MULTIPLE HEREDITARY DEVELOPMENTAL ANGIOMATA (TELANGIECTASES) OF THE SKIN AND MUCOUS MEMBRANES ASSOCIATED WITH RECURRING HÆMORRHAGES.

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Mrs. Sarah S., of Jewish origin, is a pale, somewhat fat woman of medium height, aged 60 years. She has a number of bright red angiomata distributed over the face, ears, lips, tongue, mucous membrane of the mouth, and the conjunctival surfaces of the four eyelids. All these angiomata are small; many, including those on the tongue and inside of the mouth, are hardly as large as an ordinary brass pin's head (punctiform angiomata); the largest ones are those on the face, the biggest of which, that on the right cheek, measures five by seven millimetres in diameter. They are all of them simple capillary angiomata (telangiectases) and hardly, if at all, raised above the general surface, with the exception of some of the larger ones (on the face), which project slightly and approach either the "spider nævus" (nævus araneus) class or a kind of small superficial cavernous nævus. On the face there are likewise a few of the common hair-like telangiectases. On the mucous membrane inside both nostrils there are some telangiectases to be seen, but on the posterior wall of the pharynx and by laryngoscopic examination of the glottis I could see nothing abnormal excepting some telangiectases on the anterior surface of the epiglottis. On the fingers, and notably under the finger-nails, there are several minute (pinpoint) red angiomata, which I had not observed until Professor W. Osler, when he recently saw the patient, kindly drew my attention to them. I have searched most of the rest of the body for angiomata, but have failed to find any excepting those already mentioned. She has varicose veins and slight œdema of the legs.

Both the patient's appearance and the examination of her blood (April 11th, 1907) show that she is anæmic; the red cells number 2,833,000 and the white cells 11,350 in the cubic millimetre of blood; the hæmoglobin-value (by Haldane's method) is 45 per cent. Dr. A. E. Boycott has very kindly examined blood films (of May 22nd, 1907) and reports that the red cells are pale and many of them undersized, but are not otherwise abnormal. He finds that there is no evidence whatever of myeloid reaction. His differential count of 500 white cells gives the following result: neutrophiles (polymorphonuclears), 42·0 per cent.; lymphocytes, 45·2 per cent.; intermediates, 7·2 per cent.; large hyalines, 2·4 per cent.; eosinophiles, 2·4 per cent.; and mast cells, 0·8 per cent.

I thought the coagulation time of the patient's blood was slightly increased as compared to that for an ordinary healthy person, but on using Sir A. E. Wright's coagulation tubes ("coagulometer") I found (May 26th, 1907) that the coagulation time was only about five and a half minutes, so that the coagulability is within the limits for normal individuals.

On auscultation of the heart a systolic murmur of uncertain nature can be heard. With this possible exception the thoracic and abdominal viscera appear healthy. The urine is free from sugar and albumin and normal in quantity. The brachial blood pressure by the Riva-Rocci apparatus (using the broad band) is about 135 millimetres of mercury. Dr. C. Markus by ophthalmoscopic examination of the right eye found a small central patch of chorio-retinitis.

The patient has been married twice, and by her two marriages has had nine children, eight of whom are living. I shall return later to the condition of these children. The menopause occurred at the early age of 38 years. At about 48 years of age she suffered from sore places of some kind on the left leg.

The biggest of the angiomata (that on her right cheek) was first observed when she was about 42 years of age. Previously to that none had been noticed; the others, she thinks, have developed at various times since then. The nose-bleeding commenced a few years before the appearance of the spots on her face. During the last six years or so she has suffered from recurrent attacks of epistaxis, an attack occurring on the average about every two or three weeks (that is, not including the occasions on which just a little blood can be seen on the handkerchief). During the last two years, and especially recently, the bleeding has been worse than it was before. There seems never to have been a tendency to any other abnormal hæmorrhages.

The patient's mother, who died at the age of 56 years, was certainly subject to attacks of epistaxis and had one or two "spots" on her face similar to those on the patient's. Three sons and one daughter of the patient suffer likewise from attacks of epistaxis and in two of the sons I have ascertained the presence of multiple angiomata of the skin and mucous membranes. In these members of her family, as in the patient herself, there is no history of any abnormal hæmorrhages except from the nose.

Of Mrs. S. S.'s nine children one only, Joseph L., springs from her first marriage. He is now about 42 years of age and is said to be healthy and not to suffer from epistaxis or angiomata. I have not seen him. He is married and has eight children, of whom the eldest, a girl, aged 19 years, has had epistaxis.

Lewis S., her second child (the first by her second marriage), is now about 37 years of age and looks a well-built man, not especially anæmic. He says that he has enjoyed fairly good health with the exception of occasional attacks of nose-bleeding, which first commenced when he was about eight years old. On examining him I found that there were two small "spider nævi" on his left cheek and a few scattered red punctiform angiomata on his neck and the mucous membrane of his gums, lips, and tongue. I could also make out a telangiectatic condition of the mucous membrane of the right side of the nose—that is, the side from which he says the bleeding always or generally occurs. He is married and has one child, a girl, aged 11 years, who has had epistaxis.

Albert S., the third child, is now aged 35 years. He is a strong-looking, not anæmic, man who has served as a soldier in India and has enjoyed good health with the exception of slight malaria contracted in India and the occasional attacks of nose-bleeding to which he has been subject since about the age of ten years. On examining him I found three small angiomata of the "spider nævus" class on his left cheek and several small angiomata situated on the conjunctival surface of the eyelids and the mucous membrane of the gums and lips, varying in size, but not exceeding about three millimetres in diameter. Some of the larger (doubtless older) of these angiomata are more bluish than the smaller. I likewise noted some ordinary hair-like telangiectases of the skin of both cheeks. Rhinoscopic examination showed a decidedly telangiectatic condition of the mucous membrane lining both nostrils. He is married and has one child, a boy four years old, who has not yet suffered from epistaxis.

Harriet R., the fourth child, is now aged about 33 years, and is said to enjoy good health and not to have any of the "spots" on her skin or to suffer from nose-bleeding. She is married and has one child, a girl aged nine years, who has not yet shown any tendency to epistaxis.

Jacob S., the fifth child, now about 31 years of age, is likewise said not to be subject to nose-bleeding or to have any of the "spots." I have not seen him.

Annie S., the sixth child, aged 28 years, suffered from disease (probably tuberculous) of the left knee-joint during childhood but otherwise has enjoyed fair health, though during the last three years or so she has had occasional slight attacks of epistaxis. She herself has not noticed any "spots" on her skin and on examining her face and hands and the mucous membranes of her mouth and nose I could find no angiomata.

Rachel, the seventh child, aged about 26 years, is said not to suffer from nose-bleeding or to have any of the "spots." I have not seen her.

Marcus, the eighth child, is said to have died at the age of three years from brain-trouble following a fall.

Manning S., the ninth child, aged 23 years, has been subject to occasional nose-bleeding since he was 18 years old. Otherwise he has had good health and now looks healthy. When I examined him recently I could find no telangiectases or angiomata on the skin of the face and hands or on the mucous membrane of the mouth and nose, but on the front of the chest I noted a few minute red nævi, such as very many or most persons have one or two of on various parts of their body.

Mrs. S. S. was treated for a time with arsenic and afterwards with an iron preparation which she thinks has done her good. She is frightened, she says, of having the

hæmorrhages from her nose altogether stopped, but although her blood coagulability is not abnormally low I am trying treatment with calcium lactate, having heard from Professor Osler of a good result apparently obtained by this method in one case. Calcium lactate (in daily doses of 15 grains) has now (July 13th) been taken for some weeks, but as yet apparently without any benefit. The patient has likewise been given a very dilute adrenalin solution to apply locally with a camel's hair brush whenever the epistaxis commences. This also has in the patient's hands apparently failed to arrest the hæmorrhage.

Remarks on the literature of the subject.—The first account of this malady in which several family groups of sufferers are referred to is that published in 1901 by Professor Osler entitled "A Family Form of Recurring Epistaxis associated with Multiple Telangiectases of the Skin and Mucous Membranes."[1] In it he has included three cases of his own. Recently Dr. Brown Kelly has reviewed the whole subject in his paper on Multiple Telangiectases of the Skin and Mucous Membranes of the Nose and Mouth,[2] which is accompanied by excellent coloured illustrations. I have arranged the following data from the published accounts under the various cases or families.

Family 1.—B. G. Babington[3] in 1865 published notes of a family showing a most remarkable tendency to epistaxis, but he makes no mention of the presence of angiomata or telangiectases of the skin or mucous membranes in any members of this family. There is therefore no certain evidence that his cases belong to the class under consideration.

Family 2.—J. Wickham Legg in 1876 read a paper before the Royal Medical and Chirurgical Society of London on a Case of Hæmophilia complicated with Multiple Nævi.[4] The patient, a musician, aged 65 years, had been subject to epistaxis from boyhood and had also shown a tendency to bleed from traumatic causes. He had numerous small nævi over the face, forehead, and various parts of the trunk which had been first noticed when he was about 41 years of age. Several members of his family suffered from epistaxis.

Family 3.—O. Chiari[5] in 1887 described the cases of two sisters, many of whose relations suffered from recurrent and severe epistaxis. He at first thought the spots on the mucous membrane in these two cases were telangiectases, but after observing two other cases he came to the conclusion that in all four cases he had to deal with blood extravasations undergoing a slow change, such as might be met with in hæmophilic patients. Brown Kelly shows reason for doubting the presence of hæmophilia in Chiari's, as also in Wickham Legg's cases.

Family 4.—Rendu[6] at the Société Medicale des Hôpitaux de Paris in 1896 brought forward the case of a man, aged 52 years, suffering from repeated epistaxis. He had small superficial angiomata of the skin of the face, neck, and thorax, and of the mucous membrane of the mouth. Angiomata were not actually seen in the nasal passages, but it was presumed that some were present there also and accounted for the frequent attacks of epistaxis. There was a history of repeated epistaxis in the patient's mother and in one brother. The latter, however, had suffered from albuminuria. The patient's father had died from "dysentery" and had been subject to melæna.

Family 5.—The first two cases described by Osler[7] were those of brothers, aged 57 and 55 years respectively. There was a history of repeated epistaxis in their father, two sisters, the child of one of the patients, and a grandniece. The face of Osler's elder patient "presented a very unusual appearance owing to the large number of dilated venules and capillary and venous telangiectases." There were numerous angiomata on the mucous membrane of the nasal septum and on the tongue. The younger of the two patients had likewise cancer of the stomach and died in the hospital. Sections of the septum of the nose from the latter case "showed many large dilated veins just beneath the epithelium."

Family 6.—Osler's third case[8] was that of a man, aged 49 years, who had been subject from childhood to attacks of epistaxis which recurred at short intervals. Except for the epistaxis he had been a healthy man. There were multiple telangiectases of the skin and of the mucous membranes of the nose and mouth. In this case cauterisation of the angiomata on the nasal septum seemed at one time to do good. So far as the patient himself knew "there were no bleeders in his family and none of the members had had serious attacks of epistaxis."

Family 7.—C. O. Hawthorne[9] has recently published a short note of the case of a woman, aged 49 years, with bright red telangiectases scattered over both cheeks and with a few on the fingers of the right hand. She had been subject to repeated epistaxis from childhood. Her father and a sister were troubled in the same way and both of them, as well as her eldest daughter, had "spots" like those on her own face. All her nine children had had more or less numerous attacks of epistaxis.

Family 8.—Brown Kelly's patients[10] were two sisters, aged 41 and 40 years respectively. Their father had been subject to attacks of epistaxis and vomiting, and he had "spots" on his face similar to those on their own faces; he was said to have died at the age of 62 years in consequence of frequent bleedings from the nose. The elder of the two patients died suddenly from syncope induced by severe epistaxis. A daughter (aged 23 years) of the elder patient had lately also developed telangiectases on her face. In Brown Kelly's patients, as in some of the other recorded cases, it does not appear that the telangiectases were present in early life. In fact, the obvious angiomata on the skin in these cases, as Brown Kelly points out, are not usually observed until the patient is approaching middle life, and "the most striking development of the telangiectases seems to take place from the age of 35 or 40 onwards."

Dr. W. Bligh[11] says that a man, aged 32 years, required his assistance on account of bleeding from the left forearm. On examination the source of the hæmorrhage was found to be a minute nævoid growth, from which arterial blood was spurting freely. Similar nævoid growths were present on the back of the left wrist, on the side of the neck, and on the forehead. The growths varied slightly in size, but the largest was no bigger than the head of a "carpet pin." He had been previously troubled with violent bleeding from more than one of the little tumours. Dr. Bligh destroyed all the little growths by cauterising them, but a few months later bleeding occurred from a fresh tumour on the face; two other fresh ones had likewise appeared upon the forehead. These were all destroyed in the same way. The patient was healthy and not hæmophilic. Dr. Bligh has kindly informed me that there is no history of angiomata in the family of his patient, but nevertheless I mention the case here as it is well known that usually "familial" diseases do occasionally present themselves in an apparently isolated fashion.

Though Dr. Bligh's case may not belong to the group under consideration it is a remarkable example of the kindred condition of multiple late developmental angiomata. An equally striking example of the latter condition is furnished by a case shown by Dr. F. J. Smith at the Medical Society of London on April 4th, 1898.[12] Dr. Smith's patient was a woman, 50 years of age, with multiple venous angiomata chiefly on the face and upper part of the trunk, varying in size from that of a pea to that of a small bean. In addition she had varicose veins of both lower extremities and both labia majora. Nine months previously, according to the patient's own account, she was perfectly well and had no varicose veins even in the legs. About that time she noticed a small painful spot on the right side of the nose, which came up like a small red pimple; soon after this some other red spots appeared on the back and shoulder and these spots later became bluish. In the case of a middle-aged man shown at the Dermatological Society of London on Dec. 12th, 1900,[13] with multiple bluish nodular angiomata on various parts of the body and face, there was a decided family history of multiple venous angiomata. Some of his little tumours dated from birth, others had

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[1] Johns Hopkins Hospital Bulletin, November, 1901, p. 333.
[2] Glasgow Medical Journal, June, 1906, vol. lxv., pp. 411–422.
[3] THE LANCET, Sept. 23rd, 1865, p. 362. I have not looked up other literature on "hereditary epistaxis."
[4] This paper was apparently omitted from the Transactions of the Royal Medical and Chirurgical Society, but a lengthy summary appeared in THE LANCET, Dec. 16th, 1876, p. 856.
[5] Erfahrungen auf dem Gebiete der Hals-und Nasenkrankheiten, Vienna, 1887, p. 60. I have only been able to refer to Brown Kelly's (loc. cit.) references to Chiari's cases and have not seen Chiari's original paper.
[6] Gazette des Hôpitaux, Paris, Nov. 24th 1896 1322.
[7] Loc. cit.
[8] Loc. cit.
[9] THE LANCET, Jan. 13th, 1906, p. 90.
[10] Loc. cit.
[11] Note on a Case of Bleeding Telangiectases, THE LANCET, Feb. 23rd, 1907, p. 506.
[12] Transactions of the Medical Society of London, vol. xxi., p. 358.
[13] I cannot find the account o thi case in British Journal of Dermatology.

only recently appeared. The occurrence in elderly persons of small multiple cavernous angiomata of the lips and mouth is probably not very rare.

The conclusions at which I arrive are the following. 1. That the disease (or morbid syndroma) which I am more especially considering in the present article affects and is transmitted by both sexes. 2. That the hæmorrhage in most cases is only from the nasal mucous membranes. 3. That in most cases the morbid syndroma is not connected with any hæmophilic tendency or any diminution of blood coagulability. 4. That the cutaneous angiomata are generally not congenital but that they are "late developmental" and usually first attract attention towards middle life. Some of the minute red spots of the capillary angioma class tend to develop into raised bluish nodules approaching the cavernous venous angioma in character. 5. That in most cases a tendency to nose-bleeding has been present from early life, or at all events many years before any cutaneous angiomata have been observed. 6. That with advancing years both the attacks of hæmorrhage and the anæmia usually become more severe. 7. That probably a kind of "vicious circle" is established, the repeated attacks of bleeding giving rise to a grave condition of anæmia, which in its turn increases the tendency to hæmorrhage. This consideration would furnish a reason for occasionally employing iron and arsenic as part of the treatment. 8. That the hereditary nature of bleeding telangiectases of the nasal mucous membrane may be compared to the occasional family tendency to suffer from hæmorrhoids, or to have ordinary varicose veins of the lower extremities, or multiple smaller varices and multiple hair-like cutaneous telangiectases on various parts of the body.[14] Hæmorrhoids in some cases may, in fact, be termed "hereditary developmental bleeding angiectases."

The study of families affected with multiple angiomata of the skin and mucous membranes might help to throw light on the general subject of pathological inheritance in man. In these cases it is the morbid tendency only which is inherited, just as it is in cases of hereditary tendency to pulmonary tuberculosis, but the study of the latter hereditary tendency is complicated and obscured by the fact that the advent of a microbe (the tubercle bacillus) is necessary for the fulfilment of the "morbid promise" or potentiality in question.

Harley-street, W.

[14] Cf. F. Parkes Weber: A Note on Cutaneous Telangiectases and their Etiology: Comparison with the Etiology of Hæmorrhoids and Ordinary Varicose Veins, Edinburgh Medical Journal, April, 1904, p. 346. In regard to the view that varicose veins are venous overgrowths, allied to venous angiomata, see A. Pearce Gould's Lettsomian Lectures in Transactions of the Medical Society of London, 1902, vol. xxv., p. 132; also W. Thorburn's remarks on Developmental Varix, Brit. Med. Jour., Nov. 17th, 1900, p. 1421; and Sir W. H. Bennett's remarks on Congenital Varix, THE LANCET, Nov. 22nd, 1902, p. 1374. For similar views regarding the etiology of hæmorrhoids see G. Reinbach, "Path.-anat. und Klinische Beitrage zur Lehre von den Hämorrhoïden," Beiträge zur Klinischen Chirurgie, Tübingen, 1897, vol. xix., p. 1; and "Hämorrhoïden im Kindesalter," Mittheilungen aus dem Grenzgebieten der Medizin und Chirurgie, Jena, 1903, vol. xii., p. 272.

Clinical Notes:

MEDICAL, SURGICAL, OBSTETRICAL, AND THERAPEUTICAL.

A CASE OF PARTIAL PERFORATION OF THE BOWEL SIMULATING APPENDICITIS; OPERATION; RECOVERY.

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THE patient, a coal hewer, aged 48 years, was admitted to the Sunderland Infirmary under my care on April 21st, 1907, complaining of a very severe pain in the right inguinal region and shooting across the abdomen. The history of the case was as follows. Whilst working in the Ryhope pit about 8 P.M. on April 18th he strained himself by slipping his foot when pushing a tub. He felt as if "something had torn in his bowels." There was no acute pain felt then nor for some hours afterwards. He continued to work until midnight—the end of the shift—with only momentary interruptions. The only other symptom which caused him any trouble was an urgent desire to micturate. He was able to pass a pint of clear urine and also had an action of the bowels. After finishing his work he was conveyed home to Ryhope by ambulance car and was seen by his medical attendant very soon afterwards. On examination there was no localised tenderness in the abdomen nor did palpation or percussion reveal anything abnormal. There was dorsal decubitus with both legs drawn up and abdominal facies. The temperature was 99° F. and the pulse was 76. Twelve hours later pain began in the hypogastrium and gradually became diffused over the abdomen. Tenderness on deep pressure was present over the appendix area. The temperature was 102° and the pulse was 80. There was no sickness. The subsequent symptoms changed but little up to the evening of the 21st (the date of his admission), the temperature fluctuating between 101° and 103° and the pulse always being below 80, with a gradual diminution in tone and volume. There was no history of any previous attack.

On admission to the infirmary the patient was in a rather collapsed condition (having driven three miles) and in great pain. There were great tenderness, rigidity, and some fulness over the right iliac fossa. He had a dry tongue and a feeling of nausea but there was no vomiting. The temperature was 101° and the pulse was 104. The symptoms pointed to appendix abscess. His abdomen was opened over the appendix area within an hour of admission. Recent inflammatory adhesions were found in the neighbourhood of the appendix. A small tear through the serous coat of the anterior part of the cæcum was found situated about two inches from the root of the appendix. The inner coats were bulging through. A purse-string suture was applied to the rent and the appendix, which was healthy, was removed for safety. For the first three days after the operation the condition of the patient was one of collapse, giving rise to some anxiety, but thereafter recovery was uninterrupted, and he was discharged quite well on May 23rd.

I have not been able to find the record of a similar case. The interesting points seem to be: an apparently simple slip causing a comparatively severe injury, the man being able to work for three or four hours after the injury, and the simulation of appendix inflammation.

I am indebted to Dr. Alfred Rutter of Ryhope and to Mr. Lionel H. Booth, my house surgeon, for the notes of the case.

Sunderland.

INJURY TO SHOULDER; HERPES ZOSTER; INFLUENZA.

BY JOHN ALLAN, M.B., CH.B. EDIN.

THE following case presents several points of interest and the sequence of events is unusual. A youth, 18 years of age, was seen on March 30th, 1907, on account of injury to his left shoulder. It appeared that when at work he fell and struck the back of his left shoulder against a block of wood. On examination no bruising was seen and no fracture or dislocation could be detected. Lead and opium lotion was applied to the part. During the next day or so he complained of pain at the back of the left shoulder and arm, but nothing definite could be made out to account for it. On April 3rd there appeared a herpetic eruption on the posterior aspect of the left side of the chest and on the left arm and forearm. This was accompanied by a good deal of pain. The temperature was slightly raised and the patient felt out of sorts generally. He was put to bed and the eruption was dusted with a simple dusting powder. He was given salicylate of sodium (15 grains) thrice daily and fresh vesicles as they appeared were painted with collodion. He progressed favourably until April 8th. On that date his temperature rose to 102·6° F., he was sick and vomited frequently, and he complained of headache and sore throat. On the following day his temperature was 103·8° and a scarlatiniform rash appeared over his body. This was most marked over the lumbar region and the buttocks. The face and neck and the extremities were not affected by the rash. The throat was very much congested and he had great difficulty in swallowing even fluids. He had pains throughout the body generally, especially in the back. Five grains of salicylate of quinine were given every four hours instead of the salicylate of sodium and a potassium chlorate gargle was prescribed. The temperature next morning was 103·2°, and thereafter it gradually fell to normal. The rash disappeared