degeneration. The patient consented to an operation. A similar incision was made as in the former case and the nerve was found lightly bound down by cicatricial tissue to the tendon of the biceps for about an inch. This was easily dissected off and freed. The wound healed in a week. The muscles were massaged daily and galvanised as soon as possible. The patient soon began to recover and at the present time (three months after the operation) he is well and can do anything. The muscles are not quite so active as are those of the other leg but there is no doubt that they will recover perfectly in time.

Injury or involvement of nerves from traumatism is not uncommon, especially after fractures (in certain situations) and after wounds when they may be divided totally or partially. The two cases recorded above are interesting because they are instances of severe injury to the subcutaneous tissue in which the skin was intact and in which there was no fracture. Such cases are not common and would be likely to occur only in instances in which the subcutaneous injury was very severe and in parts where the nerve was exposed. In both these cases the injury occurred whilst playing football and from the very exposed position of the peroneal nerve as it winds round the head of the fibula it is remarkable that it does not occur more often. In both cases the cause of the paralysis was the result of pressure from firm cicatricial adhesions which produced loss of conductivity in the nerve. The continuity of the nerves was intact and in neither case had they been torn across, which would have been quite possible. It is impossible to tell from clinical evidence whether a nerve has lost continuity or conductivity. Pressure if of a sufficient degree will produce the same effect as complete division of the nerve.

In Case 1 there was complete anaesthesia in the skin supplied by the anterior tibial branch, and at the operation a high division was found and this branch was very much more involved than the other, and no doubt the nerve itself from its appearance had been severely contused and damaged at the time of the accident in addition to the subsequent pressure from cicatricial tissue. In the other parts in both cases the anaesthesia was only partial though the paralysis was complete, and it might be argued that this showed clinically that the nerve was not completely divided, as slight impulses must be passing, otherwise anaesthesia would be complete. Some hold that the partial sensation is derived from other nerves supplying the affected region. Arloing and Tripier¹ described a free anastomosis on the skin of all the nerves entering the dog's paw, which would accord with the latter view. Kennedy² describes cases of injury to the musculo-spiral nerve as a complication of fracture of the humerus in which the anomalies which may be found in the distribution of sensation are very well shown in a series of cases.

Sensation always returns quickly and may be complete long before any improvement takes place in the muscles. This slowness of the muscles to recover probably depends on the muscles themselves and varies with the amount of degeneration; the more advanced it is the longer the period of recovery. In the first case the improvement in the muscles began sooner than one would have expected considering the condition of the nerve found at the operation and the length of time between the injury and the operation.

As long as the muscles give a response to the galvanic current there is hope of recovery and the operation is worth doing. If there is commencing reaction of degeneration in any case with complete paralysis and no sign of improvement six weeks after the injury the operation should be performed without waiting any longer. The sooner the operation is done the sooner is recovery likely to take place, and *vice versa*.

In some cases the muscles show no sign of recovery for three or four months after the operation and the recovery may not be perfect for 12 or 18 months. Speaking generally it may be stated in giving a prognosis in any given case that if the injury has occurred within three or four months recovery will be perfect, but after that time the result will be doubtful. If the muscles will not respond to galvanism at all they will never recover and the operation will then, if undertaken, be of no use except that sensation may return. Sensation has returned in one instance in a case which was operated upon nine years after the injury.

The most important part of the operation is complete asepsis. If this fails the operation will also probably fail. Massage and galvanism are very important afterwards and should be begun as soon as possible. When voluntary power begins to return in the muscles they should be used. It will help them to recover.

Nottingham.

A CASE OF RECKLINGHAUSEN'S DISEASE COMPLICATED WITH A SARCOMATOUS GROWTH INVOLVING THE BRACHIAL PLEXUS.

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IN Recklinghausen's disease there is a combination of multiple lesions affecting the integument and the nervous system. Though the association is sufficiently coherent to justify its being classed as a special disease there is a considerable variation in the prominence of the individual symptoms and many cases have therefore to be regarded as incomplete.

The cardinal signs of this morbid entity which was first isolated by von Recklinghausen¹ are: 1. Tumours of the skin of the nature of molluscum fibrosum. 2. Subcutaneous tumours situated on the nerves, either plexiform or fibro-neuromata; these are multiple and usually of small size, but one of them may reach a large size and require removal. They may occur on the peripheral nerves, on the larger nerve trunks, or close to the nerve-roots, and on the cranial and sympathetic nerves. 3. Pigmentation of the skin; this occurs in two forms—as patches of considerable size and in small specks somewhat resembling freckles. When one of these three essential lesions is absent the disease has been spoken of as incomplete. In addition there are phenomena of secondary importance, such as pain, arthralgia, alteration of sensation, and impaired mental activity. After this brief *résumé* of the nature of Recklinghausen's disease or, as it has sometimes been called, generalised neuro-fibromatosis,² an account of a case recently under observation in St. George's Hospital which presented some interesting features will be given.

A man, aged 32 years, had had all his life a mass of molluscum fibrosum covering the back of the left side of the head and neck and upper part of the left front of the chest and left shoulder. This growth had not increased in size except in so far as it had developed equally with the rest of the patient's body. The skin of the body was generally brownish, but scattered all over the trunk, limbs, head, and neck there were areas where the pigmentation was concentrated either into patches of considerable size, as large as a shilling or in a few instances larger, or in small discrete patches resembling freckles. Neither the hands nor the feet were pigmented. This peculiar pigmentation was not more marked on one side of the body than on the other except that the two largest areas were over the inferior spine of the right scapula and to the right of the base of the sacrum; the latter area measured two inches by two inches. In this respect it contrasted with the localisation of the cutaneous tumours which, as will be seen, chiefly affected the left side. There was more pigmentation on the back than on the front of the body and it seemed to me that the pigmentation had increased since he had been under observation. The patient himself was not aware of the pigmentation, so it is impossible to say how long it had existed or whether it was congenital. One of his children, a boy of about three years of age, whom I examined, had similar areas of pigmentation, but no tumours. Under the skin of the back on the left side of the body there were a number of smooth, firm subcutaneous tumours readily movable under the skin. They were not tender and ordinary pressure did not give rise to any pain in the course of the nerves of the back. They appeared to be multiple fibro-neuromata on the intercostal nerves. Some of them were

¹ Comptes Rendus de l'Académie, tome lxxvii., 1868, page 1058.

² Brit. Med. Jour., Nov. 5th, 1898.

¹ Die multiple Fibrome der Haut und ihre Beziehung zu den multiplen Neuromen, Festschrift, Berlin, 1882.

² For an excellent clinical account of the disease see P. Marie's *Leçons de Clinique Médicale*, Hôtel Dieu, 1894-95, p. 242.

somewhat elongated. There were a few similar subcutaneous tumours under the skin on the right side of the abdomen in front and a larger single tumour of the size of a florin on the inner side of the right groin. There was no correspondence between the distribution of these tumours and the pigmentation of the skin. The patient was perfectly intelligent and showed no sign of any mental deficiency. There had been no diarrhoea, a symptom which had been present in some of the recorded cases. So far the case presented the association of the three pathological conditions which go to make up the morbid entity of Recklinghausen's disease, but the molluscum fibrosum and the pigmentation were much better marked than the multiple subcutaneous fibro-neuromata.

To pass now to the unusual features presented by this case. The patient had had an exostosis on the front of the sternum towards the left border at the junction of the second rib cartilage all his life. In September, 1898, he noticed drooping of the left upper eyelid and about the same time his voice altered and became hoarse and he was under treatment at a special hospital. In December numbness and swelling of the left hand with loss of power developed. When he came under my care in April, 1899, he was found to have a hard tumour, growing apparently from the left side of the vertebral column at about the base of the first dorsal or last cervical vertebra. This tumour lay underneath the molluscous growth on the left supra-clavicular fossa. Skiagrams which were kindly taken by Mr. Addyman did not throw any light on the nature of the deeply-seated growth in the neck, probably because any shadow due to the growth was obscured by that of the spinal column.

Reproduction of photograph of patient with Recklinghausen's disease, showing the molluscum fibrosum and pseudo-ptosis on the left side.

It appeared to be the cause of the following pressure symptoms. 1. Complete ulnar nerve paralysis. 2. Pain in the course of the ulnar nerve. 3. Transient swelling of the left hand, together with a red mottled aspect of the skin. 4. Slight diminution in the size of the pulse in the left radial. 5. Paralysis of part of the left sympathetic chain giving rise: (a) To loss of power of dilating the pupil. The left pupil was small, contracted slightly on exposure to light, and though it could be dilated by atropine did not dilate when darkened. The

patient was perfectly well able to accommodate with that eye for reading. (b) To pseudo ptosis; the left upper eyelid could be raised by the levator palpebrae, but as seen in the accompanying illustration which is reproduced from a photograph (for which I am greatly indebted to Mr. H. G. Drake Brockman) it is relaxed and suggests ordinary ptosis. This relaxation of the upper eyelid can be explained as the result of paralysis of the involuntary muscular fibres in the upper eyelid. The symptom in this case was therefore the exact opposite of Stellwag's sign, or retraction of the upper eyelid as seen in Graves's disease. There was no dilatation of the vessels on the left side of the face or alteration of perspiration or increased growth of hair. The local vaso-motor mechanism had therefore compensated for the paralyzing effects of pressure on the main trunk. 6. The rapid pulse rate which was constantly 128. It has been suggested that the frequent febrile pulse rate of 120 was due to the inhibitory action of the vagus being removed. It seemed reasonable to believe that the cardiac branches of the left vagus were so compressed by the tumour as to bring about a similar result. At the post-mortem examination there were numerous neuromata on the right vagus, so it was unnecessary to suppose that all the cardio-inhibitory fibres ran in the left vagus. 7. Alteration of voice and brassy aneurysmal cough. My colleague, Mr. Sheild, reported that there was paresis, not paralysis, of the left vocal cord, but added that he regarded this as being more probably due to pressure on the trachea or bronchi than to involvement of the recurrent laryngeal nerve. 8. Pressure on the left bronchus as shown by stridor over the left lung.

The patient's right ear presented a scarred appearance resembling that seen after Raynaud's disease, which was due to a chilblain some few winters back. The second and third ribs on the left side of the chest in front were unduly moveable and displaceable, as if some absorption of their substance had taken place near their junction with the sternum.

Progress and termination of the case.—The growth in the neck steadily increased in size and threatened to obstruct the inlet to the thorax on the left side. The patient got thinner and progressively shorter of breath. The tumour was at first thought to be a cervical rib or exostosis, but the possibility of its being a neuroma on the brachial plexus or a sarcoma was raised and became more probable as the growth developed. The question of surgical interference and removal of the growth was considered by my colleague, Mr. G. R. Turner, but it was thought that the great difficulties of removing a growth in that situation and the special difficulty of operating on the part, inasmuch as it was covered by molluscous growth, which would not only render asepsis difficult but probably give rise to very free hæmorrhage, rendered any attempt at removal inadvisable.

On July 2nd and 3rd the patient had several severe attacks of dyspnoea which threatened his life. Mr. G. R. Turner was asked to see the patient in consultation and kindly consented to perform an exploratory operation with a view of, if possible, relieving the pressure symptoms. At the operation a growth was found displacing the trachea towards the right and having such widespread connexions that any attempt at complete removal was out of the question. A little of the growth was removed and was found to have the structure of a spindle-celled sarcoma. Tracheotomy was performed and a stiff rubber tube was introduced. The man was somewhat relieved, but on July 6th his temperature went up to 102° F., his pulse became very rapid (180), and he died on July 7th at 2 A.M. from collapse.

A *necropsy* was very carefully performed by Dr. W. S. Lazarus-Barlow. The body was found to be thin and showed little subcutaneous fat. Small subcutaneous tumours were removed from the left side of the back and from the right side of the abdomen anteriorly. Some, but not all of them, were seen to be on small nerve filaments. They were firm and of a greyish translucent colour, somewhat resembling boiled sago grains. Microscopically they were found to be fibro-neuromata with a certain amount of myxomatous tissue. The tumour in the right groin was found to be merely fat. The tumour in the neck was systematically dissected. It was a diffuse white growth varying in consistency, in places firm, especially near the brachial plexus, while the more outlying parts of the growth were soft. It involved the inner and to a lesser extent the other cords of the brachial plexus. The growth spread down into the thorax, occupying the superior mediastinum on the left side. Microscopically the

growth was a spindle-celled sarcoma with a fair amount of fibrous tissue. It obliterated the left internal jugular vein at its opening into the innominate vein, displacing and compressing the trachea, surrounding the left bronchus, invading the left lung, and destroying the left vagus and its recurrent and cardiac branches. The thoracic duct was involved at its termination and its trunk was dilated, but there were no chylous effusions anywhere. There was recent ante-mortem thrombosis in both innominate veins. The left phrenic nerve was involved. The posterior wall of the trachea was adherent to the growth and there was recent ulceration on the lateral walls of the trachea some three inches below the tracheotomy wound. This may have been due to the pressure of the tube which was in the trachea for three and a half days. The anterior wall of the oesophagus in its middle third was raised up into a prominence by a separate outlying mass of soft growth; this, it would have seemed, must have caused dysphagia, but the patient had been able to take his food without any difficulty until tracheotomy was performed. There were a number of enlarged glands in the immediate neighbourhood of the main tumour. The growth extended down in front of, and behind, the arch of the aorta on the left side, surrounded the left common carotid artery, and was closely adherent to the bodies of the fifth, sixth, and seventh cervical vertebræ which were eroded. The cervical sympathetic was extensively destroyed in this situation.

The roots of the brachial plexus were enlarged, more especially the eighth cervical and first dorsal. These two roots were united into a firm mass of growth which was examined microscopically and found to be composed of nerve bundles embedded in, and separated from each other by, fibrous tissue; in parts there was spindle-celled growth. These appearances suggested that a fibro-neuroma had undergone sarcomatous change. Sections of the thickened trunks of the plexus at a little distance from this mass of growth showed that they were infiltrated with a spindle-celled sarcomatous growth. The affected nerve roots inside the theca vertebralis were also enlarged, but in other parts of the cord the nerve roots were normal. No neuromata were found on the cauda equina or on any of the cranial nerves inside the skull. The spinal cord and the brain were normal. The right vagus was not involved in the growth but showed a number of neuromata in its course and presented a moniliform outline. The left vagus also showed neuromatous enlargement and ran into the thoracic extension of the growth over the arch of the aorta.

Where the growth started it is not easy to say with absolute certainty; it involved the brachial plexus and very probably started in connexion with a fibro-neuroma on the roots of the eighth cervical and first dorsal nerves close to their exit from the spinal column. From the microscopic appearances in this case and from the occurrence in other examples of Recklinghausen's disease of large neuromata on the brachial plexus, which may become sarcomatous, I am inclined to believe that the sarcomatous growth in this case was a malignant neuroma. Though careful dissection of the parts after removal from the body did not positively establish this view no other manifest origin was found. So although the growth might have begun in the surrounding connective tissues and spread to the brachial plexus it seems more reasonable to believe that the growth originated in connexion with the brachial plexus and then spread to the neighbouring parts.

The exostosis felt during life at the junction of the second left costal cartilage with the sternum was found not to be a true exostosis, but to be due to a forward projection of that chondro-sternal articulation. The second and third costal cartilages on the left side were intact though rather slender. The sternum was pushed forward by the intra-thoracic growth. The left lung was adherent to the intra-thoracic prolongation of the growth which surrounded its root and was invaded at one place by an outlying process of growth. The left bronchus was much narrowed. Both the lungs showed bronchitis. The heart (nine ounces) was normal. The abdominal organs were normal with the exception of some pigmentation of Peyer's patches in the ileum and of the solitary follicles in the colon. The adrenal bodies were quite healthy. On the abdominal sympathetic around the aorta there were a number of neuromata, but none was seen on the intestines or on the stomach. Dr. Lazarus-Barlow examined microscopic sections of these neuromata on the sympathetic and found that, like those on the spinal nerves, they were pseudo-neuromata.

Remarks.—It has been suggested by Feindel³ that Recklinghausen's disease is the outcome of a congenital malformation of the ectoderm. This hypothesis has quite recently been supported by Feindel and Oppenheim.⁴ The more probable theory seems to me to be that put forward 12 years ago by Dr. Payne⁵—viz., that the disease is due to a developmental vice on the part of the mesoblast in the corium and in the nerves. This accounts for the fibro-neuromata and the molluscous growths, while the pigmentation of the skin is allied to the common pigmented moles that occasionally develop into malignant melanotic growths. Recklinghausen's disease, therefore, may be regarded as depending on a congenital hyperplasia and tumour formation arising in the mesoblast at its junction with the epiblast—viz., in the skin and nerves. In this case it appears probable that as a result of progressive changes taking place the growth in the neck, at first an innocent neuroma, eventually became a sarcoma. This late development of sarcoma on congenital defects is, of course, well known to occur in pigmented moles of the skin. It also occurs in another defect of the mesoblast—viz., in multiple exostoses. An interesting example of this occurred in 1890 under the care of Mr. Warrington Haward who kindly allows me to refer to his case. A man, aged 33 years, had had exostoses all his life; one of them had pressed on the sciatic nerve and had given rise to pain for five years; the exostosis was therefore removed but a spindle-celled sarcoma developed on its site and eventually proved fatal. The patient and his son, aged seven years, also with exostoses, were shown at the Clinical Society of London by the late Mr. C. E. Cotes on Oct. 24th, 1890.⁶

Sarcomatous change in neuromata also occurs. Goldmann⁷ in his monograph gives a careful description of a malignant neuroma in a woman, aged 54 years, the subject of multiple neuromata, and Bowlby⁸ quotes examples of multiple neuromata associated with sarcomatous change. It is probable that many cases described as multiple neuromata show in addition some degree of molluscous growth and pigmentation, but that the more important clinical feature being connected with the neuromata the other changes attract little or no attention. In the present case the molluscous growth was a very striking feature and at first sight threw into the shade the small subcutaneous tumours and the pigmentation of the skin.

Other points of interest in the case are: 1. That the brunt of the tumour growth fell on the left side of the body, as shown by the molluscum fibrosum, the fibro-neuromata, and the tumour in the neck, while the pigmentation was bilateral. 2. The curious condition of pseudo-ptosis due to paralysis not of the levator palpebræ but of the unstriated muscular fibres in the upper eyelid, a condition which is the exact converse of Stellwag's symptom in exophthalmic goitre.

Harley-street, W.

INTERSCAPULO-THORACIC AMPUTATION.

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My personal experience of this operation is limited to two cases, in both of which there was little or no evidence of shock, with rapid recovery, the patient being out of bed with the wound healed *per primam* in the second week. Elaborate details of the incision and general technique of the operation were given by Paul Berger in a monograph on the subject in 1887 and these may also be found in the textbooks; but in neither of my cases, nor in the other operations of this kind which I have seen performed, were the

³ Thèse, Paris, No. 104, 1896.

⁴ Archives Générales de Médecine, July, 1898, p. 77; Sur les Formes Incomplètes de la Neuro-fibromatose.

⁵ Transactions of the Pathological Society of London, vol. xxxviii., p. 69.

⁶ Transactions of the Clinical Society of London, vol. xxiv., p. 228.

⁷ Beiträge zur klinischen Chirurgie, Band x., Heft i., redigiert von P. Bruns, 1892.

⁸ Injuries and Diseases of Nerves, 1889, p. 496.