Two cases of hemiatrophia facialis / by Walter H. Jessop and Oswald A. Browne.

Ernest Hart [?] from the author
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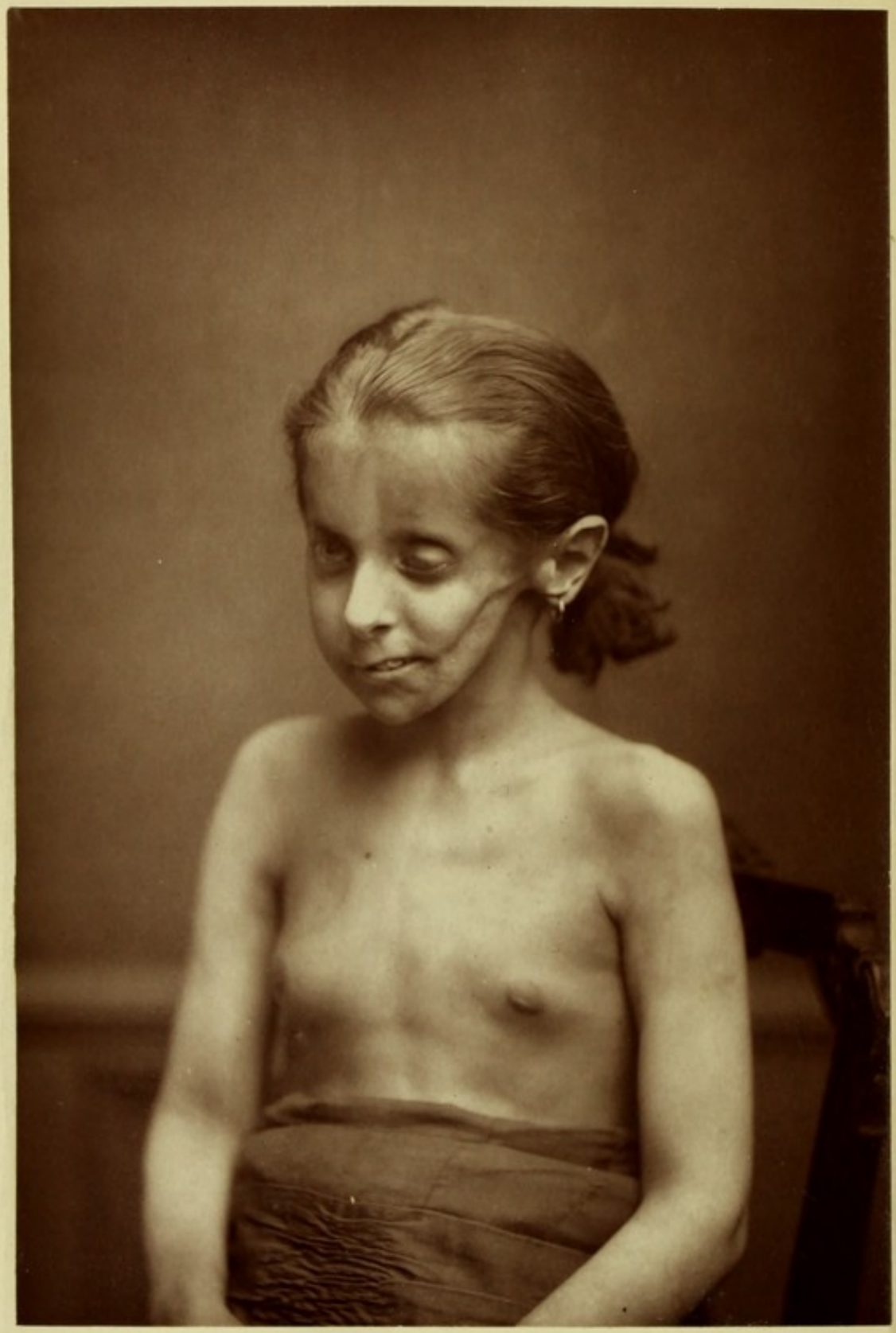
Jessop & Browne

Hemiatrophia



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TWO CASES OF HEMIATROPHIA FACIALIS.

BY

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AND

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We are deeply indebted to the kindness of Dr. Gee for the use of the notes of both cases, and to Mr. Streatfeild for the early notes of Case I., under whose care she was at the Royal Ophthalmic Hospital, Moorfields. Our thanks are also due to Mr. Gell and Mr. Santi for the excellent notes they have taken as clinical clerks of the respective cases.

Case I.

Agnes B., aged 12, was admitted into Mary Ward, under Dr. Gee in September, 1882, and is the fifth of seven children, one died at five months of congestion of the brain, a son, aged 14, has gone out of his mind during the last two months; father very excitable, mother has been out of her mind since her last confinement, two years ago; was much excited and subject to delusions before and after the birth of the child Agnes.

The father gives the following account of the child:—

She was quite well till five years ago, when a strip of slightly drawn shining yellow skin was for the first time noticed on the side of her left cheek; increasing for a year, and then the colour entirely disappearing.

About the same time a similar patch was noticed along the middle of the dorsum of the right hand, followed by wasting and rigidity of the middle finger. Shortly after this grey hairs were noticed on left side of head, and then commenced the wasting of the left side of the face, which has from that time

till three months ago increased, the latest changes being increase in the number of grey hairs.

Two years ago the sight of the left eye was affected.

In June she applied at the Royal Ophthalmic Hospital, Moorfields, and was admitted under the care of Mr. Streatfeild for an opacity of her left cornea. This opacity, which was suspected to be a film of lead, was sliced off by Mr. Streatfeild, leaving the eye in its present condition as detailed below.

Patient is a bright, intelligent, good-tempered child of medium height and build, and apparently in the best health and spirits; has never had any illness except this; stands or sits with her head slightly inclined, and face bent to the left.

On looking at right side of face she presents the appearance of a healthy well-nourished girl of 12, but on left side that of a decrepit and haggard old woman.

The atrophy which distinguishes this case is **unilateral**, being confined to the left side of head and neck and of upper part of trunk, with the exception of the adjacent side of the right middle and ring fingers.

The right side, with the exception above stated, is well-nourished, the skin, muscles, bones, &c., being well developed.

The cyrtometric tracing of the head at the level of the occipital protuberance, and of the superciliary ridges, shows a marked diminution in the left half circumference; a deep groove runs in an antero-posterior direction, marking the frontal and parietal bones, a little to the left of the sagittal suture.

The hair is dark, distinctly thinner on the left side, and there are several grey hairs on the left side.

Left side.—Skin has a glazed withered appearance, somewhat darker colour, and not so moveable over the bones as on the right; the subcutaneous tissue is considerably atrophied, and, besides the muscles, the bones have joined in the general atrophy.

Anterior part of occipito-frontalis is quite atrophied, also the corrugator supercilii; small cutaneous veins well marked, and almost total absence of lanugo, and small hairs.

Eyebrow, very few hairs; great depression above upper lid.

Anterior branch of temporal artery very prominent, but normal size.

Upper eyelid, slight ptosis, and can only be raised about half-way; wasting of tissues composing both eyelids; eyelashes fairly well developed in upper lid, scarcely any in lower lid; extrinsic ocular muscles act well.

Conjunctival vessels injected, slight circumcorneal zone.

Cornea hazy and vascular, towards outer and lower part

numerous vessels going to an ulcerated surface, which is apparently healing, and is the seat of the lead deposit removed by Mr. Streatfeild. Anterior chamber very shallow.

Vision.—Perception of light.

Nose.—Tip inclined to left. Ala nasi much atrophied, even to the cartilages.

Temporal fossa.—Great wasting of temporal muscle. Zygomatic process much atrophied.

Over situation of buccinator muscle is a round soft swelling, which is especially apparent from the extensive atrophy around, and is chiefly fat.

Deep groove extending from inner canthus toexternal angle of mouth, due to atrophy of all the tissues.

Ear.—Stands out from side of head, being made more prominent by obliteration of angle of jaw, and extensive atrophy of soft parts around; whole substance, including cartilage, atrophied. Hearing good.

Mouth.—Drawn up towards left side; the two upper central incisor teeth are seen through the chink due to atrophy of upper lip.

Labellum only developed on right side, giving rise to small prominence.

Upper lip.—Extensively atrophied; puckering of skin near angle; hairs scarcely developed over middle.

Lower lip.—Atrophy of all parts, including mucous surface

Chin.—Great atrophy of soft parts.

Lower jaw.—Total obliteration of angle; jaw slopes from symphysis to condyle without any break.

The tongue, on protrusion, is strongly pushed over to the left side; no apparent atrophy, or of the soft palate. Taste equal, and good.

The teeth appear normal, but the upper back teeth fall within the lower when the mouth is closed; the left alveolar border is flattened, and the line of teeth is thus deformed.

The muscles of the left neck are wasted; the skin and subcutaneous tissue, also atrophied, is stretched over them.

The chest seems slightly tilted over to the left, and there is wasting of the left mamma; a thin white line is seen, as if drawn with a finger-nail, marking the middle line of the chest and abdomen; the muscles of the left arm are more flabby than the right; very distinct wasting of the left pectoralis major; the acromial extremity of the clavicle is unusually prominent; slight glossiness of the skin at the upper part of the arm; the left deltoid, supra-spinatus, and infra-spinatus muscles are distinctly wasted.

Right hand.—Dorsal surface—skin in middle is atrophied and

shining, the tendons of the extensor communis digitorum to the middle and ring fingers being plainly visible; depression over dorsal interossei of second and third spaces; atrophy of middle finger and outer half of ring finger, the inner half of ring finger being normal.

Palmar aspect—great depression over centre of palm, contraction and thickening of skin, almost simulating Dupuytren's contraction; middle finger and outer side of ring finger atrophied, apparently involving all the tissues. Can extend and flex these fingers completely, though not so powerfully as other fingers.

The cutaneous sensibility is unimpaired over the atrophied parts; the faradic excitability of the atrophied muscles is good.

In order to test the sweat glands, $\frac{1}{8}$ of a grain of nitrate of pilocarpin was injected under the skin of the left arm. In five minutes beads of sweat formed on the upper lip, chin, and nose of the right side only; the skin of the right side of the face became also generally moister, that over the left remaining unaltered; the right side of the lips became moist, the left being dry. In about fifteen minutes the skin of the left side of the face and neck became slightly moister, and beads of sweat formed on parts of the left side of the nose and lips; slight flush on right side of face, but none on left. Little or no increase of salivation.

Measurements.

Tip of nose to outer canthus . .	left	$2\frac{1}{8}''$	right	$2\frac{5}{8}''$
Tragus to outer canthus . . .	,,	$2\frac{3}{4}''$	,,	$3\frac{1}{8}''$
Tragus to angle of mouth . .	,,	$3\frac{1}{4}''$	,,	$4''$
Fissure of mouth	,,	$\frac{7}{8}''$	,,	$1\frac{1}{8}''$
Palpebral fissure	,,	$\frac{7}{8}''$	,,	$1''$
Width of zygoma about middle .	,,	$\frac{2}{8}''$	,,	$\frac{3}{8}''$

All the organs of the chest and abdomen are normal.

Catamenia have not yet commenced.

Case II.

Elizabeth B., aged 48, unmarried, monthly nurse; was admitted into Mary Ward, under Dr. Gee, in November, 1882. Patient gives the following history:—When two years old, slept in a damp nightcap for three or four hours; on the day following the left eye was inflamed, and became so much worse that she had partial loss of vision in that eye.

The ocular pain was very severe, and affected her head. At the spots where the pain in the head was most intense, loss of hair permanently occurred—viz., (1) just above left frontal eminence, close to the middle line; (2) behind left ear, over mastoid

process; (3) a little to the left of mesial line of the head, about the lambdoid suture. These spots are all on the left side of the head, and the hair on that side is thinner and scantier than elsewhere.

At five years of age, the pain in the head becoming gradually more severe, she became totally blind, and continued so for six months, after which time she recovered her sight and could see well. Between ages of two and five patient had measles, scarlet fever, and constant hacking cough.

With disappearance of ocular trouble she had pain and inflammation of left side of chest, with severe cough, accompanied by occasional hæmoptysis, and had blisters frequently applied There is now at base of sternum a square scar caused by constant blistering.

As regards the facial atrophy scarcely any history can be obtained; patient says that when her eye became first affected at two years old, sinking in of the left cheek was observed. She had teeth in both jaws up to sixteen years; had toothache frequently; soon after sixteen the teeth on the left side began to drop out, and had all done so by eighteen. The right teeth were not lost till some years afterwards.

The facial atrophy increased up till thirteen, when it reached its present condition.

At nine years old underwent an operation on left eye at St. Thomas's Hospital, probably for lachrymal obstruction.

With the exception of almost continuous cough patient has been in good health since fifteen years old.

Family history good.

Patient is a healthy looking, well-nourished woman, with the exception of left side of face, of medium height, and average intelligence; holds her face always turned to the left, thus hiding her deformity.

The right side of the face gives one the idea of perfect health, the muscles, bones and skin being apparently normal, as they are also in every part of the body, except the left half of the face.

The viscera of the body are also apparently healthy.

The following notes have reference to the left side:—

The hair, which is dark, is scantier than on the right, and absent for 3″ radius above ear.

No marked demarcation along mesial line of forehead, skin much wrinkled, prominence of inner part of frontal eminence, but depression towards outer part; no difference in cyrtometric tracing of skull.

Elevation of eyebrow, depression over corrugator supercilii.

Eye rather protruded, lagophthalmos; upper lid atrophied,

absence of furrows in skin of upper half, veins very prominent, entropion, nearly entire absence of lashes; punctum patent, (never remembers eyelashes, but said to have had them at two years old), thickening of palpebral conjunctiva.

Lower lid.—Outer half nearly altogether atrophied, palpebral conjunctiva visible only at extreme outer and inner canthi, in middle no demarcation between it and skin, punctum everted and of no use.

Cornea nebulous; iris acts well; pupil normal, and equal to right.

Lens.—Normal.

Fundus.—Myopic crescent.

Vision.—Large objects; has had the eye continually shaded for 33 years.

Nose.—Slight deviation of septum to left, wasting of muscles, especially of alæ; aperture of anterior nares contracted.

Temporal fossa.—Great wasting of temporal muscle, and prominence of temporal artery, but apparently normal size.

Zygoma much atrophied, and feels like thin sharp bar through stretched and atrophied skin; malar bone atrophied in whole length; below malar bone, great depression over buccinator, which seems quite atrophied.

Ear.—Stands away from rest of face, and of greater width than right, but small, and well developed, except absence of lobule. Hearing good on this side, but total deafness on right side; till four years ago could hear well on both sides.

Mouth.—Upper lip angle drawn up, labellum towards left, at angle a mass of muscle and fat stands out over position of orbicularis, in contrast to the buccinator depression. Lower lip more affected than upper, thinning of mucous surface.

Chin.—Line of demarcation towards left of middle line, great depression due to atrophy of soft parts, and also of inferior maxilla, especially at angle; masseter much atrophied.

Gums.—Alveolar borders atrophied and edentulous; mucous membrane of the cheek is adherent to the upper border of the lower alveolus; cannot bite food on this side.

Tongue.—Much atrophied, in fact the right side apparently has increased in size, and so taken the place of the left, the latter being only half the width of the former. Taste equal and good on both sides. The pillars of the fauces and soft palate atrophied, uvula deviates to the left.

No atrophy or degeneration of the skin visible below the face. Whole of left side of body distinctly hyperæsthetic; in drawing a pin lightly over left side of face, patient visibly winces, and complains of pain, but on right side the same action gives no

pain. The left facial muscles react well to faradism, much more so than the right.

Measurements.

Tip of nose to outer canthus . .	left	$2\frac{1}{8}''$	. .	right	$2\frac{3}{4}''$
Tragus to outer canthus . . .	,,	$2\frac{1}{2}''$	. .	,,	$3\frac{1}{4}''$
Tragus to angle of mouth . .	,,	$3''$	. .	,,	$3\frac{3}{4}''$
Fissure of mouth	,,	$\frac{5}{8}''$	. .	,,	$1''$
Palpebral fissure	,,	$\frac{7}{8}''$	. .	,,	$1\frac{1}{8}''$
Width of zygoma about middle .	,,	$\frac{1}{4}''$	. .	,,	$\frac{3}{8}''$
Mid chin to angle of jaw . . .	,,	$3''$	. .	,,	$3\frac{1}{2}''$
Tragus to angle of jaw . . .	,,	$1\frac{1}{2}''$	. .	,,	$2\frac{1}{4}''$

Remarks by W. H. Jessop.—Hemiatrophia facialis is the name generally given to the disease of which the above-described cases are two excellent types; this term, though unsatisfactory, expresses the chief and most marked feature of all the recorded examples, and has usually been associated with the adjunct unilateral. The notes of Case I., however, show for the first time that the disease, besides invading other structures on the same side as the facial atrophy, may extend to the opposite side of the body.

The first case was recorded in 1825 by Parry,[1] and since that time forty-five cases are reported, only two being English,[2] the rest being nearly all German; of these twenty-six are women, seventeen men, and two children, six not stated: twenty-nine occurred on the left side, thirteen on the right, in three cases the side not given; thirty-five cases commenced before the age of twenty, and thirteen of these under three years; one case began at thirty-five, another at sixty-two, and in one only was the atrophy from birth;[3] in one case was hereditary shown by Seeligmüller as the sister of the child's mother had hemiatrophia.

Some of the supposed or predisposing causes were, falls on the side of the head afterwards affected, a shot in left shoulder, burn of left half of face and shoulder, apoplexy, epilepsy, chorea, scarlet fever complicated with abscess of tonsil, measles, paralysis (infantile ?), erysipelas.

In one case syphilis[4] was apparently the cause, and, besides the partial left-sided atrophy in temporal and masseteric regions, there was paralysis of the sixth nerve.

[1] Parry, Collections from the Unpublished Writings, i. p. 478.
[2] Moore, Dublin Quarterly Journal, 1852. Buzzard, Clinical Society's Transactions, 1874.
[3] Emminghaus, Arch. f. klin. Med., Bd. xi. S. 103.
[4] Von Graefe, Berl. klin. Wochenschr., 1868, No. xi. p. 129.

Most of the cases were preceded or accompanied by neuralgic pains along the cutaneous strands of the trigeminus, toothache being a prominent symptom.

In many, as in Case I., a pigmentation of yellowish colour was the first sign, and this disappeared as atrophy commenced; an early sign is the sinking in of the eye on the affected side.

The chief characteristics of this disease are its usually occurring without any known reason, generally on the left side of the face, in females during the early years of life, neuralgic pains, especially toothache, yellowish pigmentation at commencement, rapid atrophy of skin, subcutaneous tissue, muscles, bones, cartilage, chief seat over muscles supplied by trigeminus and facial nerves, ocular mischief, hyperæsthesia or normal sensation, never anæsthesia, muscular irritability not diminished.

The ocular troubles are early symptoms in the disease, and consist of narrowing of the palpebral fissure, sinking in of the eyes, atrophy of the orbit, dilated or contracted pupil, corneal nebulæ and ulcers, conjunctivitis, lagophthamos, and later, as the result of these trophic changes, destruction of the eye.

With regard to the two cases recorded above, most of these general signs were noted, but, in addition, the following rarer facts :—In Case I. the atrophy, besides invading the left face, extended down the neck, over the clavicle to the chest and shoulder; the atrophied left lower jaw had complete absence of its angle. But the most remarkable point is the atrophy of the right middle and ring fingers, without any sign of atrophy elsewhere on the right side.

As to Case II., the extreme atrophy of the left half of the tongue deserves mention, also that of the soft palate.

With regard to the causation of these interesting cases many theories have been advanced. Amongst which are the following:—

1. Essential atrophy of all the structures.
2. Atrophy commencing in the subcutaneous tissue by formation of fibrous tissue, which contracts and destroys the endings of the blood-vessels, thus causing atrophy of the skin and afterwards of the muscles.
3. Lesion of the trophic nerves, giving rise to a trophoneurosis (Romberg).
4. Irritation of the trigeminus and facial nerves.
5. Irritation of the cervical sympathetic (Seeligmüller).
6. Arrest of development.

With regard to 2, the atrophy is quicker than a general fibrosis would account for, and also there is no thickening of the skin or subcutaneous tissue, unfortunately I can find no account of microscopical sections of the parts.

As to 4, though many cases have their atrophy limited to the regions supplied by these nerves, yet, in Case I., such a lesion could not explain the atrophy below the face, and it would be hard to imagine why the atrophic changes should suddenly appear in the course of the cervical nerves.

5. Many of the symptoms of this disease may be accounted for by a sympathetic irritation, such as alteration of size of pupil and lid, sinking in of eye due to atrophy of orbital fat, altered pigmentation, disordered action of the sweat glands, scanty lachrymation. Samuel says lesion of vaso-motor nerves, causing diminution of blood circulation, cannot be conceived as the cause of this disease—inflammation, softening, gangrene would ensue, but not simple atrophy.

In irritation of the sympathetic there is always diminution of sensibility, and the constriction of the vessels after a time passes off into dilatation. No lesion of the sympathetic has yet been found giving rise to such extensive atrophy, though in some recorded cases the cervical sympathetic has been so pressed on by growths as to be obliterated.

One point in reference to its being the primary cause has been observed—namely, that pressure over the cervical ganglia has produced pain and tenderness; also that long-continued application of a continuous current to the cervical sympathetic ganglia has produced improvement in the symptoms.

As to 6, in the case of apoplexy at 62 years of age,[1] which developed this disease, the skin, muscles, bones, and cartilage all quickly shared in the complete left facial atrophy.

Besides this example in contradiction to the theory, in our Case I., the complete absence of the angle of the inferior maxillary bone is an impossibility, if the atrophy only commenced at five years old. The father, a highly educated man, brought a photograph of the child at five years old, previous to the atrophic changes, showing that the two sides of the face were similar and well developed up to that time; he was also very positive as to the time of commencement of the disease, and the early signs.

In trying to account for the lesion in the two cases the following facts with regard to the nerve supply of the affected parts may be considered:—

The atrophy in each case commenced over the region supplied by the trigeminus, this being ushered in by neuralgic pains especially along the dental branches. The skin has not suffered at any time from anæsthesia, but was altered as to its pigmen-

[1] Pissling, Zeitsch. d. Wien. Aerzte, 1852, S. 494.

tation. The eye in both cases was the seat of early mischief, and, after undergoing conjunctival irritation, ulcers and nebulæ formed on the corneæ. In Case II., the cornea was only superficially ulcerated; but in Case I. this ulceration has involved the deeper layers of the cornea, and the mischief is still progressing, though the eye is kept protected, and the atrophy elsewhere has stopped. In Case I., besides the disorganization of the eye, giving rise to loss of tension, there is marked sinking in of the eye, due probably to wasting of the orbital fat, and in both there is narrowing of the palpebral fissure.

In Case II. the soft palate and uvula are also affected.

All these regions come superficially in the distribution of the trigeminus and its branches.

In both cases there is marked atrophy over the parotid region, behind the ear, and affecting the structure of that organ.

In Case I. the changes extend to the neck, over the clavicle, to the thorax and shoulder.

These changes occur in the distribution of the superficial cervical nerves—namely, the auriculo-parotidean, small occipital, superficial cervical, sternal, clavicular, and acromial branches, the last-named passing over the acromion to the outer and back part of the shoulder.

Thus, as far as the left side is concerned, in each case we are enabled to limit the atrophic areas to the distribution superficially of the trigeminus and cervical nerves.

Physiological and pathological research may help us by the enumeration of the following facts:—

Meissner[1] divided the internal fibres of the trigeminus and found trophic changes in the eye, but not if these fibres were unhurt. He thus supposes that these internal fibres are special tropic fibres.

Samuel[2] irritated the Casserian ganglion by needles, and afterwards myosis, hyperæsthesia of the lids, conjunctiva and cornea, inflammation commenced, giving rise to ulcer of cornea and panophthalmitis.

Bock relates a case of sclerosis and enlargement of the Casserian ganglion giving rise to violent neuralgia; no anæsthesia of face, but ulcer of cornea, and subsequent perforation of eye.

Friedreich cites a case of sarcomatous tumours pressing on left trigeminus, in which there was great pain in left face, lachrymation and conjunctivitis; sensibility was always normal in left face.

[1] Meissner, G., Henle & Pfeufer's Zeitsch xxxix. 96–104.
[2] Samuel, Die trophischen Nerven, 1860, p. 61.

Charcot[1] gives several instances proving the same facts.

These instances and several others apparently start us on a theory for the ocular troubles—namely, stimulation of the Casserian ganglion, which will give all the necessary effects. Many have tried to explain these ocular trophic changes as due to anæsthesia, and then the presence of foreign bodies irritating and producing the ulceration. But in these experimental cases, and also in our clinical ones, the sensibility is normal, or rather exaggerated. If we could prove the existence of purely trophic nerves, we should then be able to account for the changes in this disease.

In Case I., the atrophic areas, being superficially supplied by the trigeminus and the cervical nerves, immediately bring before us the similarity between these nerves—viz., that each has a motor and sensory portion, and also that in the sensory part is developed a ganglion containing the usual ganglionic cells.

The hitherto accepted view of these ganglia is that they preside over the nourishment of the roots of their respective nerves.

A theory I would dare to put forward is that in these cases changes have taken place in these ganglia, giving rise, probably by sclerosis, to irritation of the nerves in connection with them. This sclerosis has been shown to exist in Bock's case. Also that from these ganglia arise trophic nerves *de novo*, and in consequence of such nerves Meissner's experiments of partially dividing the trigeminus were attended by the symptoms above recorded.

In Case I., then, the areas of disease are supplied by the Casserian ganglion, and those on the posterior roots of the second, third, and fourth cervical nerves, by this theory. A difficulty immediately presents itself as to why these nerves especially should be signalled out, and how they are connected. The intimate connection of the sympathetic system would provide the track for the continuity of the disease from the Casserian ganglion to the other ganglia.

The theory of the trophic nature of the ganglia will fit in with the phenomena observed in our two cases, as far as the left sides are concerned; but there is the atrophy of the right middle and ring fingers of Case I. to be accounted for. This, I would suggest, might be due to some congestion of the ganglion presiding over that area, and its subsequent sclerosis, which would not be harder to imagine than the known sclerosis of large cells in the anterior cornua, giving rise to atrophy of sometimes single muscles apparently indiscriminately picked out.

[1] Charcot sur les Maladies du Système Nerveux, 1880, p. 18.

I have tried to show the reasons for trophic nerves starting these lesions from consideration of the superficial areas involved. In these cases the skin was first affected, the muscles, bones, &c., being affected secondarily, and probably in the same manner.

Thus the theory upheld is that of trophoneurosis as put forward some years ago by Romberg, the sympathetic, however, playing a prominent part by its intimate connections and conducting strands.