GIANT CELL TUMOR OF THE SPHENOID BONE
REPORT OF A CASE

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GIANT cell tumors of bone are most commonly found in the long bones but occasionally they occur in the skull and elsewhere. According to Wattles the bones affected are, in order of frequency, long bones, small bones, maxilla, mandible, vertebrae, ribs, scapula, clavicle and the ethmoid, sphenoid, temporal and frontal bones. From a search of the records in the surgical pathologic laboratory of the Johns Hopkins Hospital for a period of thirty-five years, Geschickter and Copeland found only 22 cases of giant cell tumor occurring in the skull. Two of these were found in the sphenoid bone and the remainder in the jaws. Details of the two tumors in the sphenoid bone were not given. Analysis of the available medical literature has failed to reveal any other cases of giant cell tumor of the sphenoid bone. This is one of the reasons that the following case is reported.

REPORT OF A CASE

G. G. a 12-year-old girl was seen by me on June 16, 1943 at the Baptist Hospital in New Orleans at the request of Dr. William H. Gillentine. About two weeks before, the child had completed the seventh grade at school at the head of her class. Shortly thereafter she began to vomit and complained of headache and double vision. The child's mother stated that the patient had been irritable for several months and that occasionally there was a one-degree rise in temperature. She had grown three inches in height in the past year and had not begun to menstruate.

Examination. A complete sixth nerve paralysis on the right was noted, the pupils were unusually large and the eyegrounds and visual fields were normal. The remainder of the physical and neurologic findings were negative.

Hematologic examination revealed a red blood cell count of 5,120,000, hemoglobin 97 per cent and white blood cell count of 7,350. Spinal fluid pressure was 140 mm. of spinal fluid. The total protein content was 32.4 mg. per cent. The gold-sol test was 2111100000. The Wassermann reaction was negative.
Roentgenograms of the skull (Fig. 1) revealed complete destruction of the sella turcica suggesting either a cerebellar tumor or a large adenoma of the pituitary gland.

Course. A tentative diagnosis of tumor destroying the sella turcica was made and the patient was discharged from the hospital June 17, 1943 for a period of observation at her home. She began to sleep much of the time during the day, vomited occasionally but experienced no further headaches. The visual fields, which were examined on June 24, 1943, showed central scotoma on the right. Visual acuity on the right was 20/50 and on the left 20/15.

Admission. On June 30, 1943 the patient was admitted to the Touro Infirmary. Two days before numbness of the left cheek had developed and vision in the right eye had begun to fail. An encephalogram made under intravenous anesthesia revealed a normal ventricular system, but a rounded mass was seen in the region of the sella turcica which had elevated the cisternae chiasmatica and interpeduncularis (Fig. 2). The impression was that the patient had an atypical pituitary adenoma, an uncalcified craniopharyngioma or a teratoma of the pituitary region.

Operation I. On July 3, 1943, under local anesthesia, exposure of the optic chiasm revealed that the diaphragma sellae was in contact with the optic nerves and chiasm but it was not sufficiently elevated to flatten the optic nerves or to separate them. The diaphragma was coagulated and the sucker was pushed through it. A large cavity occupied the region of the sella turcica. The fluid contents of the cavity were aspirated by the sucker. A portion of tissue from the tumor adhered to the sucker and this was sent to the pathologist for histologic diagnosis. From the hole in the diaphragma there was brisk, persistent bleeding which was controlled by plugging the hole with a piece of temporal muscle. The dura was closed with cotton and then sutured to the periosteum. The bone flap was laid in place; the scalp was closed in two layers with cotton.

Pathological Report. Dr. J. R. Schenken made a diagnosis of giant cell tumor of bone (Fig. 3). This diagnosis was also made by Dr. Granville Bennett of the Tulane Medical School.
Course. The patient was discharged from the hospital July 17, 1943. Shortly after operation she gradually became completely blind in the right eye and paralysis of the third nerve appeared. Dilation of the left pupil followed.

Light roentgen therapy was then given by Dr. Meyer Teitelbaum: 1800 R to each of three ports, the daily dosage being 300 R alternating between the three fields. This was done by means of a small cone attached to a roentgen-ray machine at a target distance of 70 cm.

Fig. 8. Low and high power views of the tissue removed from the tumor at the first operation.

About Sept. 15, 1943 vision in the left eye began to fail. The child continued to grow progressively worse. She lost considerable weight and complained of abdominal pain and nausea in addition to headache and numbness of the face. Hypodermic injections of pituitary extract were given for several weeks without improvement in the symptoms.

In September 1943, Dr. George T. Pack and Dr. W. H. Stewart of New York City were consulted by mail. They agreed that the growth presented the microscopic appearance of benign giant cell tumor and suggested heavy doses of irradiation. This was carried out but without apparent effect upon the rate of growth of the tumor.

Roentgenograms of the skull on Jan. 7, 1944 showed a considerable increase in the destruction of the floor of the skull (Fig. 4). Examination with the pharyngoscope on this date showed pronounced bulging of the posterior wall of the nasopharynx. By this time the patient was completely blind but not confined to bed.

As the child continued to get worse, her parents conferred with various surgeons throughout the country regarding her condition.

Operation II. On March 11, 1944 in St. Louis, she was operated on by Dr. Roland M. Klemme. The operative note, which he was kind enough to send to me, is briefly abstracted. Following anesthetization with avertin, 0.5 per cent solution of procaine hydrochloride and a few drops of ether, a rather large unilateral right frontal flap was turned down, part of the previous flap being utilized. Elevation of the brain, extradurally, uncovered an enormous tumor completely filling the floors of the frontal and temporal fossae. With the electro-surgical knife, the tumor was excised from the bone and the entire temporal fossa was emptied as well as most of the frontal fossa along the greater wing of the sphenoid. All bleeding points were
controlled with the electrosurgical knife; little bleeding occurred. It was immediately ap-
parent that the optic nerves and chiasm were completely involved, so the right optic nerve
was dissected out in its entirety. The posterior third of the bony orbit of the eye had been
completely eroded. All the tumor around the globe of the eye was removed. The incision was
then carried forward and the tumor, extending down into the sphenoid and both antra to the
soft palate, was excavated. After the optic chiasm and the nerves had been freed, the
sheath of the optic nerve was split on the right, and the nerve lay perfectly free and
seemed to return to its normal size. The one on the left was so constricted that this pro-
cedure had little effect. The balance of the tumor was removed piecemeal and there
was a lot of bone in conjunction with the tumor. Erosion of the petrous tip of the
temporal bone could be seen on the right side and the same condition appeared to ex-
ist on the left. The floor of the temporal fossa was completely eroded and the soft
palate could be felt. The posterior third of the bony orbit of each eye had been com-
pletely eroded, and the sphenoids and ethmoids were gone. About two-thirds of the
clvius was completely eroded and the tumor was removed from this area in its
entirety. At the end of the procedure the optic chiasm and both optic nerves hung
suspended in mid air unsupported from the base of the brain to the globes of each eye.
The patient's condition at the end of the operation was excellent. The blood pressure
was 120/85, pulse around 90 and respira-
tions were normal.

Course. The child died forty-eight hours after operation.

Autopsy. Postmortem examination was made by Dr. L. S. Walsh. Only the part of his
findings concerning the lesion will be given. The brain weighed 1300 grams. The basal arach-
noid and vasculature were fixed to a mass which displaced cerebral tissue superiorly and in-
dented the floor of the third ventricle and optic chiasm. Gross coronal sections of the fore-
brain revealed nothing unusual. Gross cross sections of the stem showed scattered subarach-
noid clots and two foci of intramedullary hemorrhage each 2 mm. in diameter.

The tumor extending from the base of the brain involved both petrous ridges, the dura
along the posterior aspect of the left petrous bone, both middle fossae with complete erosion
of the floors, the medial halves of both orbital plates, the ethmoid bone, sphenoid bone and
nasal concha. The tumor reached the palate anteriorly and the pharynx posteriorly and ex-
tended along both carotid canals and jugular foramina to the retropharyngeal regions.

Histology. The cytologic examination of the specimen showed that it consisted of fibro-
blastic matrix in which enormous numbers of giant cells were found. There were spicules of
bone scattered here and there. The diagnosis was giant cell tumor.

COMMENT

This case presents several interesting features. The rarity of occurrence
of giant cell tumors has already received comment. Most giant cell tumors
of the skull are in the mandible. However, one case in the sphenoid bone$^9$ and
two in the frontal bone\textsuperscript{1,2,7} have been reported in addition to the two cases involving the sphenoid bones briefly mentioned by Geschickter and Copeland,\textsuperscript{4} which have already been referred to in the opening paragraph.

Another interesting aspect of this case is the fact that the sella turcica was completely destroyed by the growth. To the neurosurgeon, destruction of the sella turcica indicates either obstructive hydrocephalus caused by a tumor in the cerebellar fossa or a large adenoma of the pituitary gland. The possibility of primary tumor of bone was not even considered in this case by the roentgenologist or the surgeon.

Additional points of interest are the lack of a history of trauma and the youthfulness of the patient. Although an occasional case of giant cell tumor of bone has been reported in children, it is generally seen in adults.

The genesis of giant cell tumors has long been a subject for speculation. Probably the most plausible theory is the one expounded by Geschickter and Copeland,\textsuperscript{4} who believe that these growths arise from resorptive processes in cartilaginous bone. However, this hypothesis fails to explain the occurrence of such a tumor in the frontal bone (membranous bone), as pointed out by Keegan and Baker.\textsuperscript{7}

That the "benign" giant cell tumor sometimes proves to be malignant in its behavior is well known.\textsuperscript{1,3,5,6,8} However, even after the tendency of this tumor to destroy the base of the skull became known, there was nothing about its microscopic appearance (Drs. Stewart, Bennett and Schenken) to indicate that it differed from the giant cell tumors that have a truly benign course.

REFERENCES

2. Fraser, J. Cited by Keegan and Baker.\textsuperscript{7}