Section of Neurology.

[*December* 13, 1928.]

A Note on the Association of Extensive Hæmangiomatous Nævus of the Skin with Cerebral (Meningeal) Hæmangioma, especially Cases of Facial Vascular Nævus with Contralateral Hemiplegia.

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Abstract.—The main type of the cases under consideration is that in which extensive capillary nævus of the skin, especially of trigeminal distribution, is associated with contralateral spastic hemiplegia, developing in early life, probably sometimes connected with intracranial hæmorrhage from the abnormal blood-vessels of a diffuse leptomeningeal angioma on the same side as the main vascular nævus of the face. Various points related to such cases are referred to.

According to Harvey Cushing (1928) blood-vessel tumours contribute only about two per cent. of all intracranial tumours. In many cases in which the existence of a hæmangiomatous condition of the cerebral meninges has been proved by operation or post-mortem examination there has been no large hæmangiomatous nævus of the skin to suggest the nature of the cerebral disease. On the other hand, in the patients in whom epilepsy, especially of the Jacksonian type (irritative lesion), or spastic hemiplegia (paralytic lesion), or both, developed in early life the hæmangiomatous nature of the intracranial disease has been sometimes suggested by the presence of extensive hæmangiomatous nævus of the skin, especially of the face (more or less trigeminal distribution) on the same side as the intracranial irritative or paralytic lesion.[1]

Such cases have been recorded by S. Kalischer, Lannois and Bernoud (two cases), Strominger, Harvey Cushing (three cases), E. A. Cockayne, Mouriquand and Bernheim, C. P. Symonds, Brushfield and Wyatt (four cases), H. J. Hugo, and T. R. Aynsley (two cases). In D. M. Greig's case (to which I shall return) instead of unilateral or asymmetrical capillary nævus there was bilateral symmetrical multiple adenoma sebaceum of the face, of Pringle's telangiectatic type. In some of these cases the nature of the intracranial disease was elucidated by a subsequent post-mortem examination or by operation.

Cushing's first case was that of a boy, aged 4, with extensive capillary nævus of the face (especially of the right side) and unilateral right-sided buphthalmus. He had developed left-sided spastic hemiplegia after what Cushing supposed to have been an attack of right-sided intracranial hæmorrhage at 2 years of age. He died from some severe intercurrent infection and the necropsy showed that the right cerebral hemisphere was smaller than the left one, and the meninges covering it were unduly vascular, "in all probability representing a nævoid condition similar to that present on the skin" (Cushing).

Cushing's second case was a boy, aged 5 years, with extensive capillary nævus of the upper right part of the face and head and unilateral right-sided buphthalmus. There were epileptiform convulsions after an attack of what was supposed to have been intracranial hæmorrhage. Improvement followed right-sided craniotomy after ligation of the external carotid artery. A very vascular condition of the dura mater was found at the operation with adhesions to the leptomeninges.

Cushing's third case was a left-handed boy, aged 8 years, with extensive capillary nævus, involving especially the right side of the face and right upper extremity; there was unilateral right-sided buphthalmus. Left-sided spastic hemiplegia, epilepsy and idiocy followed what was supposed to have been an attack of right-sided intracranial hæmorrhage at 9 months of age. No operation was attempted and the nature of the intracranial lesion could only be presumed.

1 In two or three cases the facial nævus has been partly pigmentary as well as hæmangiomatous.

In Strominger's case, as quoted by Cushing, there was extensive left-sided facial capillary nævus, with spastic hemiplegia (presumably of the right side). Death occurred from some intercurrent affection and at the necropsy the cerebral meninges (presumably on the left side) were found thickened and excessively vascular.

In Kalischer's case the patient died at 1½ years from bronchopneumonia after whooping-cough. There was an extensive capillary nævus of the left half of the face. Convulsions commencing on the right side had been observed since the age of 6 months, and for the last quarter year of its life the child had had right hemiplegia. The necropsy showed that the left cerebral hemisphere was smaller than the right and was covered by a diffuse meningeal angioma. The dura mater was very adherent to the skull, and the left frontal bone was greatly thickened and very adherent to the scalp (the side of the capillary nævus). Up to the age of 6 months the child had seemed normal in regard to the nervous system. But it does not seem certain that the subsequent epileptic attacks (of the Jacksonian type) and the ultimate right spastic hemiplegia were due to intracranial hæmorrhage from the meningeal angioma on the left side.

A left-sided meningeal angioma was found at the necropsy of one of Brushfield and Wyatt's cases (Case 3), and there were other cerebral defects noted. Congenital spastic hemiplegia had been present on the right side.

In Hugo's case there were angio-endotheliomata found in the brain, kidneys and liver, but apparently there was no typical meningeal angioma.

In Steinheil's case, quoted by Kalischer and others, the patient was a man, aged 49, who had no typical vascular nævus of the skin, but, since childhood, there had been aneurysm-like enlargements and ecstasies of the right frontal, temporal and external carotid arteries. He had had epileptic attacks for years before his death, and at the necropsy some of the arteries of the brain were found enlarged and there was a racemose arterial angioma of the front third of the right frontal lobe. In this case, though typical capillary nævus of the skin was apparently absent, there were developmental extra-cranial arterial abnormalities on the same side as the racemose arterial angioma of the brain. The case possesses a further special point of interest, in so far as it tends to show that meningeal angioma may be associated in the same patient with another kind of congenital or developmental arterial abnormality in the brain or other parts of the body, namely, "weak spots" in the arterial walls, an occasional cause of developmental or so-called congenital (sometimes actually congenital) aneurysms in otherwise healthy young individuals.

In Greig's case with the multiple adenoma sebaceum, the patient was a man, aged 18, the subject of convulsions for fifteen years, and of right hemiplegia for eleven years. The convulsions were of the type of Jacksonian epilepsy, commencing in the right hand, and operation showed a meningeal angioma covering the left Rolandic area. The sebaceous adenomata of the face might well have led one to suppose that the cerebral disease was of the nature of tuberous sclerosis, but it would have had to be *unilateral* tuberous sclerosis.[2]

I described the following case in 1922, when I wrote: "It is highly probable that the congenital cerebral disease is in some way connected with the presence of a vascular nævus of the meninges or brain on the left side—of the same nature as the extensive vascular nævus of the patient's body." But I have since (summer, 1928) obtained better skiagrams of the patient's head and, as a result, it may now be stated with practical certainty that the disease of the left side of the brain is of the nature of a diffuse meningeal angioma, comparable with the condition which was discovered post-mortem on the left side of the brain in Kalischer's case. This case of mine is, I think, the only one as yet described in which so great a difference between the two sides of the brain has been demonstrated by X-ray examination

2 Incidentally, Drs. G. Brown and T. W. Sheldon have shown me that partially calcified foci of tuberous sclerosis in the brain may give radiographic shadows.

during life, without the aid of injection of air, etc.[3] The radiograms, which were poorly reproduced in my paper of 1922, were the first reported of the kind. Similar unilateral cerebral changes, but not quite so marked or extensive, have been demonstrated by radiograms in V. Dimitri's case (1923), in A. M. Marque's two cases (1927), in No. 2 of Brushfield and Wyatt's cases, in T. W. Sheldon's case, and in certain, as yet, unpublished cases (including cases of L. B. Rawling and C. P. Symonds).[4]

The patient, R. R., a woman, now aged 28, is of a Hebrew family. Her mother is active-looking, well-developed, and has a moderate degree of xanthelasma

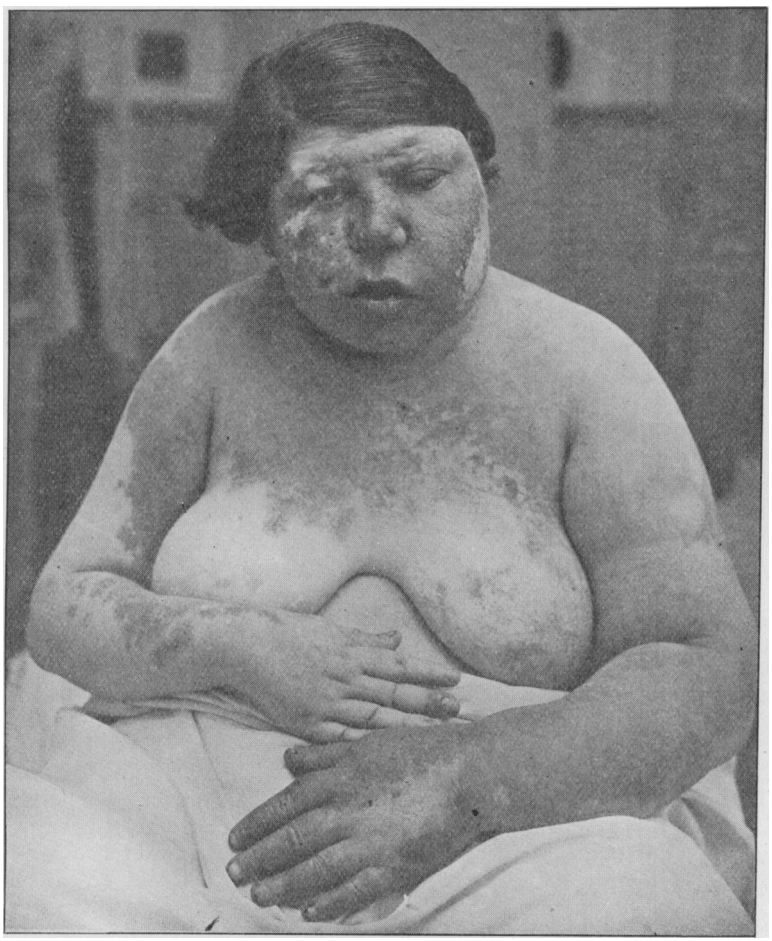

FIG. 1.

palpebrarum. A sister of the patient is said to have become insane "after air-raids." The mother says she has also two healthy normal children; none have died or were stillborn; she had two or three miscarriages. The patient has very widespread capillary nævus, chiefly of the superficial "port-wine stain" type (fig. 1). On the back of the trunk this port-wine angioma is almost entirely limited by the median line to the left side; in front the distribution, though very irregular, is more extensive on the left than on the right side. In parts, notably in the left cheek,

3 The radiograms have been reproduced in figs. 2 and 3 without any touching up.

4 *Cf.* H. Cushing and P. Bailey, "Tumours Arising from the Blood-vessels of the Brain," 1928, p. 26

there is, besides the "port-wine staining," a condition of more deeply seated thickening, though possibly also of angiomatous nature. She has right-sided spastic hemiplegia, apparently of congenital or almost congenital origin. The right limbs are smaller and notably shorter than the left limbs; in fact, there is a condition of unilateral right-sided hypotrophy, as one would expect, associated with the right-sided spastic hemiplegia.

The mammary regions are bulky, doubtless owing to her general obesity. But in 1922 I wrote: there is absence of pubic and axillary hair, and she has never menstruated; the sexual organs (rectal examination by Dr. Scheu) are probably infantile.

The left eyeball (1922) was larger than the right owing to congenital glaucoma or buphthalmus. Dr. C. Markus reported that the right eye was normal, but that there was atrophy and glaucomatous excavation of the left optic disc. She was blind in the left eye. There was so-called heterochromia iridis, the upper three-quarters of the left iris being brown, the lower quarter-sector of the left iris and the whole of the right iris being grey-blue. In August, 1926, Dr. Markus had to excise the left (buphthalmic and blind) eye on account of purulent corneal ulceration and severe pain. By macroscopic and microscopic examination of the excised eye there was no evidence of any retinal or other angioma-formation in it.

The patient is certainly not an idiot, although there may be some mental deficiency. She can speak well and in conversation seems not to be deficient in understanding simple matters and in ordinary emotions; she has never been to school, has not learned to read and write, and on account of the great paresis in her right upper extremity, can do only a little housework. There is much less paresis in the right lower limb than in the upper one. The knee-jerks are very active on both sides. The plantar reflex (1922) on the left side is of the normal flexor type; that on the right side is of the extensor type.

Whilst the patient was under observation in hospital in 1922 there were no convulsions, nor was there apparently a history of any kind of fits. Her pulse was 68 to 88, and her respiration was 24 per minute. Her brachial systolic blood-pressure was 115 mm. Hg. The blood-serum gave a completely negative Wassermann reaction. The blood-count showed slight excess of white corpuscles. The urine (quantity was about normal), when tested, was of specific gravity 1025; acid; free from sugar; containing a trace of albumin (probably due to presence of some vaginal discharge). No alimentary glycosuria followed the ingestion of 100 grm. dextrose; but the urine contained a trace of sugar after a meal of 200 grm. dextrose.

An X-ray examination of the head was made with Dr. James Metcalfe's help in 1922. The skull showed marked prognathism of the "anthropoid" or "simian" type; that is to say that the prognathism involved a projection of both jaws beyond the vertical line of the ideal orthognathous skull—a character of the lower races of mankind and anthropoid apes, not to be confused with the so-called "inferior prognathism" which is sometimes a family characteristic. The pituitary fossa was extremely small or infantile, a feature that might be considered in connexion with the causation of the patient's obesity and defective sexual development (see above). The left cerebral hemisphere appeared sclerosed; at all events, it was more opaque and gave a somewhat deeper shadow than the right hemisphere. It seemed to occupy only about two-thirds of the left half of the cranial cavity and to be surrounded by cerebro-spinal fluid (external hydrocephalus). But skiagrams of the skull, taken in June, 1928 (figs. 2 and 3), whilst confirming the findings of 1922 in regard to the prognathism and size of the pituitary fossa, have been much more successful in demonstrating the abnormal condition of the left side of the brain. The whole of the left side, excluding the cerebellum, appears to be small, occupying only part of the cranial cavity, as if sclerosed or bound down by thickened or partially calcified leptomeninges. The skiagrams strongly suggest that the left

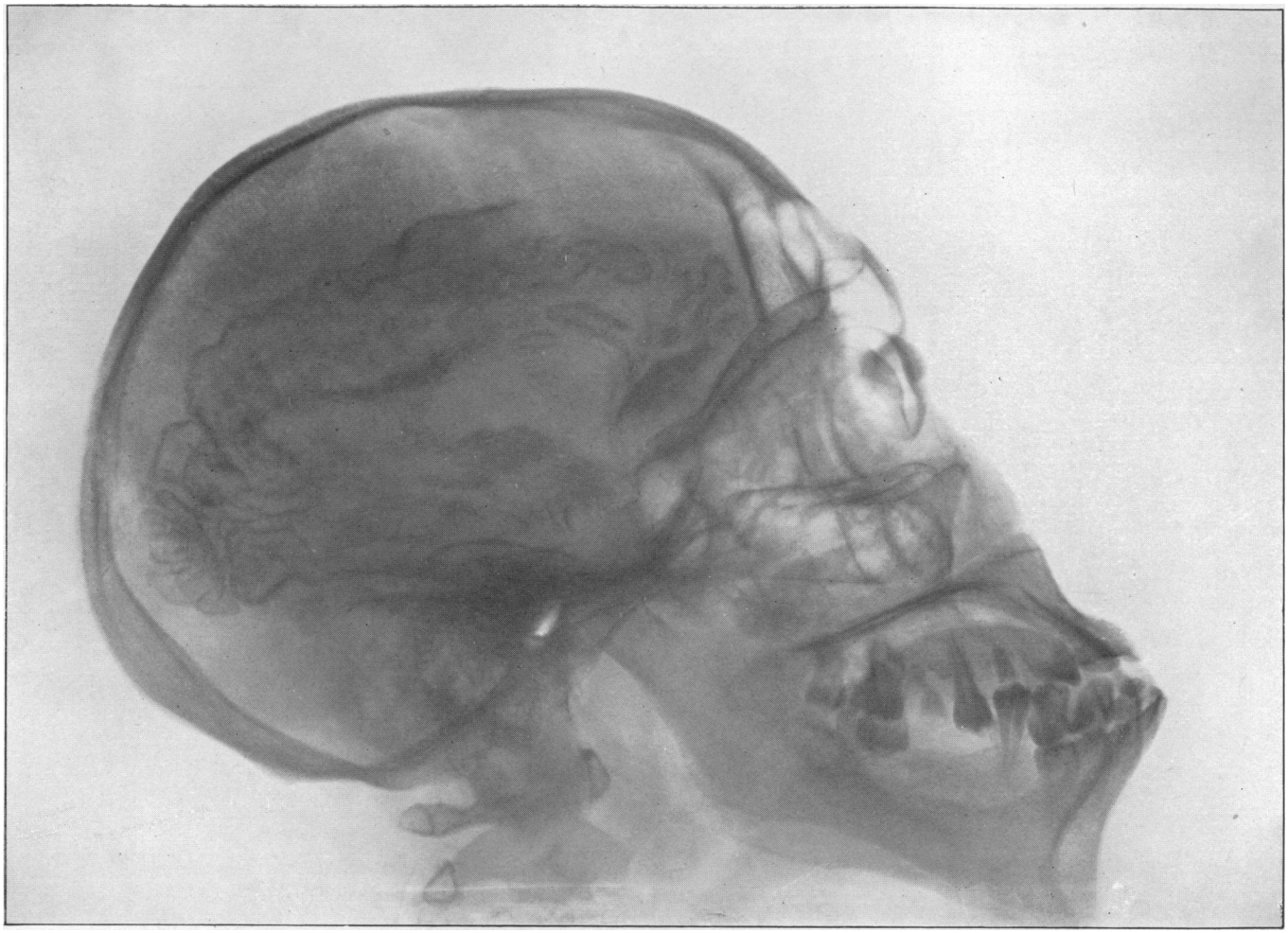

FIG. 2.—Dr. Weber's Case: Lateral Skiagram, to show the intracranial calcification (description in the text).

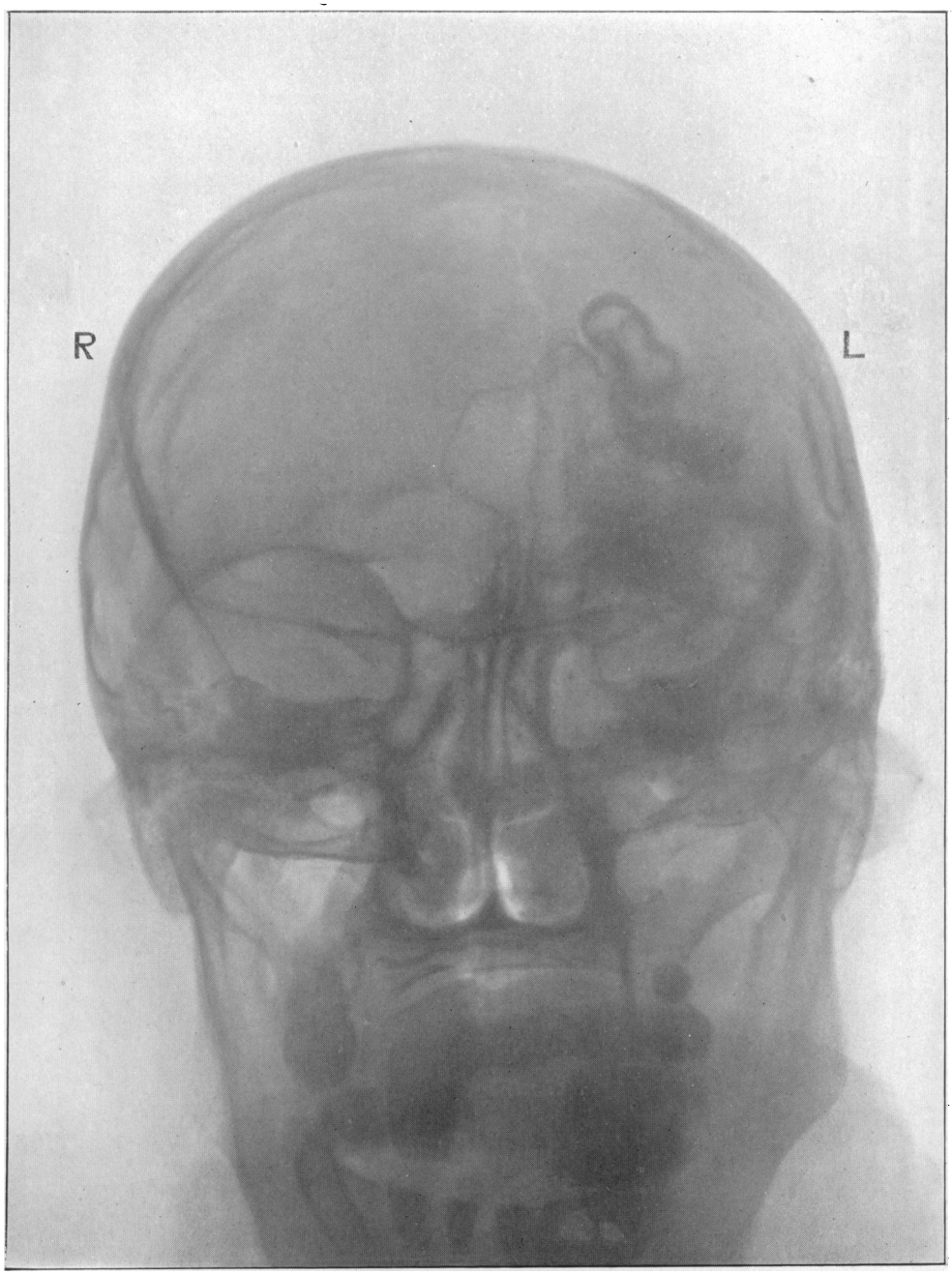

FIG. 3.—Dr. Weber's Case: Antero-posterior Skiagram (description in the text).

cerebral hemisphere is in a condition analogous to that of the left hemisphere discovered on post-mortem examination in Kalischer's case; i.e., smaller than the right hemisphere and bound down by abnormal leptomeninges, the site of diffuse angiectatic hæmangioma. In the present case there is probably some calcareous deposit in the vessels of the meningeal angioma, as there was in Nielsen's recent case of racemose arterial angioma of the inferior cerebellar artery in a man aged 43. The shadowing, apparently due to calcareous deposit, does not extend beyond the tentorium cerebelli, and there is no radiographic sign that the cerebellum is affected. The calcareous deposit may be in the walls of angiectatic vessels or in their thrombosed channels, or it may be extravascular and a result of hæmorrhages or thrombotic necroses.

The lateral radiogram of the posterior portion of the head in No. 2 of Brushfield

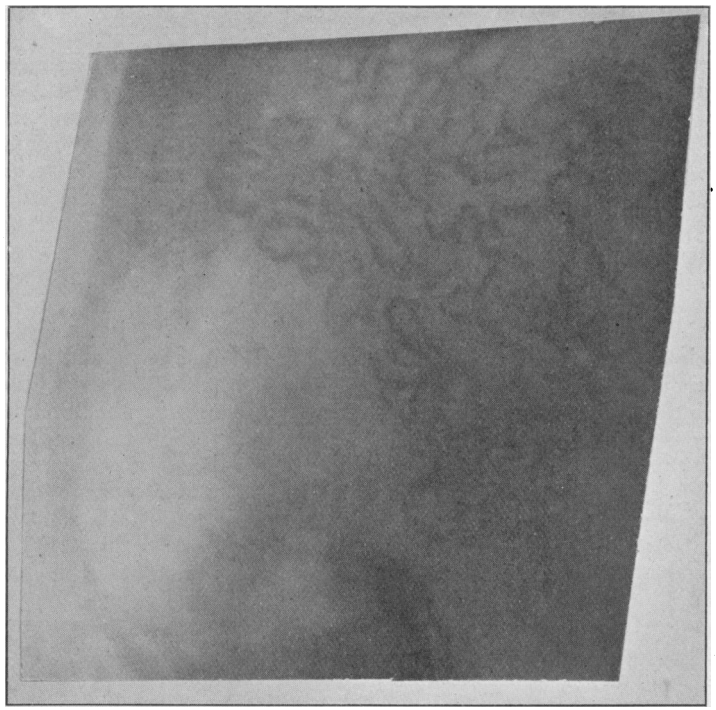

FIG. 4.—Lateral skiagram of the posterior part of the brain in Brushfield and Wyatt's second case, kindly furnished by Dr. Brushfield. It shows typical appearance of a partially calcified leptomeningeal racemose aneurysm in the occipital region, above the tentorium cerebelli.

and Wyatt's cases (a girl, aged 11) I am able to illustrate here (fig. 4) by the kindness of Dr. Brushfield. In all cases of large facial hæmangioma a radiogram of the brain should be taken, especially if any operation on the face be contemplated, but in the interpretation it must be remembered that (deep) facial as well as intracranial hæmangiomata may be partially calcified.

Unilateral congenital glaucoma on the side of the main capillary nævus of the skin and the cerebral disease has been found in other similar cases, namely in all three of Cushing's, two of Brushfield and Wyatt's (their first and second), and both of Aynsley's cases. Galezowski, in 1898, showed a girl, aged 9 months, in whom extensive capillary nævus of the face was associated with unilateral left-sided buphthalmus. It would be interesting to know if signs of unilateral (left-sided)

intracranial disease subsequently developed. In all Brushfield and Wyatt's cases, and in those of Hugo and Aynsley, and one of Cushing's there was idiocy or imbecility. There may be mental deficiency, but there is not idiocy in my case.

Direct or indirect involvement of the basal region in the neighbourhood of the pituitary gland in my case may explain the obesity and defect in sexual development. A combination of obesity with infantile hæmiplegia (Babonneix, Hutinel and Widiez) might sometimes be explained in the same way.

Prognathism of the anthropoid type has, I believe, not been shown by skiagrams in other cases, excepting in one of Brushfield and Wyatt's cases (Brushfield, *Proc. Roy. Soc. Med.*, 1929, xxii (Sect. Dis. in Child.), p. 13, discussion).

Unilateral intracranial hæmorrhage from abnormal blood-vessels connected with the meningeal angiomatous condition, occurring in early or in intra-uterine life, may have been the original cause of the spastic hemiplegia and epileptic fits (especially those of Jacksonian type) in some of the cases. The history in Cushing's cases supports this view, as does the history in Brushfield and Wyatt's first case, where the left hemiplegia was first noticed after the child had had a severe left-side convulsive attack at the age of six weeks. But the history given in Kalischer's and some other cases hardly does so. Intracranial hæmorrhage from angiomatous blood-vessels was the final cause of death in certain cases recorded by Wirgman, A. Esser, O. Latham, Worster-Drought and Dickson, and L. Nielsen. It is at least probable that the spastic hemiplegia arising during intrauterine or early life in some of the cases at present under consideration was due to intracranial hæmorrhage from the abnormal blood-vessels of a diffuse leptomeningeal angioma of the opposite cerebral hemisphere, that is to say, on the same side as the main vascular nævus of the face.

I have not attempted to enter into the whole subject of intracranial hæmangiomata associated with hæmangiomata in the skin or other parts of the body (Lindau, Schuback) or associated with multiple abnormalities, such as in Schubert's recent case in which a cerebral angiomatous tumour and one in the medulla oblongata were associated with a cystic pancreas and a small hypernephroma. Schuback refers to the association of capillary angiomata, but not cavernous angiomata, with syringomyelia. A remarkable example of partially angiomatous tumours of the skin and cerebral meninges, associated with other abnormalities, has recently been described by Drs. G. Brown and J. N. Jacobson. Their case was that of an imbecile girl, who died in a condition of status epilepticus at the age of $4\frac{1}{2}$ years. She had nævus-like patches distributed on the face, neck and shoulders of the left side; microscopically their structure appeared to be intermediate between cavernous angioma and simple pigmented mole. She also had a common hairy mole on the nape of the neck, a lymphangioma-like stripe on the chin, and a little dermoid-like tumour on the cornea of the left eye. In addition to these external abnormalities there were two small symmetrically situated intracranial tumours, apparently of angiolipoma-like structure, one of them attached by a pedicle to the pia-arachnoid in the cerebello-pontine angle on each side of the brain. The authors give references to similar pedunculated tumours of the cerebral meninges described by German pathologists.

These cases of Schuback, Schubert, and Brown and Jacobson, are examples of, or at least allied to, the so-called "Lindau's syndrome"—a term introduced since the publication of A. Lindau's "Studies on Cerebellar Cysts" in 1926. In Lindau's syndrome hæmangiomatous or hæmangiomatous-cystic disease of the cerebellum is associated with capillary angioma of the retina, and sometimes with hæmangiomatous and cystic lesions in other parts of the body, such as hæmangiomata in other parts of the nervous system, renal and pancreatic cysts and suprarenal adenoma. H. Cushing and P. Bailey ("Hæmangiomas of Cerebellum and Retina," *Archives of Ophthalmology*, New York, 1928, lvii, p. 447) confirm the association of retinal hæmangiomatosis with hæmangioma of the cerebellum, and illustrate the occasional

hereditary or familial nature of "Lindau's syndrome." They quote a new paper (1927), by G. F. Rochat, on familial retinal and cerebellar angiomatosis and refer to an equally recent (1927) article on "Lindau's syndrome," by Wohlwill (both published in Germany).[5] F. H. Leavitt (*Archives of Neurology and Psychiatry*, Chicago, 1928, xix, p. 617) has recorded the occurrence of a cerebellar tumour (I have unfortunately not seen the original description) in two almost certainly homologous (uni-ovular) male twins. It is now clear that patients in whom retinal hæmangiomatosis ("Hippel's disease"), which is sometimes a familial condition, has been noted (several instances have been recorded in scattered publications in England), should be re-examined in regard to the possible co-existence of intracranial lesions. The relation of retinal hæmangiomatosis to Lindau's syndrome seems to me analogous to that of unilateral buphthalmus or congenital glaucoma to the syndrome which forms the main subject of my present communication, namely, cutaneous and cerebral meningeal hæmangiomatosis.[6]

At the Société de Neurologie (Paris) on November 29, 1928, Lhermitte and Cornil showed a man, aged 39, with syringomyelia and syringobulbia and with multiple cutaneous pigmentary and vascular nævi.

With the intracranial hæmangiomata associated with hæmangiomata elsewhere may be compared the rare cases of angiectatic hæmangiomata of the spinal meninges associated with hæmangiomata of the skin or elsewhere. In Stanley Cobb's case the hæmangioma of the spinal cord was associated with cutaneous nævi of the same metamere. In Berenbruch's case a spinal cord angioma was associated with scattered angiolipomata and with an angioma of the kidney capsule. Chaput clinically described the case of a man with a large superficial angioma of the right flank and with atrophy of the right leg, the latter being conceivably due to pressure of a spinal meningeal angioma. In E. Herman's case of cavernoma cerebri there was spinal meningeal hæmorrhage possibly related to an abnormal vascularity in the spinal meninges of angiomatous nature.

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5 Other papers on the subject are quoted in Cushing and Bailey's magnificent monograph, "Tumours Arising from the Blood-vessels of the Brain" (1928), including H. Kufs, "Ueber heredofamiliäre Angiomatose des Gehirns und der Retina, &c.," *Zeitschr. f. d. ges. Neurol. u. Psychiatrie*, 1928, cxiii, 651-686.

6 A difference in colour between the two irides was present in my case, in one of Brushfield and Wyatt's cases, and in T. W. Sheldon's case (see ADDENDUM).

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ADDENDUM.—Dr. G. L. S. Kohnstam kindly tells me of a man, aged 45, a grocer's assistant, who has hæmangiectatic nævi of the lower lip. Three years ago he commenced to suffer from occasional epileptic convulsions of the Jacksonian type, beginning in the left hand. The Wassermann reaction was negative. On exploratory operation, Mr. H. S. Souttar found a condition resembling racemose arterial angioma of the leptomeninges over the middle of the motor cortex on the right side of the brain. A beneficial result was obtained by the local extradural insertion of radium.

Dr. C. Worster-Drought has kindly sent me the following notes of two cases in which extensive right-sided hæmangiomatous nævus of the skin was associated with left-sided (contra-lateral) Jacksonian epilepsy.

Case I.—Extensive Right-sided (mainly) Cutaneous Hæmangioma with Left-sided Jacksonian Epilepsy probably due to Right Cerebral Hæmangioma.

G. G., a girl, aged 6, seen March, 1928.

Present Complaint.—"Fits."

History.—The first fit occurred when she was 14 days old, and she has since had attacks on an average of one in two months, except during the last twelve months when a fit has occurred about every fourteen days. The cutaneous hæmangiomata have been present since birth.

Previous History.—Labour prolonged, otherwise normal. Measles at two years.

Family History.—Seventh child of a family of seven, all alive and well excepting one, who died age 7 weeks from summer diarrhœa. No similar condition or "fits" known in family. Mother died six months ago (aged 35) from "heart-failure."

Medical Examination.—Normal size and development. Extensive cutaneous hæmangioma involving right side of face and neck (only a slight patch on left cheek), practically the whole of right side of trunk and right arm (on the left side there is only a small patch below the clavicle and faint naevi on the left arm), right leg almost entirely involved and the left leg to a slight extent only). There is a massive vein running from the manubrium sterni downwards slightly to the left of the mid-line and bifurcating below the umbilicus, each branch disappearing on the anterior aspect of each thigh.

Central Nervous System.—Pupils dilated, left larger than right, reactions to light and accommodation normal. (The right conjunctiva shows slight injection between 3 and 6 o'clock.) Slight nystagmus on deviation of eyes to right. Other cranial nerves normal. Sensation, as far as can be tested, normal. Upper limbs normal. Abdominal reflexes and coördination normal. Lower limbs normal in motor power and tone. Left knee-jerk brisker than right. Ankle-jerks equal and normal. No ankle clonus, plantars flexor.

Other Systems.—Heart, lungs and abdominal organs nil abnormal. Pulse 90. Blood-pressure 95/75.

X-ray of Skull.—Nothing abnormal.

Cerebro-spinal Fluid.—Definitely blood-stained, the blood being intimately mixed with the fluid which was ejected during lumbar-puncture with pulsations synchronous with the heart-beat. 180,000 red blood-cells per c.mm., 120 white cells. Wassermann reaction negative.

Description of Fit.—Two days before a fit she becomes restless and difficult to manage. The right eye becomes injected (pink). At onset of attack head and eyes turn to right, falls to ground, becomes rigid, left-sided clonic movements follow, the arm being more affected than the leg. She is incontinent and has frequently bitten her tongue. As a rule similar fits follow one after another with intervals of sleep, the whole series being spread over six to nine hours.

Case II.—Right-sided (exclusively) Cutaneous Hæmangioma, with Left-sided Jacksonian Epilepsy, probably due to Right Cerebral Hæmangioma.

C. H. W., male, aged 22; seen March, 1922.

Present Complaint.—"Fits" and occasional occipital headache.

History.—The cutaneous hæmangioma (see description) has been present since birth. "Fits" began only four years ago, at the age of 18, and has since averaged one per month.

Previous Illnesses.—Fistula in ano, 1916; operation.

Family History.—Negative. No similar condition known in family.

Physical Examination.—Intelligent and of normal mentality. Extensive cutaneous hæmangiomata, *exclusively right-sided*, as follows: (1) Involving whole of forehead and neck

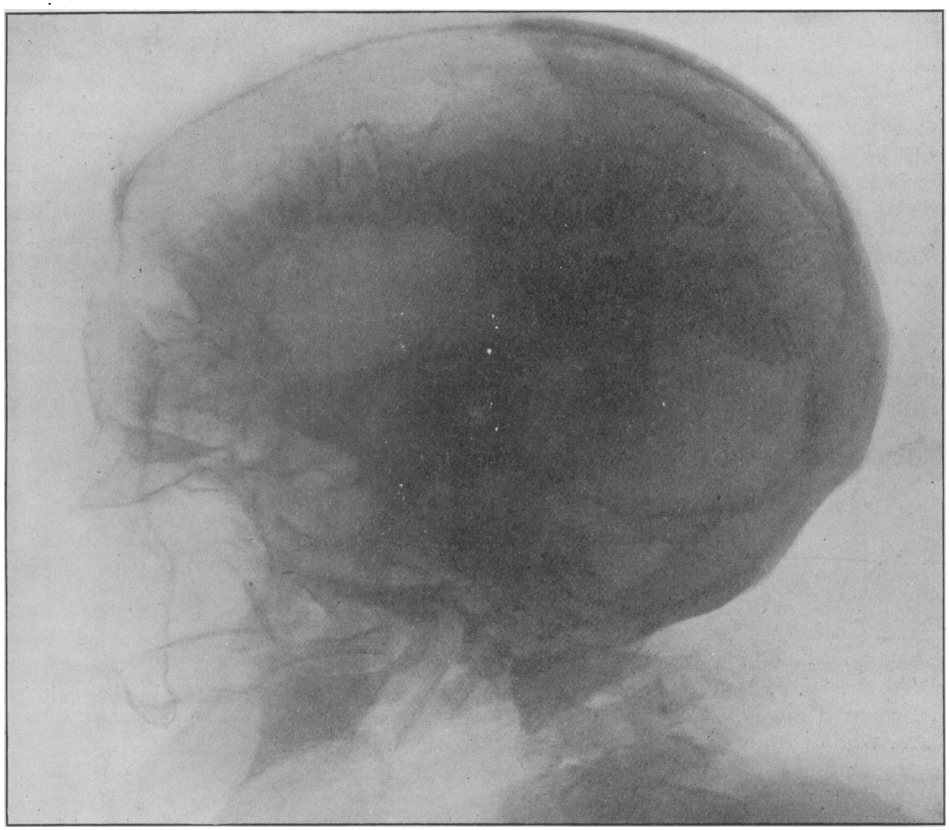

FIG. 5.—Dr. T. W. Sheldon's Case (description on p. 34).

down to level of clavicle; (2) whole of trunk in front from fifth rib to groin and back from spine of scapula downwards to iliac crest, with extension on to axillary part of right arm; (3) right leg from groin downwards, mainly on anterior, posterior and inner aspects (outer side escaping).

Central Nervous System.—Cranial nerves normal. Upper limbs: Motor weakness of right arm compared with left, but no inequality of deep reflexes. Sensation normal. Left knee- and ankle-jerks definitely brisker than right. No ankle clonus. Plantars flexor. Abdominal reflexes: right greater than left. Coördination normal.

Other Systems.—Nothing abnormal.

Skiagram of Skull.—Nothing abnormal.

Description of Fit.—Feeling of malaise for one hour or so before onset of attack, with *aura of multi-coloured lights in left eye*, then sudden onset with cry and fall and left-sided

clonic movements affecting mainly the *left* side of the face and left arm. The fits usually occur singly and last from three to twelve minutes.

Dr. T. W. Sheldon has kindly sent me the following note of a congenital low-grade imbecile in Tooting Bec Hospital:—

A man, H. P., aged 67, born with left-sided facial hæmangiomatous nævus and complete right-sided hemiparesis.[7] Roentgen-ray examination shows what seems to be a left-sided racemose meningeal hæmangioma, extending from the frontal region backwards to just above the level of the tentorium cerebelli. There is likewise a peculiar density of bone (not an artifact) in the posterior parietal region, and Dr. Sheldon adds that repeated attempts to nullify an unnatural thickening in the petrous area have failed (fig. 5). The case is an excellent example of unilateral vascular nævus (involving the face and the ocular conjunctiva), with contralateral hemiparesis and X-ray evidence of a meningeal racemose hæmangioma on the same side as the facial nævus. The age of the patient is also interesting. Wassermann reaction negative.

From H. Cushing and P. Bailey's recent monograph on "Tumours Arising from the Blood-vessels of the Brain" (1928, p. 26), I must quote the following:—

"Three remarkable examples of congenital facial nævi associated with epilepsy and in each of which a calcified lesion was radiographically disclosed in the occipital lobe have been reported from the Argentine, the first of them by V. Dimitri (*Rev. Assoc. méd. Argent.*, 1923, xxxvi, p. 63), and the other two by A. M. Marque (*Rev. Oto-Neuro-Oftalm.*, Buenos-Aires, 1927, i, p. 202). The patients were all children, of 13, 6, and 10 years respectively at the time the calcification was first observed."

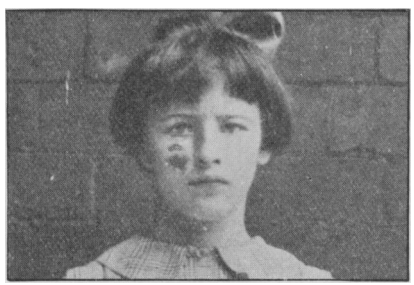

FIG. 6.

To Dr. W. J. N. Vincent of the South Yorkshire Mental Hospital (through Dr. G. Brown) I am indebted for the account of an imbecile and epileptic girl, aged 16 years, who died of tuberculous meningitis in 1926. She had a facial hæmangiomatous nævus confined to the distribution of the second division of the trigeminal nerve on the right side (see fig. 6), and was the subject of severe epileptic fits, which were general and not of the focal or Jacksonian type. There was no hemiplegia. On examining the brain at the necropsy, besides evidence of meningeal tuberculosis, there was an extensive localized hæmangiectatic condition of the vessels of the pia-arachnoid over the convexities of the cerebral hemispheres, especially over the post-Rolandic and parietal regions of the left hemisphere. The left hemisphere was smaller than the right hemisphere. It is interesting that in this case the meningeal hæmangiomatous condition was mainly contralateral to the facial nævus.

Discussion.—Dr. C. P. SYMONDS: Though familiar with the association of nævi in the trigeminal field with contralateral epilepsy or hemiparesis, of which I have seen several examples since the case (already referred to by Dr. Parkes Weber) which I showed at the Association of Physicians Meeting in 1925, it was not until September, 1928, that I first made acquaintance with an X-ray picture similar to that just shown. The patient, a boy aged

7 The case was shown by Dr. T. Brushfield (*Proc. Roy. Soc. Med.* (Sect. Dis. in Child.), 1929, xxii, 12), and these notes, with the figure, are given here by kind permission of Dr. E. Beresford.

11, was brought to me by Dr. W. H. Beach, of Hassocks, on account of a visual defect for which he had already been seen by Mr. Foster Moore. The characteristic X-ray appearances were discovered by Dr. Prowse, of Brighton.

The patient was born with a nævus affecting almost the whole of the left side of the face. This was contained within the field of the left trigeminal nerve, but by no means filled it. The upper and lower eyelids were both affected and the greater part of the cheek, including parts supplied by the third division of the nerve. Apart from this disfigurement he appeared to be in every way normal. He is at school and up to the standard of his fellows in work and play. His father is an invalid and is said to be suffering from tabes with optic atrophy. The patient's Wassermann reaction in the blood is, however, negative, and there are no clinical indications of syphilitic disease.

He was taken to see Mr. Foster Moore in September, 1928, on account of imperfect vision. The examination showed an incomplete right homonymous hemianopia (see charts, fig. 7). In addition to this there was a slight dislocation of the lens in the right eye and a congenital coloboma of the left disc. He suggested to Dr. Beach that the likeliest cause was a nævus of the visual cortex and advised that a skiogram of the skull should be taken. This was carried out by Dr. Prowse. I saw him a few days later and found nothing to add to the list of his physical defects except that the abdominal reflexes on the right side were diminished as compared with those on the left. Mentally he appeared normal. There was no defect of speech either on the receptive or the expressive side.

Stereoscopic X-rays make it clear that the shadows revealed in the lateral and antero-posterior views are those of a plexus lying throughout in close relation to the skull.

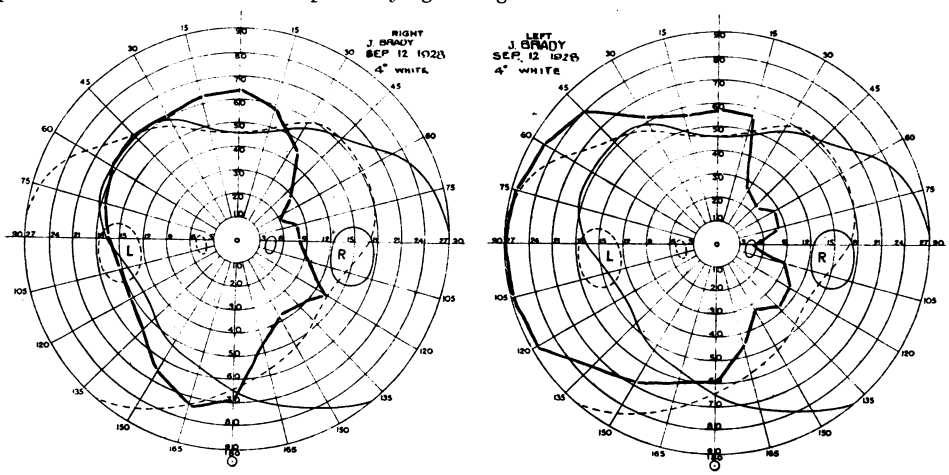

FIG. 7.—Dr. Symonds' Case: Visual Fields by Mr. R. Foster Moore.

A search through the literature of X-rays and of neurology provided no analogous picture, the nearest approach being the report of Dr. Parkes Weber's case already published in the *Journal of Neurology and Psychopathology*, 1922, but a few suggestive clues were obtained from papers dealing with calcifications of angiomata generally. Ruggles [1] reported five such cases in 1919. The clinical data are not recorded, but the diagnosis in each case was of an angiomatous tumour; one of these was of the jaw, the others of trunk or limbs. The X-ray pictures in each instance showed "multiple small cyst-like masses varying in diameter from a millimetre to a centimetre or more, with a thin shell, and an irregular mass in the centre, giving an appearance suggestive of encysted parasites. These spots are scattered throughout the growth and probably represent calcification of thrombi in the loops of the cavernous type of hæmangioma." Similar appearances in hæmangiomas of trunk or limbs have been recorded by Wakely [2], Ravold [3], Dunn [4], and Johnson [5]. I am indebted to Mr. Foster Moore for permission to quote his observations and reproduce the visual fields, and to Dr. Prowse for the X-ray pictures (figs. 8 and 9).

REFERENCES.

[1] RUGGLES, *Amer. Journ. Roent.*, 1919, vi, 512. [2] WAKELY, *Arch. Rad. and Elect.-therap.*, 1921, xxv, 363. [3] RAVOLD, *Rad.*, 1924, iii, 231. [4] DUNN, *Ann. Surg.*, 1925, lxxxii, 880. [5] JOHNSON, *Journ. Amer. Med. Assoc.*, 1928, xc, 1108.

Dr. R. M. STEWART said that some time ago he had had the opportunity of seeing Dr. Brushfield's cases, to several of which Dr. Parkes Weber had referred. About a year ago a case of this type had been under his care. The patient was an epileptic imbecile with an infantile hemiplegia on the right side and an extensive nævus of the body, particularly well marked in the left trigeminal area of the face, the line of demarcation between the normal skin and the nævus being sharply defined.

It was anticipated that post-mortem examination would disclose a cerebral or meningeal hæmangioma, but no evidence of such a condition was found. The left cerebral hemisphere was considerably smaller than the right, and there was evidence of an old softening in the region of the left superior parietal gyrus, but little else was found. He (Dr. Stewart) concluded from this case that the association of infantile hemiplegia with extensive nævus of the skin might occur without an accompanying intracranial hæmangioma.

Mr. L. BATHE RAWLING said that he had experience of two cases. The first was that of a boy of 6 years old, who since birth had suffered from attacks of stupor and convulsions. There was some bulging of the right temporal region, and this was explored. Some bone was cut away, exposing normal dura. When opened, a large angiomatous plexus was seen, extending beyond the area of brain exposed, consisting mainly of large tortuous veins. The wound was closed. This was in 1920.

In 1926 the boy was admitted to hospital with a large hæmatoma of the right eye region, right face and extending on to the neck. It was obvious that some vessel had burst, and that the blood had escaped freely outwards. He recovered and is now attending school, better in all respects.

The second case was that of a boy, aged 10, one of twin sons, and regarded at first as the healthier twin, though he had a nævus of right face, involving exactly the area supplied by the first and second divisions of the fifth nerve. When 6 months old he had had a series of convulsions, with left-sided Jacksonian fits. The left arm had remained weak, and he had never used it till he was nearly 4 years old. Every year up to 1926 he had had one or more of these convulsive attacks, all of which had presumably been due to the bursting of a vessel and hæmorrhage over the surface of the brain. Since 1926 the condition had improved and he had not had any severe seizure. The mentality, however, was poor—he was very irritable, only obeyed his nurse, and liked to throw things about. He was weak on the left side of the body and spastic. In reference to treatment, it seemed obvious that operative measures were useless and highly dangerous. It was proposed to send him to the Radium Institute, where Dr. Pinch was undertaking the case, and it will be highly interesting to see if any benefit accrues. The skiagram showed a mass of tortuous vessels over the right side of the brain, most marked in the occipital region and entirely above the tentorium cerebelli, with smaller scattered vessels, all somewhat calcareous, over the mid-brain and the frontal region (see fig. 10).

There were five specimens in the museum of St. Bartholomew's Hospital illustrating various degrees of hæmangioma of the brain, and apparently these angiomata extended, as a rule, deeply into the brain substance, and were seldom truly superficial in position.

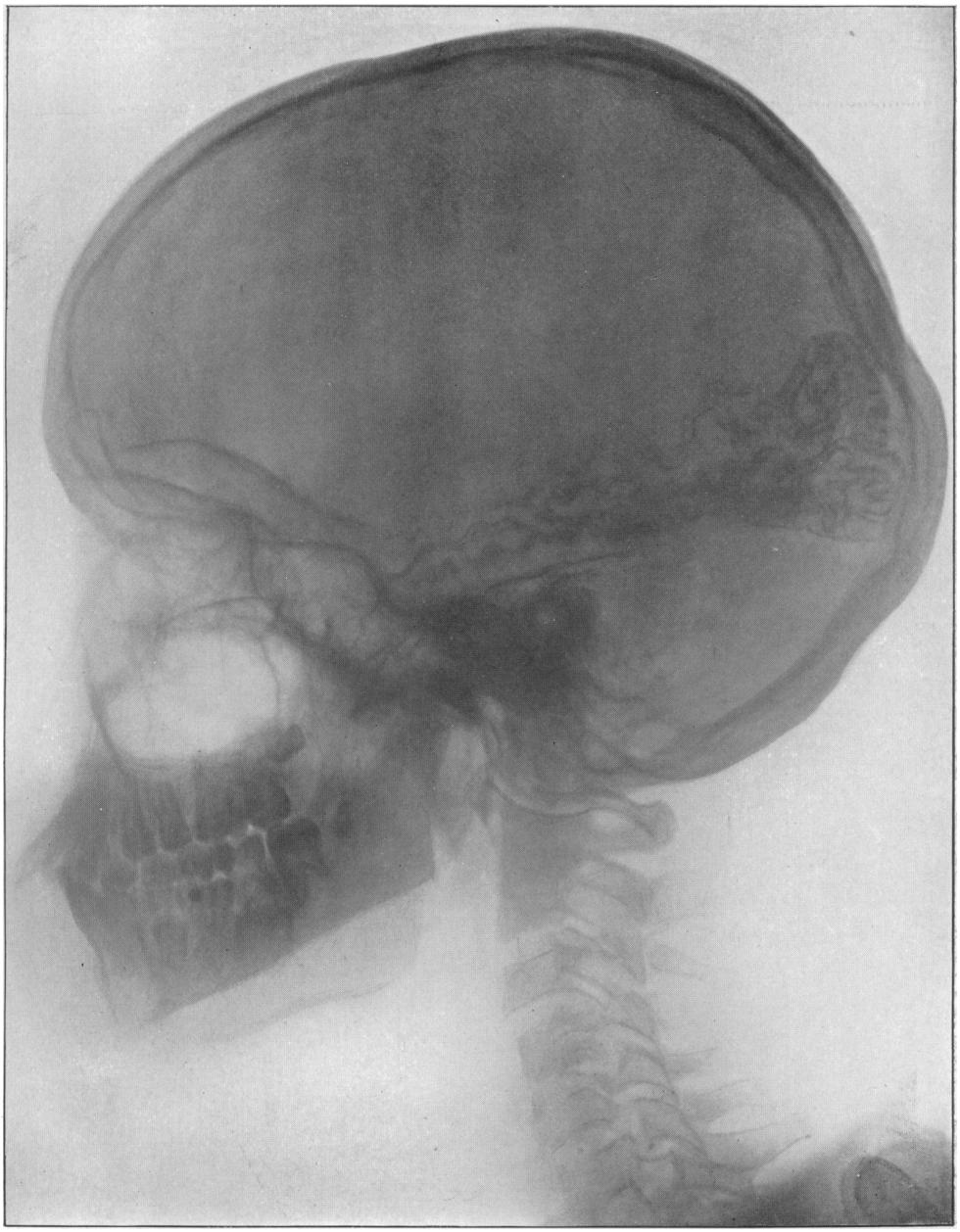

FIG. 8.—Dr. Symonds' Case: Lateral Skiagram.

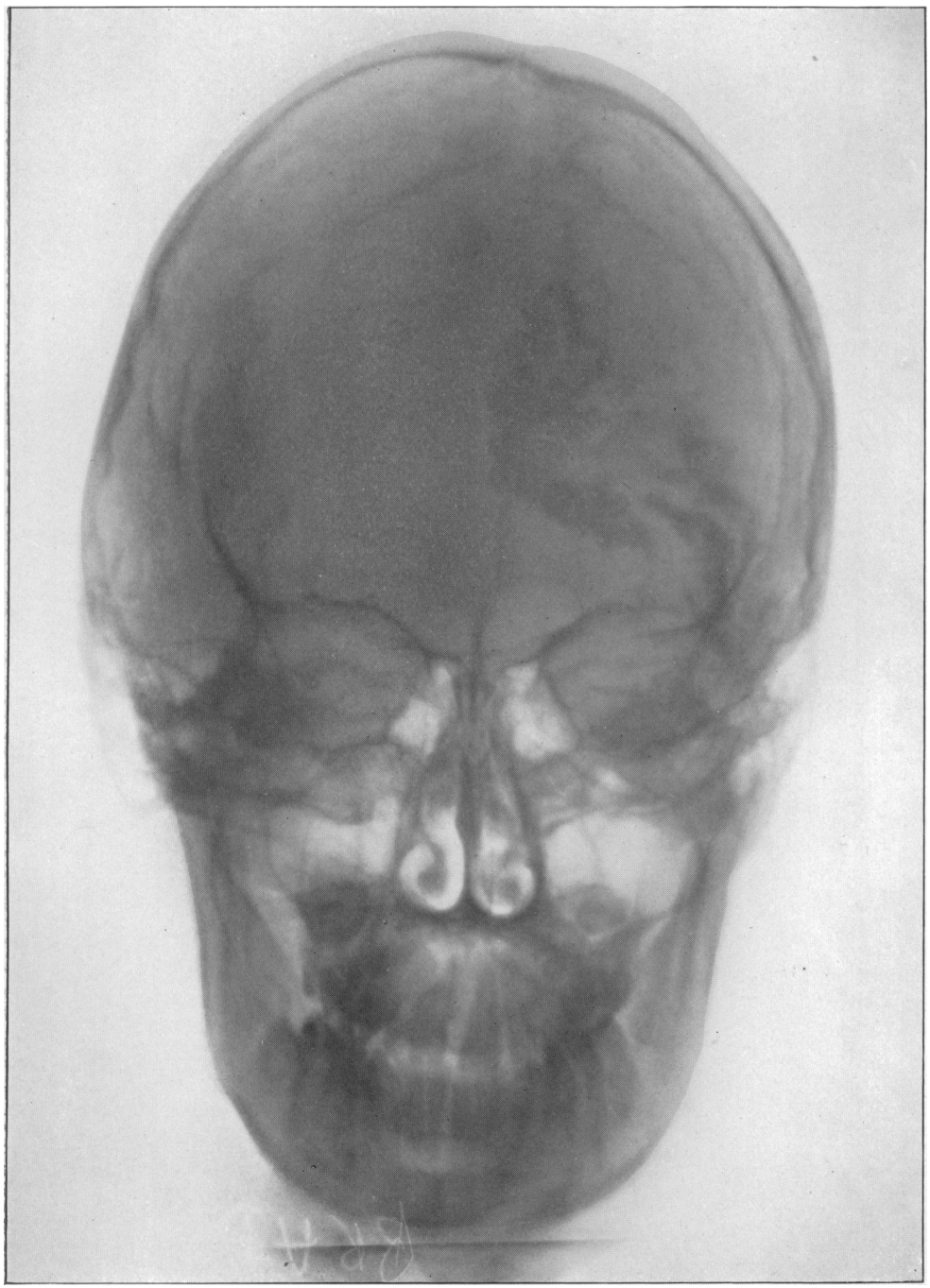

FIG. 9.—Dr. Symonds' Case: Antero-posterior Skiagram.

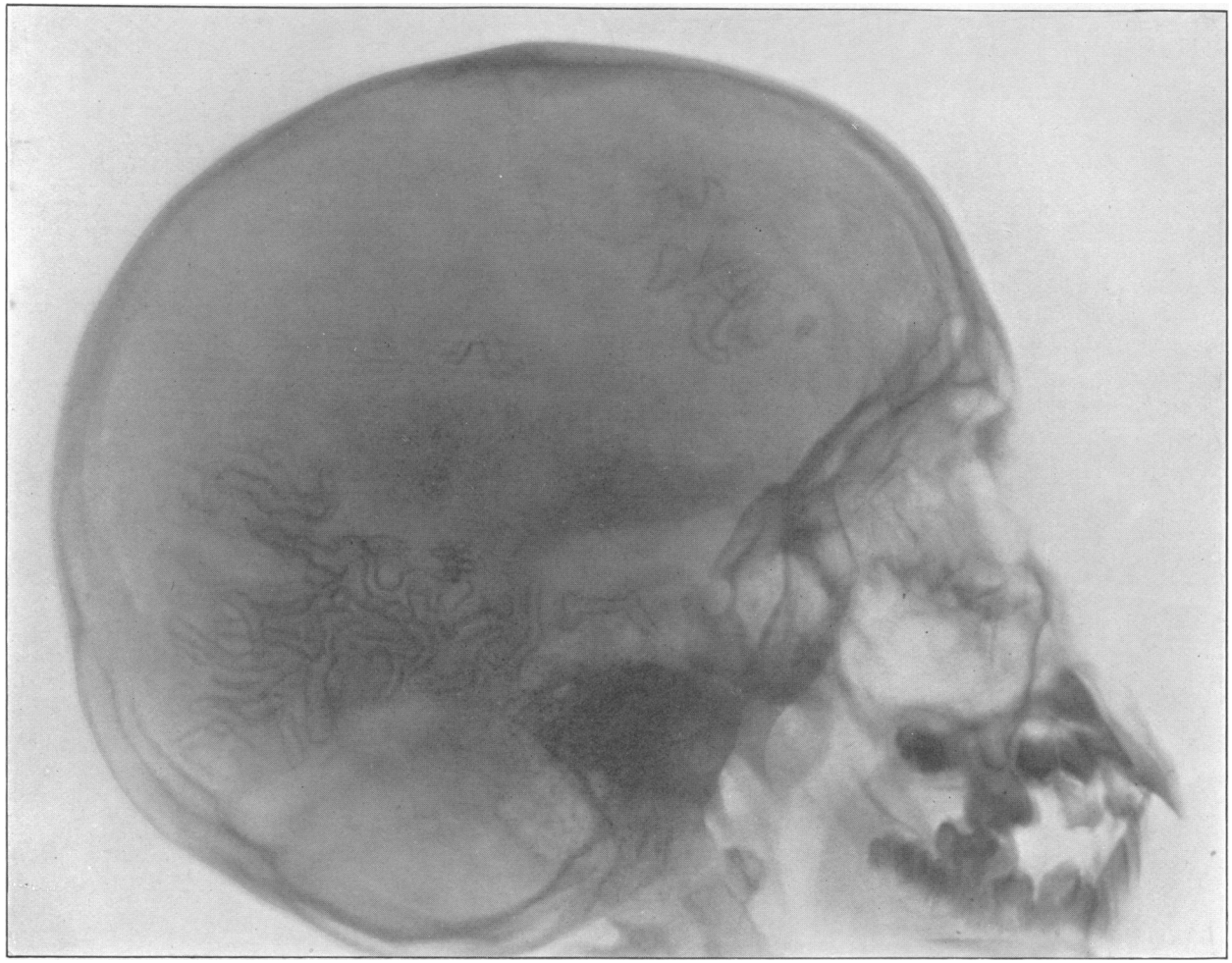

FIG. 10.—Mr. L. B. Rawling's Case: Lateral Skiagram (description in text).