LIPOMATOSIS OF THE CENTRAL NERVOUS SYSTEM

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Lipomas of the central nervous system are not uncommon, approximately 80 cases having been reported. The most usual site is the dorsal surface of the corpus callosum, although they have frequently been found in the region of the tuber cinereum, the mammillary bodies, and the midbrain. Lipomas of the spinal cord unassociated with spina bifida are much less common, only 12 cases having been recorded. Involvement of both the brain and spinal cord is extremely unusual.

In 1935 Krainer (1) summarized the literature on lipomas of the brain and spinal cord and added two cases of his own. Since then publications by Misch (2), Sperling and Alpers (3), and Scherer (4) have appeared.

Misch reported two cases occurring in the region of the foramen magnum. The first was an extradural lipoma $2.5 \times 4 \times 1$ cm. in a fourteen-year-old girl. It originated in the lower end of the spheno-occipital region and extended across the width of the clivus, compressing the medulla in its upper part. The second patient, a woman of thirty-four, had an intradural tumor, $2.5 \times 1.7 \times 2.2$ cm., lobulated, firm and yellow, with branching vessels on its surface. It filled the greater part of the right side of the foramen magnum and reached from the upper margin of the jugular foramen to a little below the atlanto-occipital articulation.

Sperling and Alpers record a lipoma $7 \times 5$ cm. as an accidental post-mortem finding in an elderly woman. The tumor was chiefly in the third ventricle and extended into the anterior horn of the right lateral ventricle, compressing the basal nuclei of the right side and excavating the medial part of the temporal lobe. The capsule of this tumor was partially calcified.

Scherer reported a spinal cord lipoma observed in a 42-year-old woman. It was 14 cm. in length and 2.7 cm. in width, extending from the cervical enlargement to the mid-dorsal level, where it tapered down and seemed to merge into the substance of the cord. A section through the cord revealed a large, obliquely oval fatty body superimposed in a dorsal direction upon a wedge-shaped compressed cord. The right posterior rootlet was enclosed within the lipoma, and for this reason the author believed that the tumor may have arisen from the pia in the region of this rootlet. The cord adjacent to the tumor was atrophic and contained small groups of fat cells.

REPORT OF A CASE

A white female, aged one year, was admitted to the University of Minnesota Hospital on Jan. 10, 1937, and died the following day.

The birth of the patient was attended with little difficulty; the head was only slightly enlarged with a defect in the skull, indicated by a protuberance of soft tissue over the left
temporal region. The infant was cyanotic and had a convulsion shortly after birth. Cyanosis and convulsions occurring as often as every half hour continued for six days, during which oxygen was continuously administered. The heart rate was rapid. A spastic paralysis of the right arm and leg was present, and congenital absence of the iris in each eye was observed.

The child was nursed at the breast for nine months, but had some difficulty in swallowing, and at no time manifested a normal reaction toward food.

At six months of age there developed a diarrhea, with elevation of temperature. Several attacks of diarrhea occurred during the next six months, lasting three to four days and on each occasion associated with fever. The head continued to enlarge and at ten months the circumference was 23 inches (58.4 cm.). The weight at this time was 16 pounds 10½ ounces (7516 gm.).

During the month prior to admission the temperature was elevated to 103° F. daily and the child cried almost continuously. Sedatives were without effect. Feeding became increasingly difficult, and during the three days prior to admission little food was taken.

**FIG. 1. PHOTOGRAPH OF THE CEREBRAL AND SPINAL LIPOMAS**

Note the large masses in the cervical region and their extension downward along the length of the cord. An opaque membrane covers parts of the inferior surface of the left cerebral hemisphere.

On admission the temperature was 105° F. and the child appeared to be in a stuporous state. The nutrition was fair, although there were signs of dehydration. The head was asymmetrically enlarged, quite typical of hydrocephalus, with prominent frontal and parietal protuberances. The fontanelles were large and irregular and separation of the suture lines was evident. Over the left temporal region was an area of decreased resistance which measured about $2 \times 5$ cm. The edges were raised and sharp and gave the impression of a bony defect. The left eye was turned inward, the tongue was displaced to the right side of the mouth, and drooling was marked. The chest was asymmetrical; the left side appeared to be flattened, the right somewhat bulging. The percussion note and breath sounds were normal, as were the heart sounds. The abdomen was negative. The right arm was spastic, with the fist clenched. The left arm moved in rhythmic fashion. The knee jerk was exaggerated on the right side. The plantar reflexes and the Chvostek and Kernig signs were negative. Fundus examination of the right eye revealed areas of patchy pigmentation, the discs being sharply outlined. The spine was abnormal, with kyphosis and scoliosis to the left. The skin showed a few small xanthomatous lesions, and there were what appeared to be abnormal pads of tissue about the right shoulder region.

The routine laboratory findings were not significant. X-ray examination revealed a thinning of the inner table of the skull with local erosion in the left posterior parietal region.
and a bulging of the skull at that point. Areas of rarefaction were present. The appearance was not characteristic of Schüller-Christian's disease, but suggested rather some local increase of pressure from within the cranium. The appearance suggested a hydrocephalus with the possibility of an intracranial tumor.

The temperature continued to rise, reaching 108° F. The patient had a series of convulsions with generalized twitchings, involving the face, arms, and lower extremities. The breathing became slow and irregular, and death occurred eight hours following admission.

Autopsy Findings: The occipitofrontal circumference of the head measured 51 cm. There was a bulging of the skull in the left parieto-occipital region near the midline, measuring $5 \times 8$ cm. In the left temporal region was a bony defect 2 cm. in diameter. The frontal fontanelle was much larger than usual, but there appeared to be no definite separation of the sutures. The iris was absent from the right eye and that of the left was defective. There were a few hemorrhagic areas in the conjunctiva and a pterygium on the medial surface of the left eye. The right upper eyelid contained a xanthoma 4 mm. in diameter, and the left lid showed three similar tumors. The left side of the chest was flattened and pushed toward the right.

Examination of the viscera was negative with the exception of the central nervous system.

There was a tremendous hydrocephalus involving primarily the left lateral ventricle. The brain tissue was thinned and distended to form a huge sac, being reduced in the parieto-occipital region to a few millimeters in thickness. The lateral surface of the left frontoparietal and inferior temporal regions was covered by a heavy, opaque, gelatinous membrane 3 mm. in thickness, extending upward into the Sylvian fissure to form firm adhesions between the temporal and frontal lobe (Fig. 1), and downward over the inferior surface of the cerebral peduncle. The right lateral ventricle was only moderately hydrocephalic, the dilatation being limited to the occipital horn. The third ventricle was distended; its floor was thinned and displaced downward and the optic tracts, as they encircled it, were displaced to the right. Both foramina of Monro were enlarged. The choroid plexus within the left lateral ventricle, close to the foramen of Monro, was enlarged and contained a yellowish irregular mass measuring $2 \times 1$ cm.

There was a fatty tumor $1 \times 1.5$ cm. in the left cerebellopontine angle, firmly adherent to the lateral surface of the pons and the anterior border of the cerebellum, which was...
indented by it. The fifth cranial nerve was completely enclosed within the fatty mass, its
fibers being compressed into a wide band, but retaining a fairly intact outline in its course
through the tumor. The left 7th and 8th cranial nerves were displaced downward and
passed around the posterior border of the new growth.

Two large tumor lobules were attached to the cervical cord at the level of the third to
the fifth segment (Fig. 1). The uppermost was the smaller, measuring $2 \times 2 \times 1$ cm. and
protruding from the right lateral border of the cord. Its caudal surface was attached to the
larger fatty mass which measured $3 \times 3 \times 2$ cm. Both were very firm and yellow.

From the inferior surface of the larger cervical tumor a column of fatty tissue 1.5 cm.
in diameter continued downward on the dorsolateral surface of the cord to the lower sacral
segments (Fig. 1). The cord was compressed in an anteroposterior direction and measured
but 4 mm. in diameter (Fig. 2). Numerous fine trabeculations seemed to pass from the
cord tissue into the fatty mass, dividing it into many elongated lobules. The cord tumor

![Image](https://via.placeholder.com/150)

**Fig. 3. An Area of the Lipoma Adjacent to the Spinal Cord**

A heavy membrane separates the two structures. Most of the fat cells appear mature and
inactive.

was covered by a fairly firm opaque membrane. The spinal rootlets, as they emerged from
the cord, passed laterally around the fatty tissue to reach the spinal ganglia, which were
situated on the lateral surface of the column of fat. In many cases small clumps of fat
followed the rootlets through the openings in the dura to form nodules alongside the spinal
ganglia (Fig. 1).

**Microscopic Examination:** The large tumor in the cervical region consisted entirely of
mature fat cells with distended cell bodies and peripherally disposed nuclei, producing the
characteristic signet-ring appearance. There were no blood vessels nor connective tissue
within this mass.

Sections through the smaller cerebellopontine angle tumor revealed a somewhat dif-
f erent picture. The tumor cells in the center and periphery were typical large adipose cells,
but as the brain tissue was approached, their nature changed. They became compressed
and smaller; the nuclei were more centrally placed and elongated, and had acquired in many
cases a definite cytoplasmic body. These cells increased in number and were very numerous
in the vicinity of the pons. In this region many small vessels were also observed. The
tumor was sharply demarcated from the cerebral tissue except for one small area where the
proliferating cells had broken into the pons but had not extended very deeply into its substance. The cerebral vessels adjacent to this lipoma were completely calcified.

The choroid plexus tumor was composed primarily of mature fat cells, but many cells seemed to be of a more embryonic character.

The spinal cord was compressed, the posterior horns being situated directly adjacent to the anterior. The nerve cells of the cord were not decreased in number, although they were injured, being irregular in shape and almost completely free of Nissl substance. There was a diffuse but equal demyelination of the white substance. The cord was separated from the lipomatous mass by a dense membrane. The tumor was composed principally of adipose tissue intermixed with smaller and apparently more actively growing cells. Radiating from the heavy lining membrane were numerous connective-tissue strands that subdivided the lipoma into lobules of varying size (Fig. 3).

The rootlets, prior to their emergence from the lipomatous tissue, were broken up into numerous small masses of nerve tissue and separated from one another by heavy fibrous bands. In the area of these fragmented rootlets, the membrane separating the cord from the tumor was deficient and numerous large fat cells had extended between the nerve bundles into the cord tissue just beneath the rootlet entrance. Fat cells were not found elsewhere within the cord.

**DISCUSSION**

One of the most striking observations concerning this tumor is its extensiveness. It involved the entire cord, the base of the brain, and the choroid plexus. From the literature it appears that there is a type of pial lipoma which characteristically involves the dorsolateral region of the cord, along the posterior rootlets, producing changes within the rootlets with fatty development and compression of the cord. The most frequent sites of spinal lipomas, however, are in the cervicodorsal and lumbosacral regions, with only localized cord involvement.

The association of lipomas of the central nervous system with developmental defects is a common observation. This occurs most frequently in spinal cord lipomas, where the tumor is associated with spina bifida. In the brain lipomas are not uncommonly associated with agenesis of the corpus callosum (Würth, 5; Huddleson, 6; Benjamin, 7; and Ernst, 8). Wolbach and Millet (9) in 1913 reported a case of lipomatosis of the spinal cord in which there was a congenital absence of the right kidney and ureter as well as a cleft palate and hare-lip. This case is of particular interest because of its close resemblance to the case here reported. It occurred in a male infant, ten months of age, in whom an autopsy revealed a lipomatous mass involving the spinal cord from the medulla to a few centimeters above the filum terminale. The cord in its greatest part (cervical) measured 2 × 1.5 cm. At the cerebellopontine angle were two whitish vascular nodules attached to the pia mater and measuring 1 × 1.5 cm. They were partially encapsulated by a thin band of connective tissue which was composed of the same material as the lesions in the medulla. Histologically the tumors were composed of mature fat cells and showed no signs of embryonic fatty tissue nor of rapid growth. In our case anomalies were also present. These consisted of lipomas of the skin, bilateral absence of the iris, and defects in the skull.

Virchow believed these tumors to be hyperplastic growths from fat cells normally found in the pia. Others have held that they develop, as a result of metaplasia, from embryonic remnants, either ectodermal or mesodermal in origin, which have been displaced during the formation of the central nervous
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system from the neural tube. Still another theory maintains that they arise by a formation of fatty tissue from the connective tissue present in the pia. Many investigators insist, however, that these tumors do not originate in adult connective tissue but are related to the development of the primitive layers of the meninges and hence have an embryonic mesenchymal origin. Scherer (4) more recently stated that lipomas probably develop from the connective-tissue apparatus of the newly formed blood vessels. He believes that in the formation of new vessels there also develops a mesenchymal reticulum. The fatty tissue in the lipomas originates through a proliferative change in this new mesenchyma.

The latter two views seem most tenable, especially since it is being recognized that new fat cells in the adult arise, not from fully differentiated fibroblasts, but from undifferentiated mesenchymal cells. Such cells in the pia, regardless of their origin, might give rise to these fatty tumors.

SUMMARY

A case of lipomatosis of the central nervous system is reported occurring in a one-year-old girl, with fatty tumors in the choroid plexus, the base of the brain, and the spinal cord.

A brief summary of the more recent literature on this subject is attempted.

BIBLIOGRAPHY