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Encephala-trigeminal angiomatosis of Sturge-Weber

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ENCEPHALO-TRIGEMINAL ANGIOMATOSIS OF STURGE-WEBER

By

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I wish to express my sincere appreciation to Dr. Howard B. Hunt (Department of Radiology), who provided the material for three of the case presentations in this paper.

I am deeply indebted to Mr. William S. Cornwell of the Eastman Kodak Company for the use of a fourth case* and for the illustrative material presented with these cases.

* The case provided through the courtesy of Mr. William S. Cornwell has been previously published: Cohen, R., and Burnip., Nevoid Amentia., Ann. West. Med. and Surg., 3: 47-49, 1949.

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I. Introduction.

The case presentations reported here were prepared under the direction of Dr. Howard B. Hunt during the summer of 1949. The material was primarily intended for publication in the Eastman Kodak Company's technical journal: "Medical Radiography and Photography."

It is presented here in an enlarged and more detailed fashion. It would appear that this condition is more important than has been previously suspected. The three cases observed here from the state of Nebraska have been seen in the space of two years.

II. Encephalo-trigeminal angiomatosis of Sturge-Weber

A. History:

The syndrome of Sturge-Weber is generally characterized by cutaneous vascular nevi ("port-wine stain", naevus flammeus, naevus vinosus) found in the areas of trigeminal nerve distribution, associated pial and cerebral hemangiomata productive of neurological disturbances, and sometimes, glaucoma in the eye on the same side where the cutaneous hemangioma is observed.

Sturge's original case, reported in 1879, a six year old girl with a right-sided facial nevus, was afflicted with Jacksonian seizures beginning in the left hand. Sturge (1879) postulated that these seizures resulted from a "port-wine" mark on the surface of the right side of the brain. Kalischer (1897) demonstrated postmortem, in a child, that a "port-wine" mark of the face and scalp was indeed accompanied by telangiectasis of the leptomeninx on the same side. Volland (1913) described the appearance of a boy with a vascular nevus on the left side of the face and contralateral hemiplegia who was feebleminded and suffered epileptic seizures. At postmortem examination, the left cerebral hemisphere was found to be smaller than the right and it was observed that the pia mater overlying the left hemisphere was the seat of angiomatous formation, while the underlying cortex was atrophic and contained many calcareous concretions.

In 1922, and again in 1929, F. Parkes Weber called attention to these associated intracranial calcifications which were discernible in roentgenograms of the skull, and were located on the same side as the facial nevus. Dimitri (1923) independently

described similar roentgenological findings.

Over two hundred cases have been reported in the literature, but the terminology of this condition remains varied and confusing. Some of the terms proposed for the disease relate to various combinations of the names of pioneer students of the subject, such as: Sturge, Weber, Dimitri, Kalischer, Krabbe, and Brushfield-Wyatt, or to various phases of the disorder, for example, in cases accompanied by mental deficiency the term: nevoid amentia, and in cases with ocular involvement: nevus flammeus with glaucoma. Other terminology has attempted to provide an anatomic or pathologic description, such as, encephalo-trigeminal angiomatosis (Green, 1945), cerebral angiomatosis with trigeminal nevus, or nevus-pial calcification (Cohen and Burnip, 1949).

B. Incidence:

Little information can be gained from the literature as regards the incidence of this particular syndrome. Watson and McCarthy (1940) in studying the incidence and type of therapeutic response of 1,363 blood vessel tumors found in a total of 1,056 patients, stated that these individuals represented five per cent of the total admissions to the Memorial Hospital (New York) over an eight year period. Sixteen per cent of these patients had multiple lesions. Of the total, nineteen instances of " port-wine stain " were observed, twelve in males, and 88 per cent were seen to occur on the face and neck. No mention was made of Sturge-Weber disease, nor was mention made of any of the attending symptoms. From this series, it would appear that the incidence of "port-wine stain" is about 0.14 per cent, but the actual incidence of Sturge-Weber disease

itself can not be ascertained from this data. No extensive series of cases has been reported, although several authors have reviewed a number of cases. Nussey and Miller (1939) stated that they reviewed 145 cases in the literature, but no details of this review are to be found in their article. O'Brien and Porter have reviewed the ophthalmological findings in several cases which were associated with glaucoma. No idea of the incidence of encephalo-trigeminal angiomatosis can be gained from these papers.

C. Etiology:

This disease is congenital in origin and although pertinent records are not to be found, it is probably hereditary in origin, as may be suspected by analogy to related conditions, as the neurofibromatosis of von Recklinghausen, the tuberous sclerosis of Bourneville, or the familial hemangioblastomatosis of the cerebellum and retina of Lindau. The condition has been reported throughout the world, but chiefly in Europe, where it is more commonly recognized than elsewhere. There appears to be a slight preponderance of males (Nussey and Miller, 1939).

D. Pathology:

The lesion fundamental to the Sturge-Weber syndrome is essentially an anomalous development of the regional blood vessels to the skin, to the meninges, and to the underlying cerebral cortex. The development of the blood vessels in the brain has been divided by Streeter (1918) into five stages:

1. Establishment of a primordial vascular plexus;

2. differentiation of the plexus into circulating channels;

3. cleavage of these channels into three systems, that is, scalp, dural, and pial vessels;

4. adjustment of the vessels to the growth of the head structures;

5. final histogenesis of the vessels.

Any anomalous development affecting the vessels of the scalp, dura, or pia would necessarily occur during or before the third stage of embryological development. Angiomatous vessels lack the usual elastic and muscular layers and consist of little more than endothelium resting directly on adventitia. The rate of blood flow through the angiomas is retarded, as proven by Green (1939) through arteriographic studies. Lichtenstein and Rosenberg (1946) have suggested that since angiomatous tissue is mesenchymal in origin, angiomas may be considered to be a type of mesenchymal dysplasia. That this dysplastic hemangiomatous tissue may develop from a small focus of primitive mesenchyme is probable in that the final defect appears segmental or metameric. This is illustrated by the observation of metamerically distributed cutaneous nevi in association with spinal cord vascular tumors, often of a congenital origin (Cross, 1947). Whether this angiomatous mesenchymal dysplasia occurs in the mesoderm of the primative streak or in the mesenchyme of the neural crest, or from both, is undecided.

Since the encephalo-trigeminal angiomatous change results from a disturbance in the development of neuro-ectodermal tissue, it may occasionally occur in combination with other such disturbances arising earlier in the developmental history, such as, the tuberous sclerosis of Bourneville, the neurofibromatosis of

von Recklinghausen, or the familial hemangioblastomatosis of Lindau (Hublein, Pendergrass, and Widmann, 1940).

The association of calcification in the cerebral cortex with the neurological disturbances of Sturge's syndrome was first described by Hebold (1913) and histologically by Krabbe in 1934, who pointed out that calcification occurs extensively through the second to fourth laminae of the cerebral cortex. Green (1945) believed that primary angiomata were present within the affected brain tissue and that they underwent calcareous degeneration. Bergstrand, Olivecrona, and Tönnis (1936) believed that the calcifications occurred primarily in the small precapillary and capillary blood vessels and attributed cerebral sclerosis to this cause.

Cushing and Bailey (1928) attributed the calcifications to disturbance of circulation through the angiomatous channels, with consequent areas of focal infarctive necrosis. These sites of maximum calcification from the second to the fourth laminae of the cortex are at the level of the junction of the central and peripheral blood supplies of the brain wherein the vessels are smaller and probably more subject to vascular lesions.

The incrustations observed in radiographs of patients suffering from this malady have been presumed to be calcium or calcium complex compounds. In some cases, calcium salts have been found to be admixed with iron, indeed, exclusive ferrugination rather than calcification has been observed.

E. Clinical Manifestations and Diagnosis:

The patient with Sturge-Weber disease is born with a capillary

cutaneous hemangioma distributed over one or all of the branches of the trigeminal nerve. The hemangioma is not necessarily limited to the face, and is frequently multiple. Extension of the nevus into the mucus membranes of the oral and nasal cavities is a common finding. Neurological symptoms usually appear during infancy, but in some cases, symptoms have first appeared as late as the third or fourth decade.

The first symptom is commonly a focal convulsive seizure which tends to recurr more and more frequently and become more extensive; the disturbance may begin as a generalized convulsive seizure. Mental deficiency ranging from mild retardation to complete idiocy occurs in over half of the reported cases. The envolved extremities usually show muscular weakness and some underdevelopment and may show hyperactive reflexes. Various neurological disturbances such as personality changes, memory defects, headache, deafness, and vertigo have been reported occasionally.

Ocular manifestations in association with capillary hemangioma were first described by Schirmer (1860). In summarizing the occurrence of glaucoma with hemangiomas of the face, O'Brien and Porter (1933) pointed out that it occurs only when there is envolvement of the ocular structures, which may include the lid, conjunctiva, episclera, iris, or choroid. It must be noted that the site of the major hemangiomatous formations is the mesenchymal layer immediately bordering the ectoderm, as the skin in the face and scalp, mucus membranes of the mouth, nose, and throat, the leptomeninges, and the choroidal coat of the eye. The visual disturbances include homonymous hemianopsia, optic nerve atrophy,

choked disc, nystagmus, ocular palsies, anisocoria, choroidal atrophy, papillitis, congenital glaucoma, and enlargement of the retinal vessels.

Glaucoma is usually of the infantile type, but occasionally devlops in later decades. It is usually unilateral (may be bilateral, however) and occurs in about one-third of cases.

Cohen and Kay (1941) list four major symptoms of encephalo-trigeminal angiomatosis and believe that at least two are necessary to confirm the diagnosis. These are: convulsions, paralysis, mental retardation, and visual disturbances. Typical radiographic findings contribute to these criteria in the diagnosis of Sturge-Weber disease.

F. Laboratory findings:

Aid in confirming the diagnosis of encephalo-trigeminal angiomatosis of Sturge-Weber is generally not provided either by the routine analysis of blood and urine specimens, or by special serological, bacteriological, or chemical laboratory studies. Mild secondary anemia has been observed, probably on a dietary basis.

G. Radiographic findings:

Radiographic demonstration of calcification (ferrugination) along the cerebral cortex ipsilateral to the facial nevus was first noted independently by Weber in 1922, and by Dimitri in 1923. Such alterations occur in about eighty per cent of cases reported in the literature.

The calcium salts are laid down in wavy, tangled lines and in irregular flakes and masses which often tend to follow the general

outline of the cortical convolutions. The angular and supramarginal gyri of the parietal region appear to be most commonly envolved (Hublein, Pendergrass, and Widmann, 1940).

A relative underdevelopment in the size of the cranial cavity results from a hypoplasia of the cerebrum, and it is indicated by an ipsilateral slight flattening of the cranial contour, a thickening of the cranium primarily along the inner table, a relative enlargement of the mastoid cells and paranasal sinuses on the envolved side, and in severe cases, a relative elevation of the petrosal and sphenoidal ridges, with occasionally a shift of the calcified pineal body toward the side of cerebral hypoplasia. The vascualr channels of the skull are usually enlarged through the region of envolvement.

H. Special technics:

Encephalographic studies may show enlargement of the lateral ventricle underlying the area of hypoplasia (Hublein, Pendergrass, and Widmann, 1940) while the subarachnoid spaces overlying the hypoplasia may be widened or obliterated by an adhesive arachnoid thickening (Green, 1945). In the absence of cerebral hypoplasia, enlargement of the ventricle and widening of the subarachnoid space would not occur.

In Green's case (1945), angiography showed increased vascularity of the superior branches of the right anterior cerebral artery and of the parietal branch of the middle cerebral artery, forming a tangle of vessels extending into the region of calcification. Diodrast remained longer in the abnormal vessels, indicating a slowing of circulation time. Angiography would appear to be most

contributing in cases unassociated with calcification.

I. Prognosis:

The unfortunate individual afflicted with the encephalo-trigeminal angiomatosis of Sturge-Weber has a fairly good life expectancy. Constant danger of fatal intracranial hemorrhage is present however. Economically and socially, the patient's prognosis is poor. Mental retardation, physical appearance, and convulsive seizures discredits the patient from constant employment, so these individuals become a financial and social burden upon their relations. Frequently, institutional care devolves on the state.

J. Treatment:

Treatment of this condition is highly unsatisfactory, and the neurological disturbances tend to progress in frequency and severity with age. Sedatives and anticonvulsant medication may help to control the milder convulsive attacks, but fail to control the seizures as they become more severe.

Extirpation of the diseased cortex has been done by Cushing (1906), Green (1945), Sachs (1915), Geyelin and Penfield (1929), Bergstrand, Olivecrona, and Tönnis (1936), and others, and has resulted in variable control of severe convulsions, and has been associated with only slight progression in the neurological symptoms. The extreme danger of fatal intracranial hemorrhage following such a procedure has been repeatedly emphasized.

Cushing and Bailey (1928) have reported disappearance of a bruit, the patient becoming symptom free for six years, following irradiation therapy, and Dyke (1935) has had some measure of success with roentgen irradiation. Shafar (1946) on the other

hand, has reported that in his experience, x-ray therapy is of no value.

Capillary hemangiomas of the skin, so-called "port-wine stains" or birthmarks, do not respond to x-ray therapy as do those composed of larger vessels, as the nevus vasculosus and the cavernous hemangioma. This appears to be due to the presence of adult epithelial lining of the capillaries. In any event, shrinkage of an angiomatous tumor becomes apparent only after three to six months and is not fully effected until after two years. Shrinkage of dilated vessels might lead to a diversion of blood flow from the angioma into the adjacent brain tissue and to a reduction of any localized compression of the brain by the angioma. Cases with extensive calcification or definite cerebral hypoplasia must be considered unsuited for treatment by radiation since the existing lesion is already associated with deficient blood supply with more or less infarctive necrosis of cerebral tissue.

CASE NUMBER I (NEBRASKA):

The patient, a white female infant, four months of age, was admitted to the Children's Memorial Hospital, Omaha, Nebraska, with a history of convulsions occurring for the first time on the previous day. The convulsive attacks lasted fifteen to forty-five minutes and were not associated with cyanosis or loss of consciousness. Since birth there had been a large blotchy reddish discoloration over her face and scalp. The patient was one of twins; her brother showed no abnormality. The birth and the mother's preceding pregnancy had been uneventful; the family history was non-contributing.

Physical examination revealed a well-nourished, although extremely irritable child who presented twitching movements of the right arm and leg. The face was envolved by an extensive dark, reddish discoloration of the skin which extended into the scalp. This discoloration was slightly, if at all, elevated and blanched somewhat with pressure. The appearance of the patient's face is illustrated in the kodachrome prints of transparencies reproduced as Figures 1, 2, and 3. The mucous membranes were unenvolved; the eyes showed no abnormalities. There was muscular weakness of the extremities, especially on the right, with hyperactive tendon reflexes on that side. The Babinski was more marked on the left.

Laboratory studies showed a mild secondary anemia.

A clinical diagnosis of Sturge-Weber syndrome was made because of an extensive hemangioma of the face probably associated with an intracranial hemangioma productive of focal convulsive seizures and

muscular weakness

The convulsions varied from mild twitching to severe generalized contractions, envolving both sides of the body at different times, and lasting from a few seconds to twenty-five minutes. Treatment consisted of elixir of luminal and seconal for sedation. Deep x-ray therapy, 200 roentgens in air at 200 kilovolts was given over the area of the left motor cortex and was directed toward shrinkage of the vessels. The child was dismissed after a five day hospital stay.

At six months of age, the patient was readmitted with recurrence of the convulsions complicated by episodes of vomiting and failure to gain weight. There had been some temporary improvement after her previous therapy, but the convulsions had recurred, primarily on the right side, and were increasing in frequency and the patient had some cyanosis. The appearance of the patient on this second admission is illustrated in the kodachrome print of a transparency reproduced as Figure 4. The anterior fontanel was small; the right arm and leg were slightly spastic. Laboratory studies showed that the anemia was not so severe as previously demonstrated.

Radiographic examination of the skull, consisting of lateral and posterior anterior projections (Figures 5 and 6), showed a striated vascular pattern through the fronto-parietal region, the vault was smooth, and the sutures appeared slightly widened. Radiographic findings were interpreted as showing increased vascularization through the frontal region of the skull and some increase in intracranial pressure.

Seisures were uncontrolled by phenobarbital and dilantin.

Feedings were necessarily administered by tube. Additional deep x-ray therapy was given through the frontal region and was directed toward ultimate shrinkage of the vessels. In as much as shrinkage of hemangiomas requires about six months for evident shrinkage and two years for full response, possible benefit from this type of therapy could not be evaluated. It seemed improbable that any significant improvement would acrue in the clinical status of the patient.

Examination six months later demonstrated that the nature of the muscular weakness had progressed, and that the convulsions had only slightly decreased in their severity. The difficulties of having such a patient in the family have increased to the point where institutionalization has been recommended.

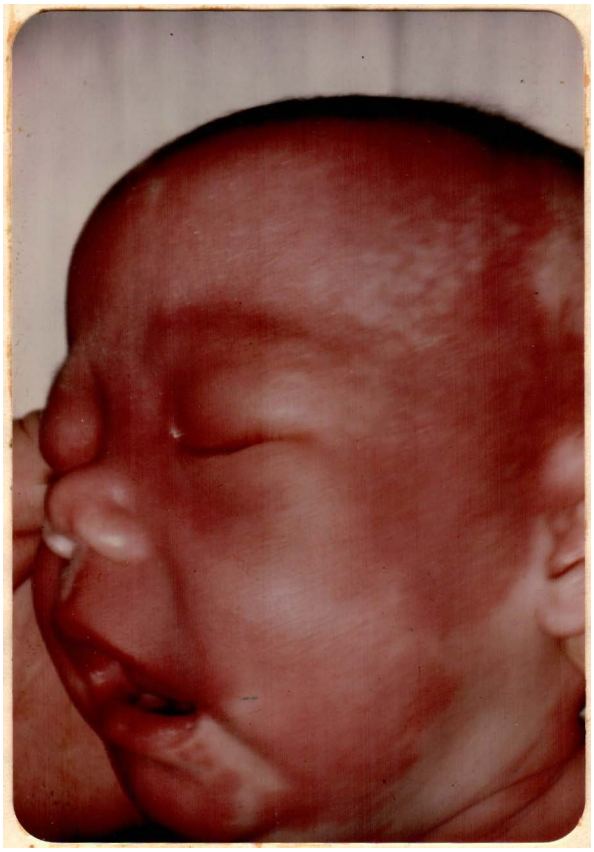


FIGURE 1

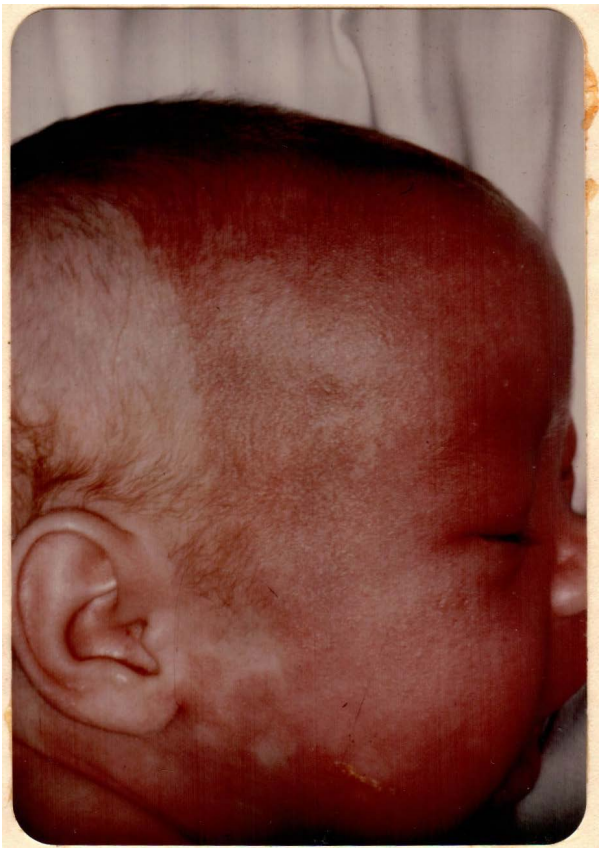


FIGURE 2

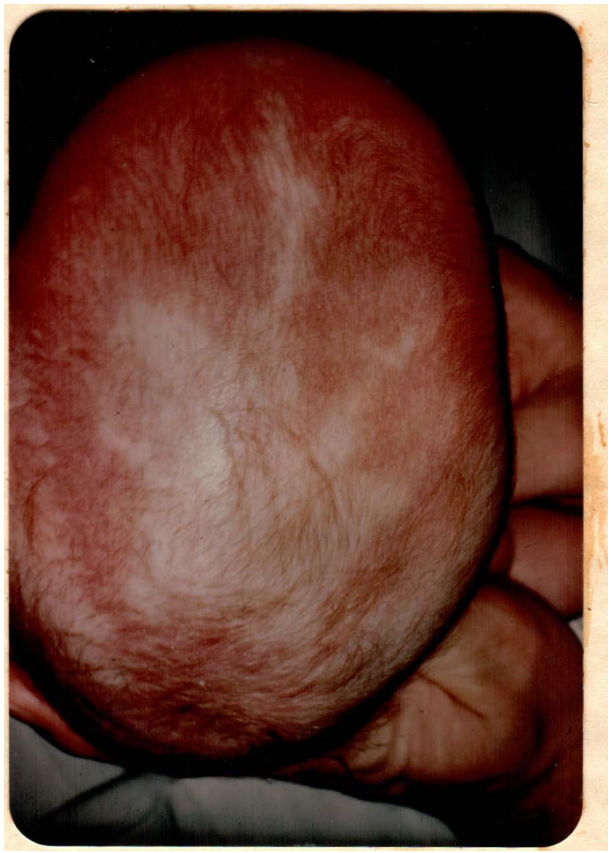


FIGURE 3

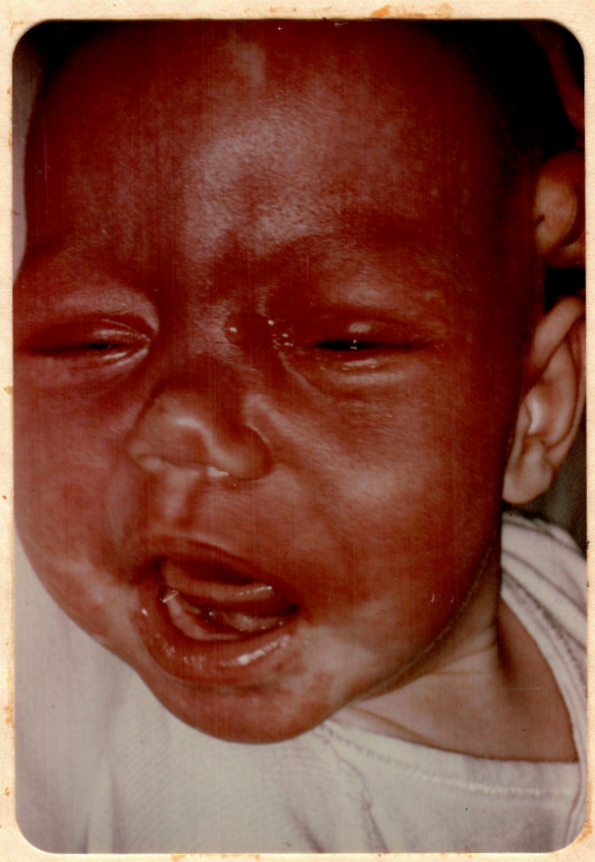


FIGURE 4

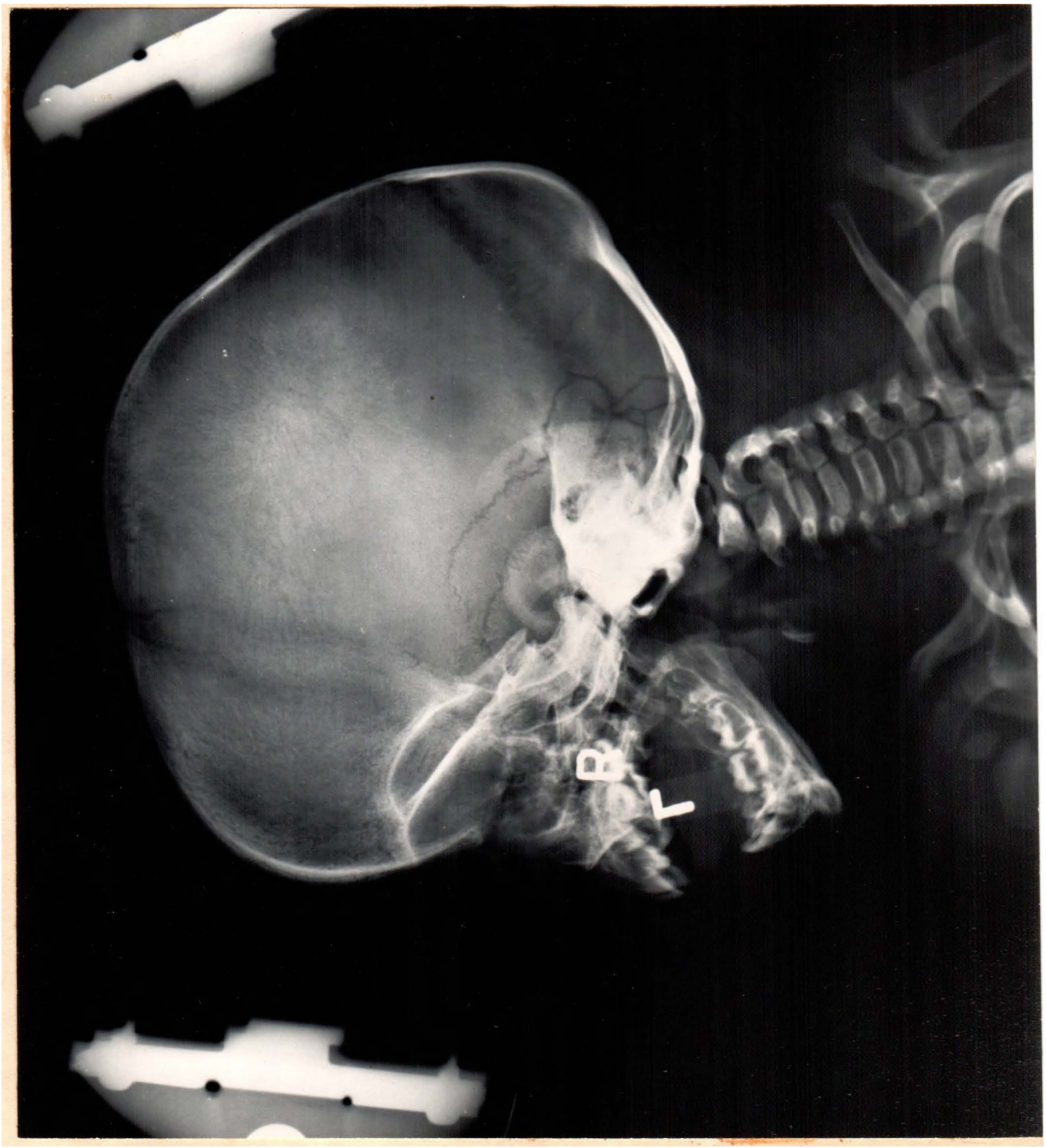


FIGURE 5

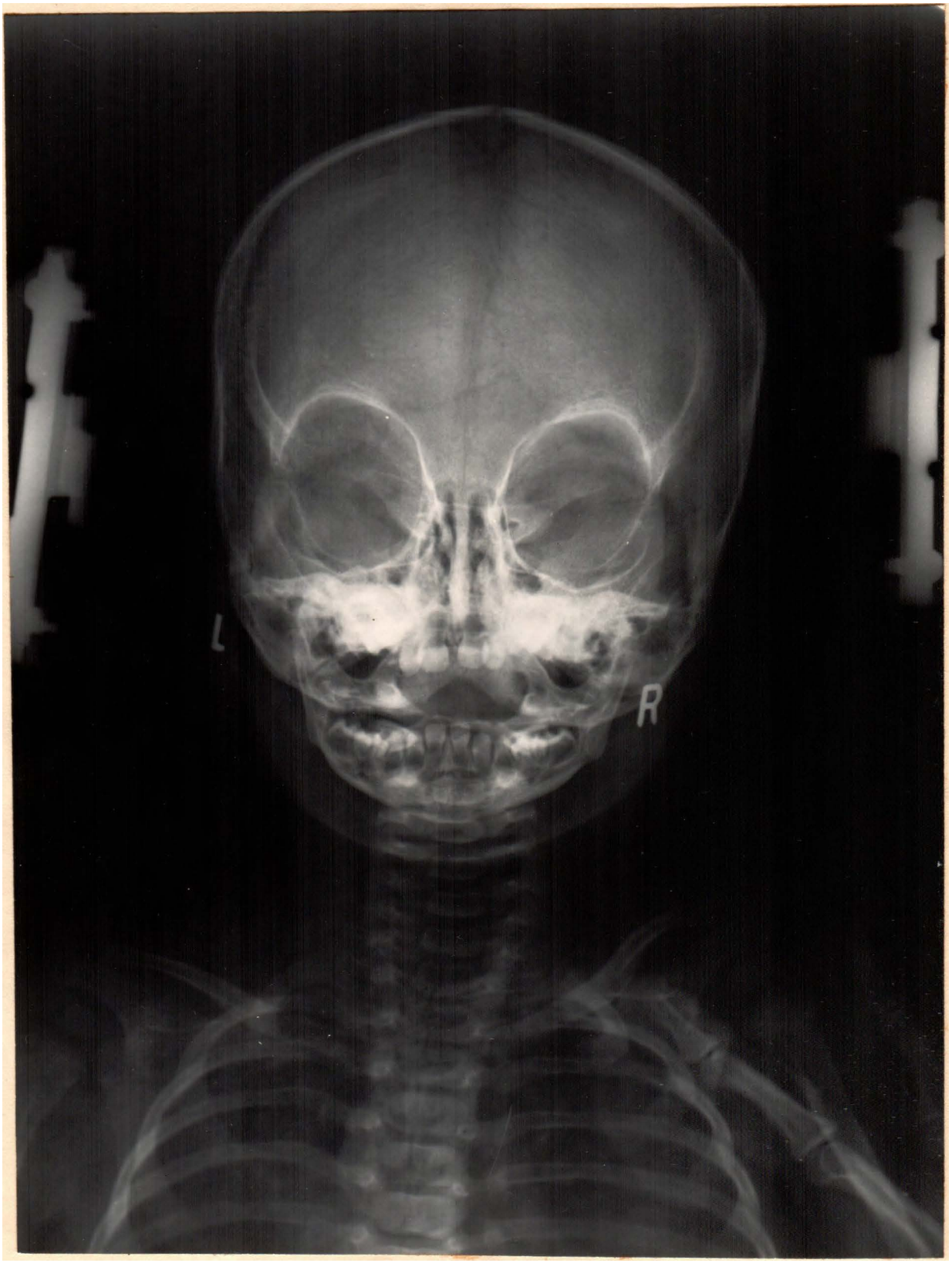


FIGURE 6

CASE NUMBER II (California)*:

The patient, a white boy six years of age, was born with a hemangioma over the left frontal region and following a fall from his bed at the age of seven months, had suffered frequent convulsions. These attacks were characterized by intermittent clonic and tonic contractions, screaming, and were often followed by periods of coma, lasting as long as forty minutes on one occasion. The patient's eyes rolled to the left during a seizure and after the child began to walk, he would stagger and fall at the onset of a convulsive episode.

The patient walked at the age of two, and began to talk at about four years. At the time of examination, however, difficulty in speech, a staggering gait, severe strabismus, the seizures, underdevelopment of the right arm and leg, and the hemangioma were the chief complaints.

The family history was essentially negative and the patient's delivery at full term was without incident.

Physical examination revealed a well-nourished boy who had a large pink hemangioma over the left frontal region in the distribution of the ophthalmic division of the trigeminal nerve. Both of the patient's right extremities were smaller in diameter and were less well-developed than the left extremities. Reflexes tended to be reduced or absent on the right side and hyperactive on the left side. External examination of the eyes revealed a

* This case was kindly provided through the courtesy of Mr. William S. Cornwell, Eastman Kodak Company, and has been previously published. (Cohen, R., and Burnip, R., Nevoid Amentia, Ann. West. Med.& Surg., 3: 47-49, 1949.)

marked internal strabismus with apparent functional impairment of the right rectus extraocular muscle. Ophthalmoscopic examination revealed increased pallor in the temporal half of the optic discs bilaterally. The blood pressure was 80 millimeters of mercury systolic over 60 millimeters of mercury diastolic.

Laboratory examination of the urine, blood cell morphology and the hemoglobin concentration of the blood revealed no pathologic alterations. Examination of the spinal fluid was negative; there were no positive reactions to skin tests with either a tuberculin or a coccidiodin antigen.

Radiographic examination of the skull in the lateral and antero-posterior projections (Figures 7 and 8) revealed evidence of intracranial calcifications in the right occipito-parietal cerebral cortex. These calcifications appeared to follow the convolutional markings of the brain and were located superior to the line of attachment of the tentorium cerebelli.

The combination of the presenting complaints and the radiographic findings led to a diagnosis of Sturge-Weber's encephalo-trigeminal angiomatosis. No therapy other than sedative medication was given to the patient, this tended to limit the number of convulsions to about one every two weeks. The patient is being observed monthly for any signs of improvement or deterioration.

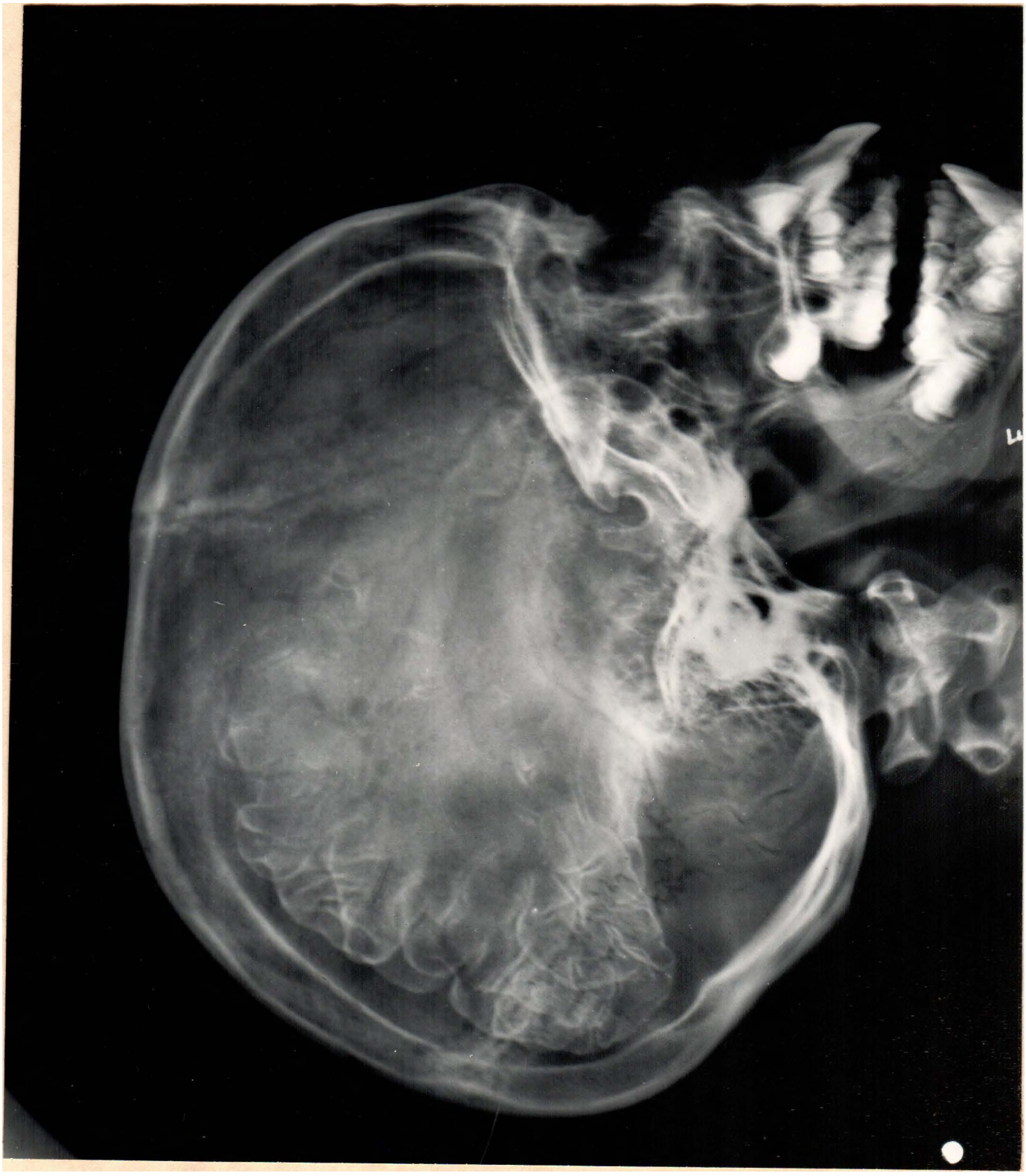


FIGURE 7

CASE NUMBER III (Nebraska):

The patient, a white schoolboy, twelve years of age, entered the Nebraska Methodist Hospital presenting a blotchy hemangioma over the left frontal region and complaining of convulsions of increasing frequency and severity. At birth, a dark reddish discoloration had been noted over this same region, which, although at first regarded as a bruise, was soon recognized as a hemangioma envolving the orbital and frontal regions extending superiorly and posteriorly to the line of the coronal suture. At the age of eight months, convulsive twitchings of the right arm began, and these continued to occur intermittently. By the time the patient was one year old, the twitchings also envolved the right leg, and at the age of three, the patient's face twitched and his eyes rolled during these seizures. Following such an attack, the right arm and leg would be limp.

The patient was placed on sedating medical treatment at the age of five, without a recurrence of the seizures during the year he was carried on treatment; furthermore, there were no convulsions during the following four years during which time the patient received no sedating medication.

The convulsive attacks recurred at the age of ten, and became progressively more frequent and severe. At the time of examination, the patient was having two to three attacks a week, each lasting from one to three minutes.

During an attack, the patient would not loose consciousness, but his eyes rolled and he was unable to answer when spoken to. The patient's coordination was fair; he rode a bicycle and participated in play. He was left handed and had some disability in the

use of the right hand. An average student in school, the patient seemed to be well adjusted in relation to his playmates and his family.

Physical examination showed a dusky, red, bltochy discoloration envolving the orbital and frontal areas of the forehead and scalp. The patient's appearance is illustrated in the kodachrome prints made from transparencies reproduced as Figures 9, 10, and 11. The discolored areas blanched on pressure and the vascularity was associated with only slight thickening of the hemangioma which was of the capillary type. The left side of the face seemed slightly more developed and fuller than did the right. The right arm and leg were slightly although definitely underdeveloped, and somewhat hypotonic and relatively weak as compared to the left limbs. The eyes showed no abnormality in tension or appearance except for some vascularity of the conjunctiva.

Radiographic studies of the skull in the lateral, posterior anterior, and antero-posterior projections (Figures 12, 13 and 14) showed irregular, tangled strands and flakes of calcification within the region of the left mid-frontal gyrus approaching the region of the anterior central gyrus and envolving an area about 15 by 15 by 45 millimeters. There was relative porosity and accentuation of the diploic patterns through the left frontal area of the skull, extending from the supraorbital region upward, backward, and laterally. No other significant abnormality of the skull was noted.

The patient's convulsions were uncontrolled by phenobarbital and dilantin in moderate doses. High voltage roentgen therapy in the

amount of 350 roentgens was delivered into the left frontal area, through anterior and lateral ports in an effort to bring about shrinkage of any underlying angioma within the meninges or brain.

The patient was observed six months following this treatment; his symptoms had not improved, neither had they progressed. Cerebral angiograms were done and these demonstrated a reduced blood supply in the region of the angiomatous calcification. The supply of this angioma appeared to be related primarily to the anterior cerebral artery. The patient was to be subjected to surgery in an attempt to remove the area of focal calcification. The results of this operative intervention are not available.

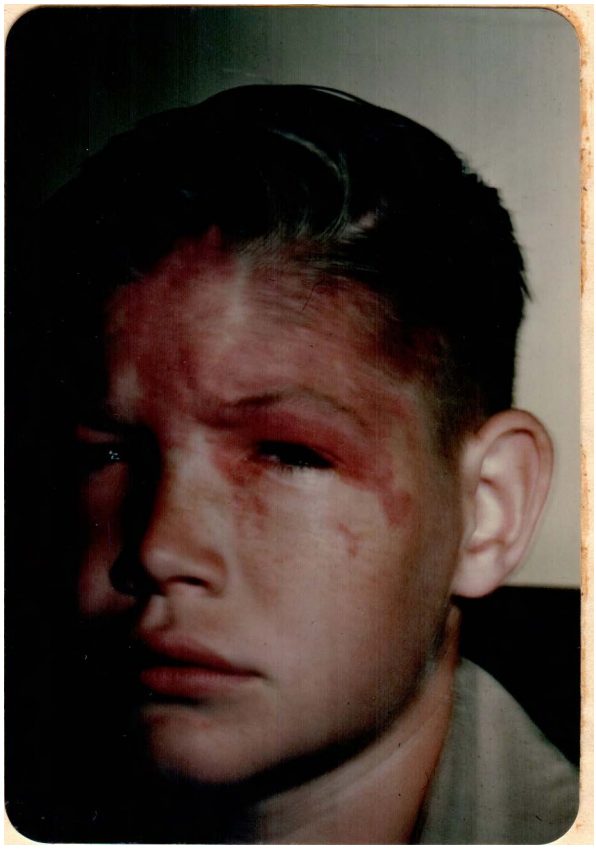


FIGURE 9

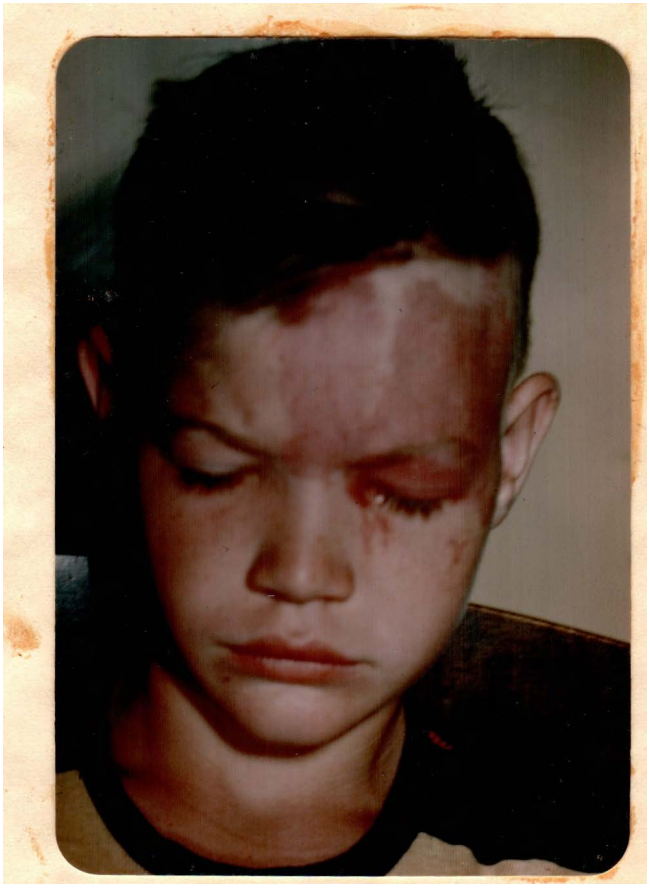
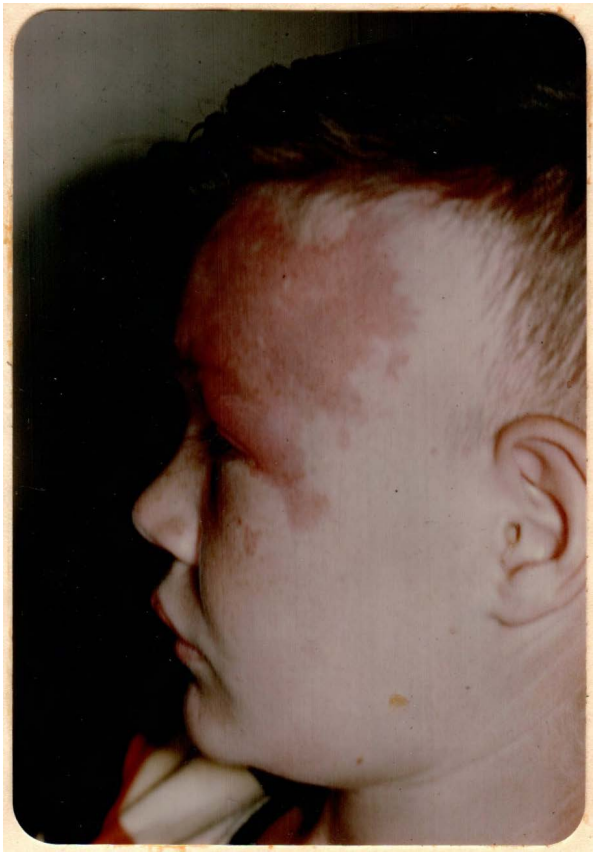


FIGURE 10



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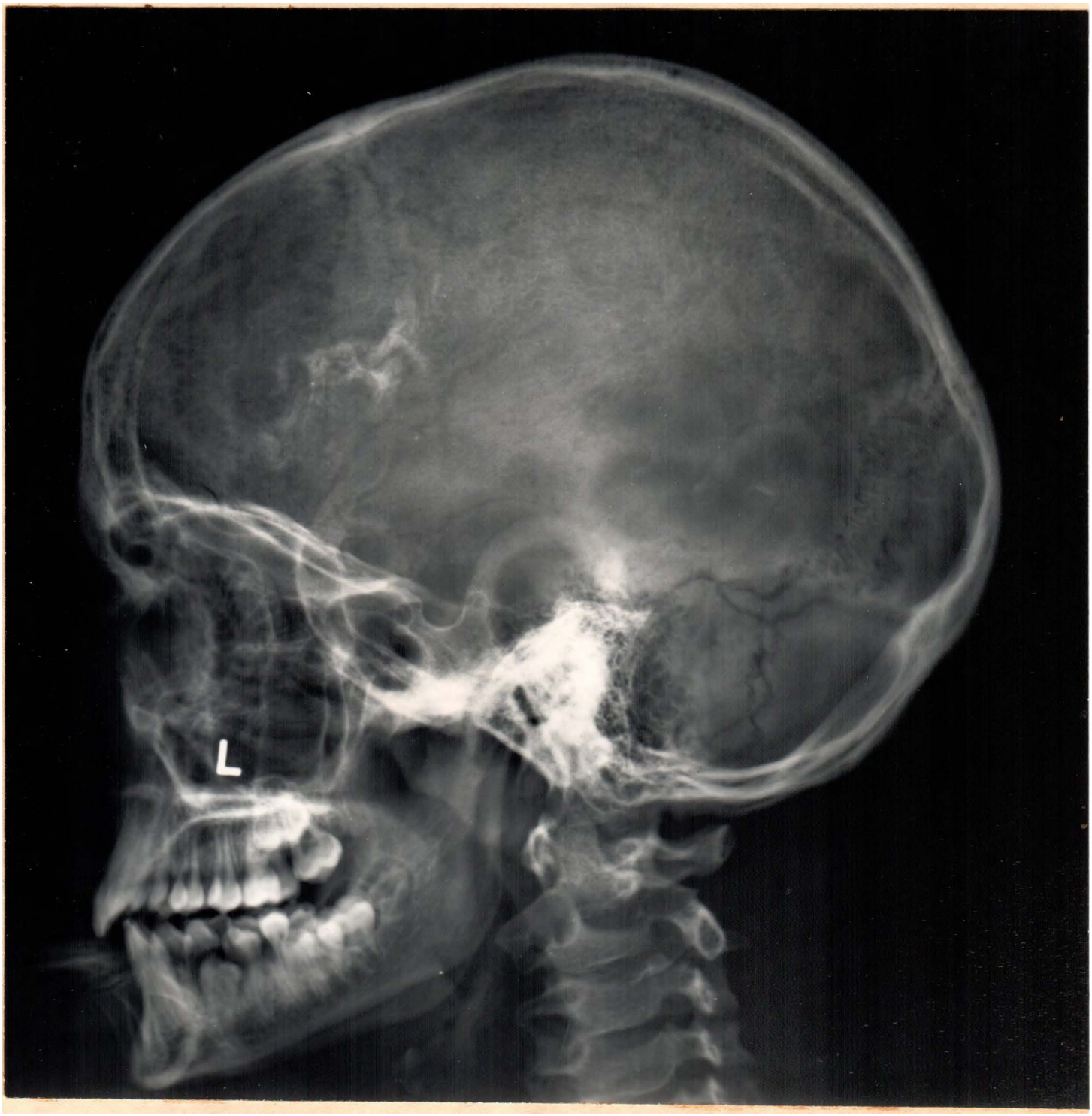

FIGURE 12

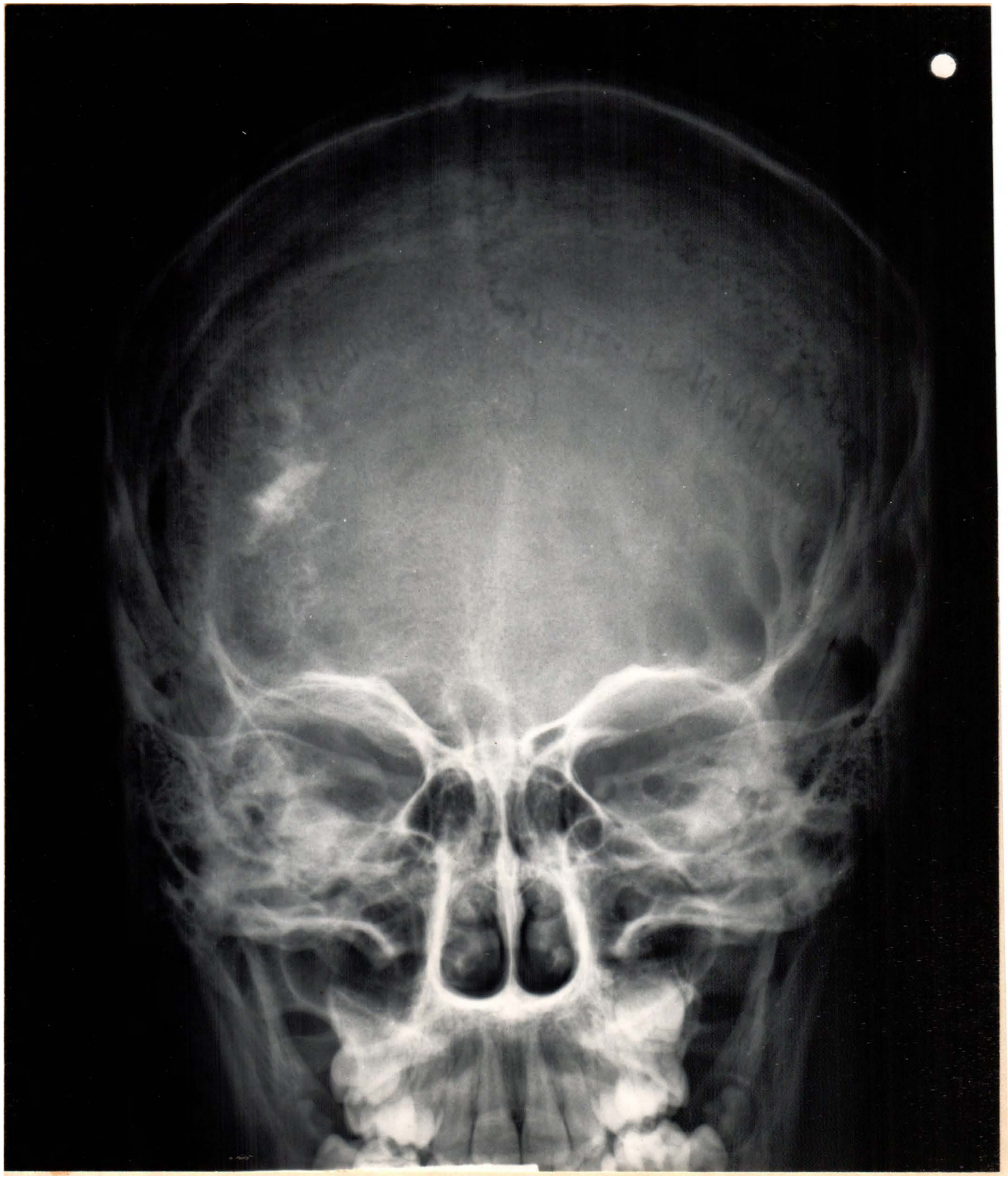

FIGURE 13

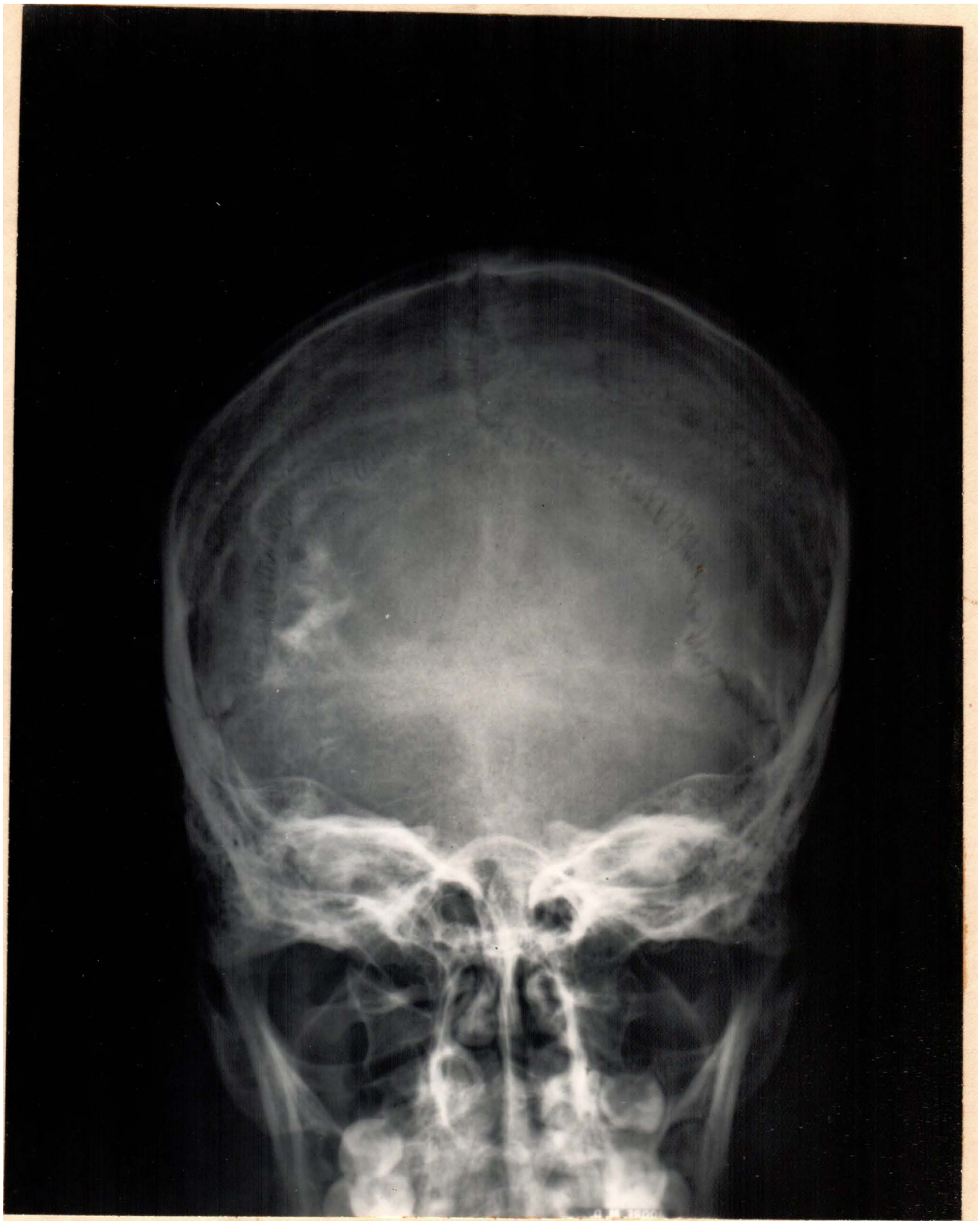

FIGURE 14

CASE NUMBER IV (Nebraska):

The patient, a single white unemployed woman, 32 years of age, entered the University of Nebraska Hospital for the first time seeking correction of a deformity, a facial birthmark, by means of plastic surgery. Since birth, she had been troubled by an extensive vascular birthmark covering the entire left side of the face, part of the right side of her face, and covering part of her shoulder on the right; a deforming fullness of her left lip, difficulty in breathing due to plugging of her nostrils; poor vision in the left eye; weakness and atrophy of the right arm and leg; and convulsive seizures which started in the left shoulder, became generalized and recurred at irregular intervals.

On physical examination, the patient was found to have a capillary hemangioma covering the left half of the face over the entire distribution of the trigeminal nerve, while on the right side of the face, a similar hemangioma was restricted to the course of the maxillary division of the fifth nerve. This hemangioma extended into and along the left half of the oral and nasal cavities. The left half of the upper lip was swollen to three or four times its usual size by a cavernous hemangioma. The appearance of the patient is illustrated in the print made from a kodachrome transparency reproduced as Figure 15. The right arm and leg were weak and their muscles atrophic; tendon reflexes were comparatively hyperactive on the right. There was evidence of some mental retardation.

The vision of the left eye was poor and had not been corrected by glasses. Ophthalmoscopic examination of this eye revealed a posterior synechia, irregular contraction of the pupil, vitreous

opacities, complete retinal detachment and evidence of an old severe chorioretinitis. Laboratory findings were non-contributing.

Radiographic studies consisting of stereoscopic lateral, posterior, and fronto-occipital projections of the skull (Figures 16, 17, and 18), presented a relative underdevelopment of the left cranial cavity and intracranial calcifications over the left posterior cerebral regions. The cranial vault on the left measured ten millimeters in thickness compared with a thickness of five millimeters on the other side, this thickness being primarily through the region of the inner table. The mastoid cells and the sphenoidal and ethmoidal sinuses were larger on the left than on the right. Vascular channels were increased, more particularly through the left side of the skull. There was calcification along the left parieto-occipital region laid down in an irregular cauliflower-like pattern conforming to the convolutional and vascular configurations of the cortex and meninges. In summary, radiographic findings indicated a hypoplasia of the left cerebrum associated with extensive meningeal and cortical calcifications through the parieto-occipital region.

The diagnosis of Sturge-Weber's encephalo-trigeminal angiomatosis was made by the neurology service. Dilantin and phenobarbital were given as anti-convulsant therapy during the patient's hospitalization. It was felt that neurosurgical and plastic surgical procedures offered no benefit. The cavernous hemangioma of the left upper lip was given 420 Roentgens in air of 80 kilovolt x-ray therapy, and this same treatment was repeated three months later. The patient was discharged to the radiotherapy outpatient service and arrange-

ments made for nursing home care.

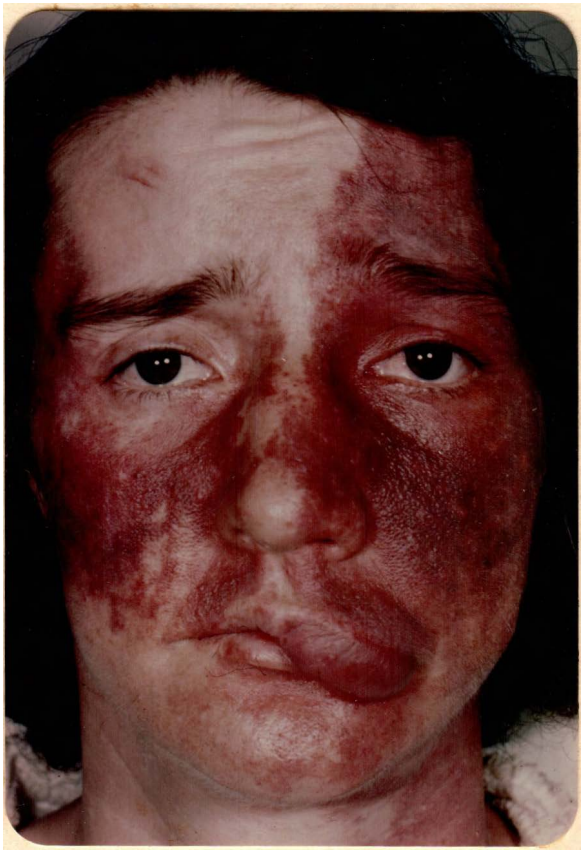

FIGURE 15

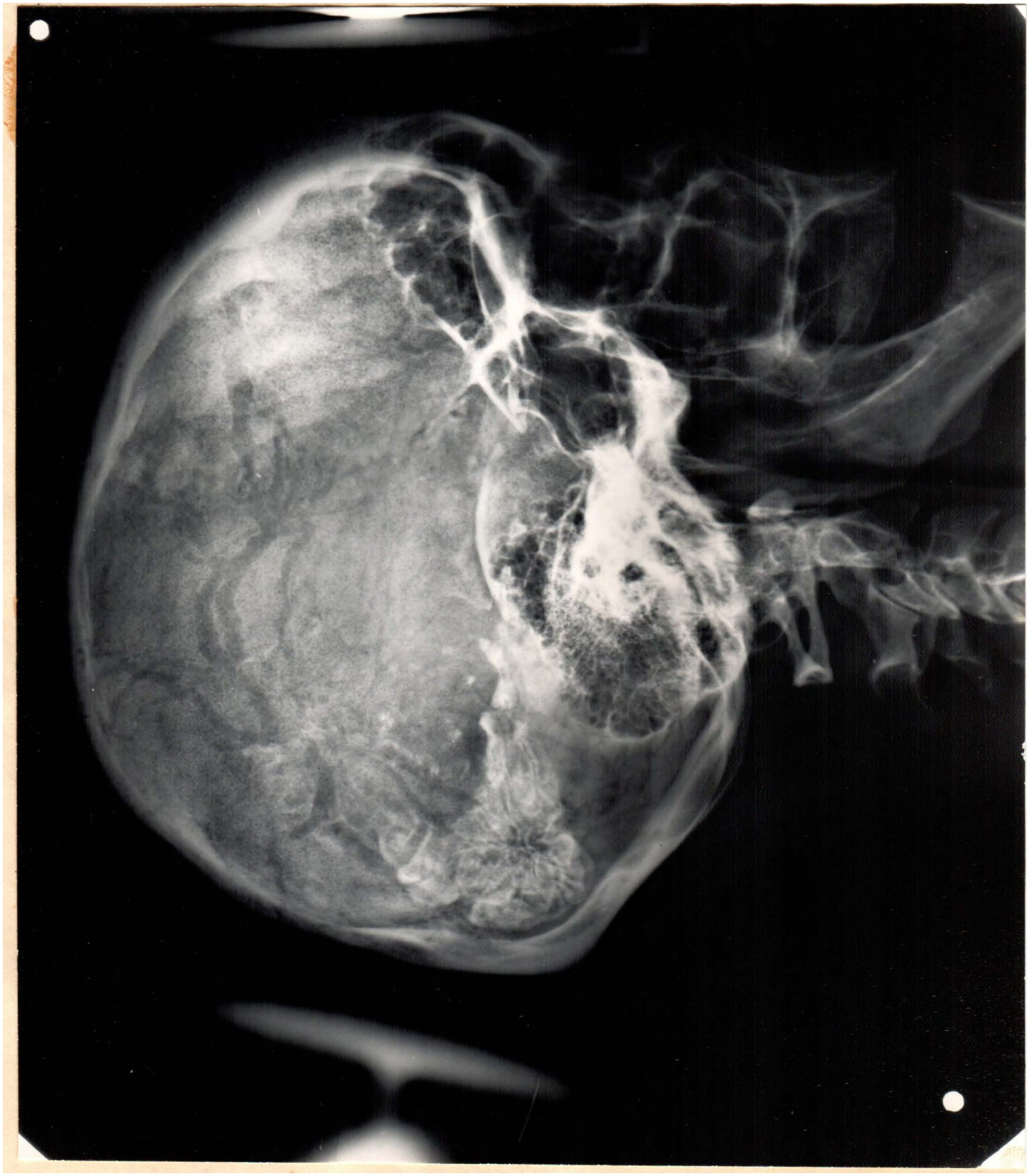

FIGURE 16

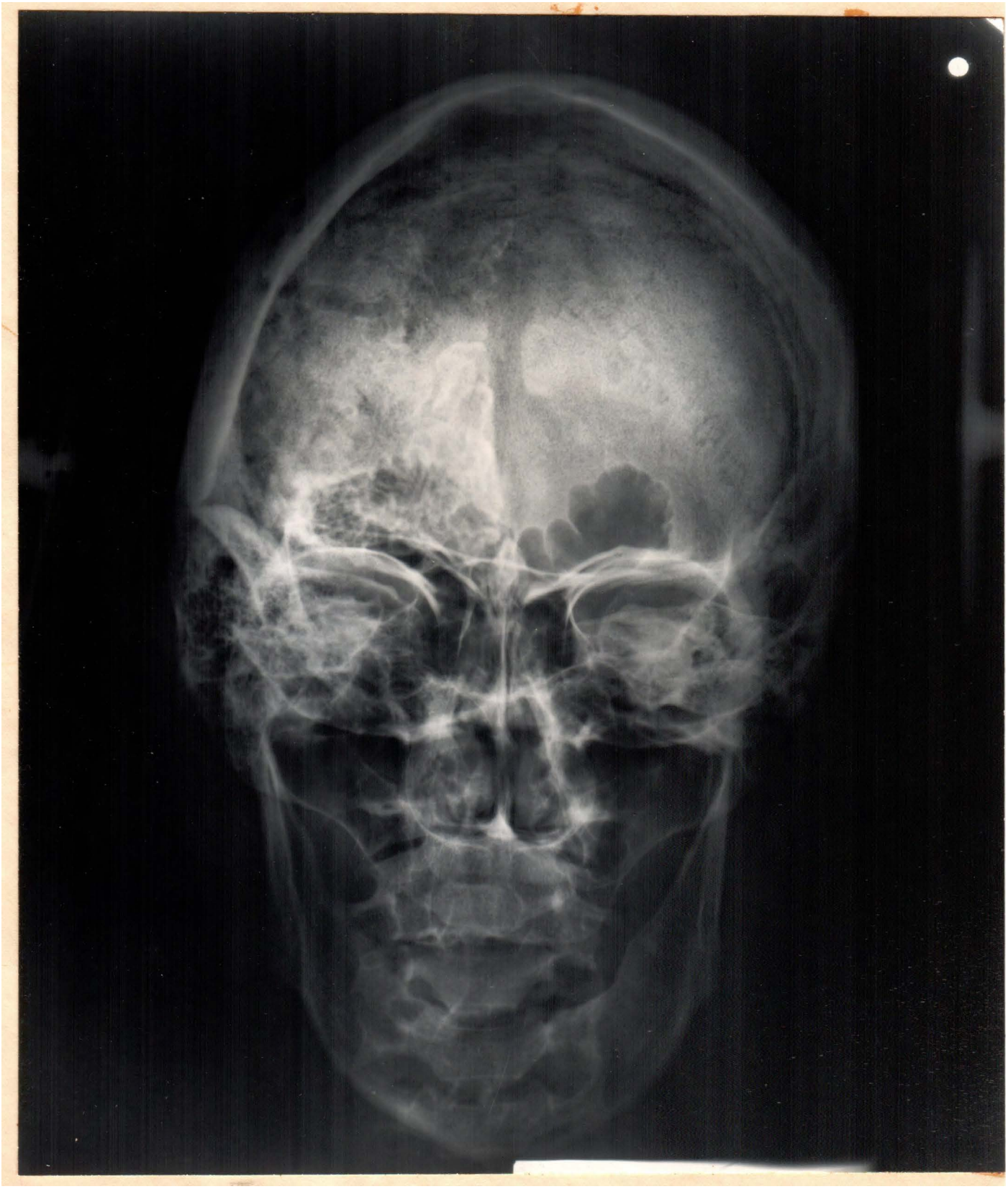

FIGURE 17

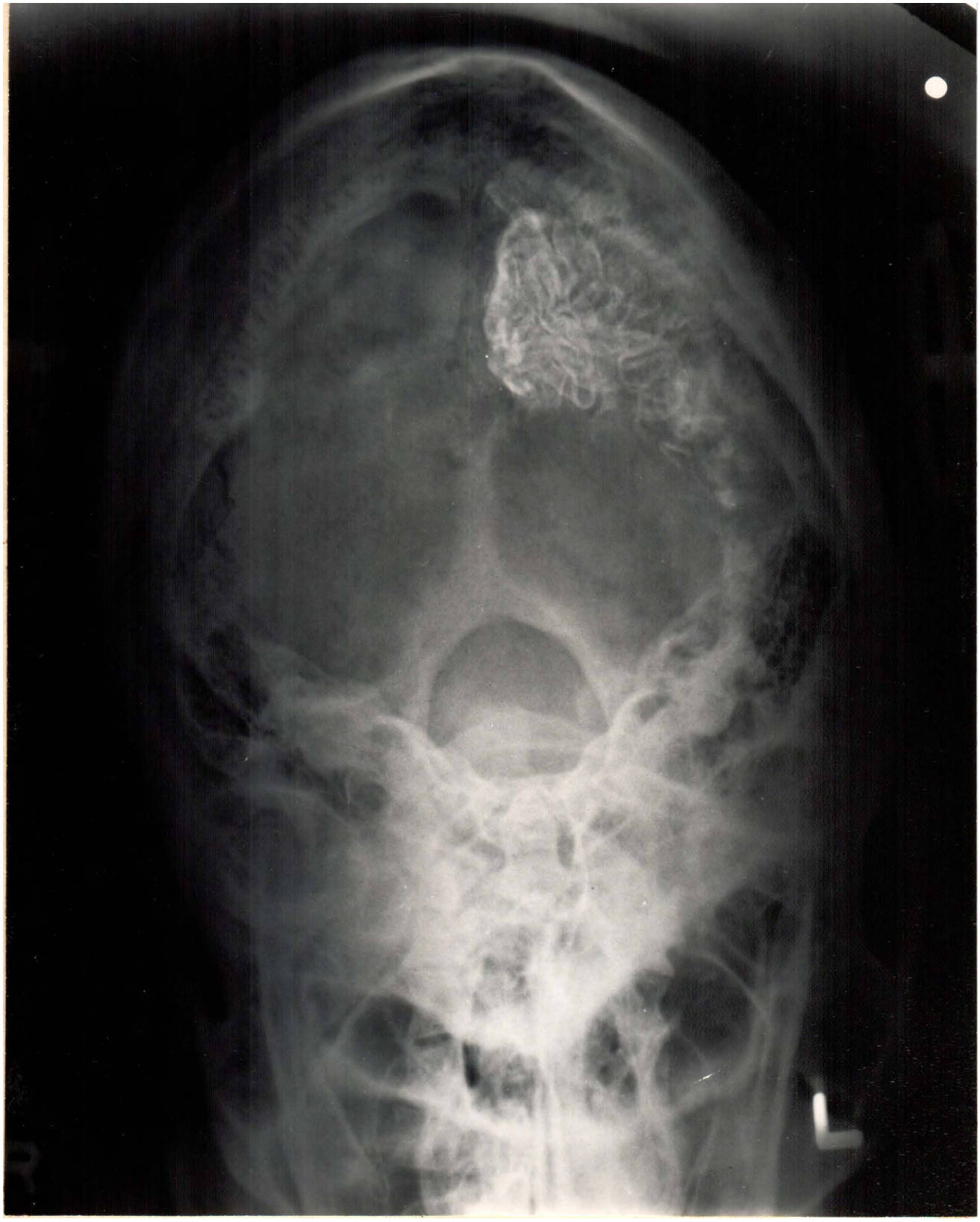

FIGURE 18

DISCUSSION:

The diagnosis in the four cases presented here is fairly certain, since in each case the diagnostic criteria are fully met, in particular with regards to Case Numbers II and IV. The appearance of the facies is for the most part characteristic, the neurological lesions typical, and the radiographic findings practically diagnostic. It will be noted that roentgen irradiation has been attempted in two cases. In the instance of Case Number I, the child is young enough so that the calcific lesions in the intracranial hemangioma are not yet observed. It is hoped that by the use of x-ray, some shrinkage of this vascular tumor might be obtained. In the instance of Case Number III, permanent changes are present in the cerebral cortex as demonstrated by radiographic examination, but the application of radiotherapy in this case was done so because although convulsive seizures were fairly severe, none of the rest of the symptomatology was as severe and incapacitating as has been seen in other cases. If any possibility of controlling the seizures might be entertained, therapy would be justifiable in the hope that the incidence of his seizures might be reduced, allowing the individual to have a more normal life than may be usually expected in encephalo-trigeminal angiomatosis. Surgical therapy has been resorted to in this case after a six month interval, since radiation therapy has not proved of immediate benefit.

The extreme severity of the lesion in Case Number IV makes any type of therapy hopeless. Shrinkage of the lip cavernous hemangioma may be expected, but the cerebral changes are too far advanced to expect any important change, had radiation therapy been attempted.

It is of interest to note that the lesions were apparent in all of these cases at the time of birth.

That three of the cases reported here have been observed in the State of Nebraska alone in the space of a little over two years time suggests that the Sturge-Weber syndrome is to be more commonly found than has been previously anticipated.

SUMMARY:

1. A brief description of a clinical entity usually termed "the Sturge-Weber syndrome" but which may better be named "the encephalo-trigeminal angiomatosis of Sturge-Weber" has been presented.

2. Four case presentations of this uncommon disease have been made. It is of interest to note that three of these have been observed in Nebraska in the course of two year's time.

3. Sedation remains the primary symptomatic treatment of this condition. Some of the problems envolved in the treatment of this condition by means of roentgen irradiation have been set forth. It may be mentioned that adequate trial of deep x-ray therapy in Sturge-Weber's encephalo-trigeminal angiomatosis has not been fully and properly evaluated.

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