are always of the morphea variety seems unlikely, but that they may often be morpheic seems highly probable. Further pathologic studies of the nervous system in cases of morphea and scleroderma coming to necropsy may reveal the essential etiology of these obscure conditions.