PRIMARY TUMORS OF THE CRANIAl BONES

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New growths originating in the cranial bones are uncommon, although the skull may be affected secondarily by benign or malignant conditions arising in other organs. The most common primary tumor in this region is the benign osteoma, which usually involves the frontal bones or frontal sinus. Rarefied areas in the roentgenogram, either solitary or multiple, may be due to angiomatous growths in the bones of the cranium or in the intracranial cavity, to cholesteatomas or to a Brodie’s abscess. Such circumscribed areas of bone destruction are more often, however, part of a generalized skeletal disease, such as parathyroidism or Schüller-Christian’s disease (xanthomatosis).

Both osteogenic and Ewing’s sarcoma may affect the cranial bones. Malignant conditions producing multiple involvement of the skull along with other bones are more common. These include multiple myeloma, chloroma, and leukemia. The skull may also be involved by direct extension in cases of meningeval and pituitary tumors and in carcinoma originating in the antrum, frontal sinus, or orbit.

Metastatic involvement of the skull occurs commonly, the breast, prostate, or lung being the usual seat of the primary tumor. Less frequently sarcoma from other bones (Ewing’s sarcoma), lymphosarcoma, or neuroblastoma may affect the cranial. The following table gives the incidence of the primary tumors of the skull in the series upon which this paper is based.

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Benign Tumors

Osteomas

In the membranous bones of the skull and jaws, benign osseous tumors may occur as a result of the production of new bone in ossify-
ing fibrous tissue. The more rapidly growing osteomas are predomi-
nantly fibrous tumors with small osteoid or osseous spicules em-
bodied in proliferating connective tissue. Some of these growths are
composed largely of spongy bone, while those of the more highly dif-
ferentiated group are formed of dense compact bone.

Of 85 tumors classed as osteomas in the surgical pathological lab-
oration of Johns Hopkins Hospital, 30 were in the region of the upper
jaws, 19 were antral or intranasal, 21 involved the lower jaws, 4 the

frontal sinuses, and 2 were intraorbital. Nine occurred on the external
surface of the cranium, in the region of the frontal bones; one was in
the mastoid region and another at the occiput. An equal number occu-
pied the inner tables of the skull or dura, producing definite intra-
cranial symptoms. Several of these were unattached to the skull and appar-
ently were dural tumors.

Osteomas involving the membranous bones differ histologically from
osteoendromata, which may occur in those areas of the skull preformed
in cartilage, and from the osteophytes produced in membranous bone
invaded by meningeal tumors.

The typical cranial osteoma is a benign, mound-like swelling occur-
ring in the frontal region in a young adult. Of the 11 patients in the
present series, 9 were between the ages of eighteen and thirty-three

Figs. 1 and 2. Osteomas of the Skull

The boy at the left was thirteen years of age, with an osteoma of the frontal bone. It
was successfully removed, leaving the inner table intact. The patient at the right, a boy of
eighteen, had a large osteoma at the vertex of the skull. There was a history of severe trauma
in this region fourteen years earlier, and a tumor had been present since the age of seven.
Fainting spells and convulsions had occurred during the past two years. Path. Nos. 56628
and 56632.
years. The youngest was thirteen years of age and the eldest seventy years. The average duration of the tumor was eighteen years and 8 of the patients attributed its onset to a severe blow or fall.

Echlin has previously reported 7 of the cases in the present series, which were treated on the neurosurgical service of Dr. Walter E. Dandy, together with a group of 19 osteomas of the outer surface of the cranium collected from the literature. A hard, immovable, painless swelling on the surface of the skull was the first sign noticed by all of these patients (Figs. 1 and 2). Sixteen of the 26 cases involved the frontal bone. The parietal or temporal bones were involved in 8 cases and the occipital in 2. The growth of the tumors was slow and progressive, but in several instances acceleration occurred following trauma or puberty. In the late stages, intracranial symptoms such as dizziness, headaches, and epileptic seizures were noted. The majority of the tumors dated back to childhood, although the patients at the time of clinical examination were adults. The usual size of the tumor at examination was between 3 and 10 cm. in diameter.

The roentgen findings are characteristic (Figs. 3, 4 and 5): a dense
Fig. 5. Roentgenogram of a huge spongy osteoma of the mastoid region
The patient was a child of twelve. Path. No. 53574.

Fig. 6. Roentgenogram of osteophytes occurring in a case of meningeal tumor
The patient, a man aged forty, had received a blow in this region fifteen years previously.
He lived four years after the appearance of intracranial symptoms. The nature of the tumor
was verified at autopsy. Path. No. 45782.
mass of new bone with a smooth convex outer border and a wavy sharply demarcated base formed by the thickened or slightly depressed inner table of the skull. In the region of the outer table is a border of decreased density over the surface of the growth, which represents the growing margin of the tumor. The tumor appears to be a continuation of the diploe.

Osteomas of the cranial surface must be differentiated in the roentgenogram from the hyperostoses resulting from meningeal tumors invading the skull. In these lesions the new bone formation shows more definitely radiating spicules which extend at right angles from both

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

**FIG. 7. PHOTOMICROGRAPH OF A SPONGY OSTEOMA SHOWING LARGE AMOUNTS OF FIBROUS TISSUE AMID BONE SPICULES**

The tissue was removed from the patient shown in Fig. 2. From Echlin, F.: Arch. Surg. 28: 372, 1934.

the inner and outer tables of the skull. The persisting and rarefied tables of the skull can be traced through the center of the radiating osteophytes (Fig. 6).

The osteomas are composed of spongy or eburnated bone. The outer margin is overlaid by fibrous tissue which fuses with the surrounding periosteum, and which in the spongy osteomas is often abundant. The bony layer is continuous with the diploe or may reach the inner table. In the eburnated osteoma there is a layer of cortical bone beneath a capsule of fibrous tissue; the tumor is usually small with its base either on the outer or inner table. The larger osteomas are of the spongy type and show cancellous bone extending to the inner table.
Microscopically, the osteomas exhibit a layer of cortical bone under-lying a zone of connective tissue. Beneath this cortical bone are tra-beea1ae of cancellous bone varying in vascularity and in the amount of intervening fibrous tissue. The eburnated osteomas have adult qui-escent lamellae of bone; the spongy osteomas, on the other hand, show cellular, vascular connective tissue separating spicules of newly formed bone surrounded by more or less orderly rows of osteoblasts (Figs. 7 and 8).

The growth of the osteomas seems to follow the physiology of ossification in membranous bone. Normally the skull increases in thickness by deposition of bone superiosteally, the latter being formed directly from preosseous connective tissue. The persistence of the overlying periosteal layer and the increase of fibrous tissue in this region (Fig. 9) in growing osteomas suggest that the tumor is a result of ossification proceeding from the periosteal or subperiosteal region of the skull. Eehlin in his study was impressed with the fact that 50 per cent of the tumors in his series arose before the eighth year of life and 75 per cent before the twelfth year. He also emphasized the relatively high frequency of osteomas in the frontal or facial bones. This age inci-
idence and distribution he related to the persistence of normal growth in the frontal and facial bones, which continue to increase in size through puberty, after many of the other bones of the skull have ceased to grow. To this latter phenomenon the increased size and more pronounced clinical features of osteomas in these regions may be attributable.

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

**Fig. 9. High-power Photomicrograph of the Surface of a Spongy Osteoma, Showing the Formation of New Bone from Fibrous Tissue**

Pathologically the osteomas of the skull are of special interest from the standpoint of both normal bone growth and tumor formation. As can often be clearly demonstrated, the compact bone surmounting the tumor is laid down only in the presence of vascularization and resorption of underlying cancellous bone. Apparently the compact upper portion of the tumor immediately below the periosteum grows at the expense of the cancellous substance beneath. Although it is usually stated that membranous bone originates directly from fibrous tissue, it is thus seen that compact cortical bone is not formed directly but only
in conjunction with the resorption of pre-existing cancellous bone. In this respect the membranous bones resemble the intracartilaginous bones, in the development of which both the resorption of calcified cartilage and the reconstruction of cancellous tissue precede the appearance of the final cortical and cancellous structures. Nevertheless, osteomas are not found in the intracartilaginous bones.\textsuperscript{1} Here the benign bone tumors show an intervening zone of cartilage between the overlying precartilaginous connective tissue and the underlying bone. Since the final stages of cortical bone formation are similar in both membranous and intracartilaginous bone, it follows that these benign tumors, the osteochondromas and the osteomas, must proceed from the more undifferentiated or embryonic portions of the persisting connective tis-

\textbf{Fig. 10. Photograph and Roentgenogram of Intracranial Hyperostoses}

The new bone is formed from the inner table. Path. No. 56034. From the Pathologic Museum of Bellevue Hospital, New York.

sues, for in the histogenesis of these lesions the entire cycle of intracartilaginous or intramembranous bone formation is repeated, rather than merely the end-stages.

\textit{Intracranial Hyperostoses and Osteochondromas}

Hyperostoses of rounded or irregular contour resembling osteomas may project from the inner table of the skull in the anterior portion of the calvarium. The frontal and parietal bones are usually involved diffusely. According to Yolton, this condition is observed more frequently in women than in men and is a compensation for atrophy of the brain. It is usually found only at autopsy in adults. The compact or spongy new bone projects inwardly from the inner table without any evidence of change in the outer table (Fig. 10). Microscopically the new bone formation is similar to that in the spongy and chondrified osteomas.

In the long bones the most common benign tumors are osteochondromas. Of more than 300 of these recorded in this laboratory only 3 involved the skull, all in the region of the occiput. Two of the patients

\textsuperscript{1}Jaffe has recently described osteoid osteomas occurring in the medullary cavity of the intracartilaginous bones. The lesions are small and do not exceed 2 cm. in diameter. Clinically they resemble Brodie’s abscess.
Fig. 11. Roentgenogram of an Osteochondroma Arising from the Basi-occipital Bone Near the Attachment of the Trapezius Muscle


Fig. 12. Photomicrograph of an Osteochondroma of the Occipital Bone. Path. No. 15977
were adults and the third was aged fifteen. All three patients have remained well for more than five years, although one of the lesions was interpreted as probably malignant from the roentgenogram (Fig. 11) and in another a diagnosis of chondrosarcoma was made from the sections (Fig. 12). Since the basioccipital bone is preformed in cartilage, osteochondromas of this region do not differ from those of the long bones.

Angiomas

Angiomas of bone, which may be either of the cavernous or capillary type, are found more often in the skull or spine than in the long bones. Of 12 angiomas of bone recorded in this laboratory, 5 involved the skull. Bucy and Capp reported, among 8 cases of primary hemangioma of bone, a cavernous angioma of the parietal bone, operated on by Cushing. In their review of the literature they found 11 angiomas of the skull among 56 cases. The majority of these involved the frontal or parietal bones and occurred in adults.

In the case recorded by Bucy and Capp the roentgenogram showed a fine honeycombed area of rarefaction and radiating spicules of bone overlying the tumor. Two cases in our series produced cyst-like expansion of both tables, and little or no apparent increase in the size of the lesion was observed over a period of years. In one instance a congenital angioma was excised from the scalp and skull of a girl aged seventeen months. The inner table of the skull was intact. In two cases previously reported by Dandy the skull, dura, and brain were involved. One of the lesions was described by Dandy as a venous aneu-
Fig. 14. Roentgenogram of a Giant-cell Tumor Eroding the Body and Wing of the Sphenoid Bone

The lesion occurred in a woman aged fifty-two, who is living, but with hemiplegia, nine years after operative removal. Path. No. 37914.

Fig. 15. Roentgenogram of a Benign Cystic Defect in the Skull in a Male Aged Twenty-six

A swelling had been present in the region since childhood. The lesion has not changed in the past three years, during which it has been under observation. The probable diagnosis is unresolved hematoma. Path. No. 44198.
Fig. 16 A. Roentgenogram showing a Multilocular Cystic Defect in the Temporal Region Produced by a Cholesteatoma. Path. No. 56636

Fig. 16 B. Photomicrograph showing the Epithelial Lining of the Tumor in Fig. 16A
rysm arising in the longitudinal sinus and the other as a cavernous angiomia in the posterior cranial fossa and extracranial occipital region (Fig. 13).

Giant-cell Tumors of the Skull

A study of over 300 giant-cell tumors in various skeletal and extra-skeletal locations indicates that there is an intimate relation between pathologic giant-cell proliferation and the physiological processes of absorption in intracartilaginous bone. With the exception of the giant-cell epulis, which forms around the roots of deciduous teeth, giant-cell tumors are usually confined to the intracartilaginous bones or to the sesamoid bones in the tendon sheaths derived from fibro-cartilage. In the skull, therefore, they should be restricted to those regions which develop from the chondrocranium.

This fact is borne out by the 4 giant-cell tumors of the skull in the present series. Two of these presented in the temporal fossa, arising from the sphenoid bone, one occurred in the mastoid region and involved the auditory meatus, and the fourth developed in the occipital region. The ages of the patients at the time of the initial examination were fourteen, forty-two, fifty-two, and seventy years. Although in three of the four cases there was a recurrence after excision, none of the patients died of tumor. Excision of the lesion in the temporal fossa resulted in a cure in the youngest patient, who is reported well twenty years later. The tumor in the body of the sphenoid bone (Fig. 14), occurring in a white female aged fifty-two years, apparently caused hemiplegia four years after operation. The patient is still living with spastic paralysis nine years later. The tumor in the mastoid region

![Image](image.jpg)

Fig. 17. Roentgenogram of Osteomyelitis of the Parietal Bone

Note the small, dense, rounded sequestrum at the lower posterior portion of the lesion. Path. No. 48466.
invaded the cranial cavity and at the third operation a portion was removed from the brain substance; the patient, however, is free of symptoms five years later.

Roentgenographically giant-cell tumors of the skull produce a sharply demarcated area of bone destruction. Microscopically they do not differ from giant-cell tumors of the long bones. We have been able to find only 2 cases of giant-cell tumors of the skull in the recent literature: one reported by Troell, the other by Fraser.

Troell's patient was a man of twenty, and the tumor had supposedly been present from birth. At the age of three it was excised and reported as giant-cell sarcoma. At the age of twenty it was about the size of an adult fist and was encased in a shell of bone. The tissue removed at this time showed only a bone cyst containing blood, cholesterol, plasma cells, and lymphocytes. This case demonstrates that a healing giant-cell tumor may be converted into a benign bone cyst. Fraser's case, occurring in a man of forty-two and of five months' duration, is of interest because of its location in membranous bone. The tumor was removed from the left half of the frontal bone following deep x-ray therapy. Microscopic examination showed blood spaces surrounded by giant cells. It is possible that this lesion was a post-traumatic vascular defect with giant-cell osteoclasts. Such a histologic picture may also be seen in inflammatory lesions of the diploe (Fig. 18 C).

Cystic Lesions of the Skull

Isolated defects in the cranial bones may be observed in roentgenograms of the skull in young adults following the discovery of a mass overlying a membranous bone (Fig. 15). There may be a history of
trauma or infection, or the mass may date from early childhood. The lesions are practically asymptomatic. Such defects are often classed as bone cysts and even after microscopic examination of the cyst wall and contents are diagnosed as xanthomas or cholesteatomas.

Cholesteatomas: Cushing restricts the term cholesteatoma to epithelial-lined cysts which may arise in the leptomeninges at the base of the skull, in the infundibula or cerebellar regions, or in the diploe, usually in the region of the petrous or squamous portion of the tem-

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

**Fig. 18 C. Photomicrograph of Tissue Removed at Operation in Case Shown in Figs. 18 A and B.**

Vascular areas such as this, surrounded by foreign body giant-cells, were numerous. Other areas showed lymphocytes, foam cells, and blood pigment, suggesting a diagnosis of xanthoma.

poral bone. They may also occur in the walls of the ventricles. Pathologically dermal and epidermal types are recognized, the dermal types enclosing epithelium and hair, the epidermal type having a transitional squamous-cell lining. Cravathier in 1829 described the dermoid type under the name pearly tumor, and Johannes Mueller in 1838 described the epidermal form, reporting three cases, one of which involved the diploe of the skull. He introduced the term cholesteatoma because of the cholesterol crystals found in the cyst contents.

Cholesteatomas involving the diploe are less common than those found in the leptomeninges. They produce clearly demarcated areas of bone resorption which may be multilocular, eroding the inner table
FIGS. 19 A AND B. FIBROSARCOMA OF THE PARIETAL BONE

The roentgenogram shows a circular defect. The photomicrograph shows a low-grade fibrosarcoma probably arising from the outer layers of the periosteum. The patient was a white female, aged thirty-four, who had first noticed a painful mass accompanied by headache two and a half months prior to operation. She died five years after excision of the tumor, from intracranial extension. Path. No. 31267.
and thinning the outer table. The margin of the growth has a wavy or lobulated outline. The lesions are most commonly found in the temporal bone (Fig. 16), but Cushing was able to collect 8 cases, including a case of his own, of epidermal cysts of the diploe, 3 of which were in the frontal, 2 in the parietal, and 3 in the occipital bone.

Microscopically the cysts are seen to be contained within a fibrous and epithelial wall under a thin external table of bone with a few adherent plaques representing the remains of the internal table. The cyst contents may be rich in cholesterin crystals. The epithelial lining shows five or more rows of transitional epithelium resting upon a wall of vascular connective tissue. The tumors apparently arise from epithelial implants carried into the bone during embryonic development.

![Fig. 20 A. Osteolytic Sarcoma of the Parietal-Occipital Region](image)

The patient was a white female, aged twenty-two, who had noticed a mass accompanied by headaches three years previously. The tumor at the time of examination was about the size of an orange. The roentgenogram shows an irregular area of bone destruction at the junction of the parietal and occipital bones. Path. No. 45834.

**Infections and Traumatic Defects:** Typical osteomyelitis of the cranial bones, with irregular new bone production, bone destruction, and small rounded sequestra, is easily recognized in the roentgenogram (Fig. 17). Such lesions are usually secondary to infections of the scalp, sinusitis, mastoiditis, or brain abscess. Syphilitic osteitis is not uncommon in the frontal and parietal bones. Confusion with primary tumors of the skull, however, may be occasioned by solitary defects of inflammatory origin (Fig. 18). Such areas of bone erosion in the present series have been observed in 12 instances several weeks after the occurrence of laryngitis, salpingitis, phlebitis, or septicemia. A palpable fluctuant tumor may be found overlying the osseous defect and aspiration or incision yields fluid mixed with granulation tissue or granulation tissue alone. Microscopic examination fails to reveal an
Figs. 20 B and C. Low-power and High-power Photomicrographs of the Tumor Shown in Fig. 20 A. Suggesting a Histologic Variant of Giant-cell Tumor

The high-power photomicrograph shows malignant cells in the stroma. The roentgenogram and the location in membranous bone were against the diagnosis of benign giant-cell tumor. The patient died fifteen months after excision of the tumor with metastasis to the lung, confirming the diagnosis of osteolytic sarcoma.
epithelial lining. The granulation tissue shows plasma cells, giant cells, lipoid-laden phagocytes, and other wandering cells. Because of involvement of the diploe the tissue may be quite vascular. Such granulation tissue is non-specific and, although sometimes interpreted as xanthoma or Schüller-Christian's disease, as osteitis fibrosa, or even giant-cell tumor, it is the result of an infection similar to Brodie's abscess of the long bones.

Defects without an epithelial lining and containing granulation tissue intermingled with blood and blood pigment may result from hemorrhage following trauma. The defect is surrounded by thin sclerotic bone, both tables of the skull being intact. Chorobski and Davis have reported such lesions of the skull being intact. We have observed two similar cases.

In rare instances osteitis fibrosa cystica accompanied by areas simulating giant-cell tumor may affect the membranous bones without demonstrable involvement of the remainder of the skeleton. Chorobski and Davis have attempted to collect such cases from the literature. The one case observed by these authors personally showed not a solitary defect but multiple recurring lesions of the cranium. Studies of the blood calcium and blood phosphorus were not made. In our own series of solitary bone cysts, approximating 200 cases, we have never observed such a solitary lesion in the membranous bones of the skull.

**Primary Sarcoma of the Cranial Bones**

Sarcoma primary in the cranial bones is rare. In a series of more than 500 cases of primary bone sarcoma, the upper and lower jaw were involved in 26 cases, but the cranial bones in only 12. With one exception the patients were children or young adults. Practically all of the cases occurred in the vault of the skull, in the region of the parietal bone, sometimes at its junction with the occipital or temporal. Eight of the 12 cases were varieties of osteogenic sarcoma, 1 was a fibrosar-
FIG. 22. OSTEOLYTIC OSTEOGENIC SARCOMA ARISING IN THE OCCIPITAL-PARIETAL REGION

This is the most common site for osteogenic sarcoma of the skull. Path. No. 40970.

FIG. 23. SCLEROSING OSTEOGENIC SARCOMA ARISING FROM THE POSTERIOR PARIETAL REGION IN A GIRL OF SIXTEEN

The inset in the lower left-hand corner shows the appearance of the patient. She had noticed swelling and pain in this region for eight months. Death occurred six months after excision of the tumor. Deep x-ray and radium therapy were without benefit. Path. No. 27325,
coma apparently arising from the outer layers of the periosteum (Fig. 19), and 3 were Ewing's sarcoma.

**Osteogenic Sarcoma**

Osteogenic sarcoma of the skull may be of the sclerosing or chondrosarcoma type. The osteolytic variety occurred twice in the present series (Fig. 20). The discovery of the growth may be preceded by headaches. The tumor increases rapidly in size but in general the duration prior to examination is longer in the skull than in other regions. All forms of osteogenic sarcoma in the present series showed

![Image](image.png)

**Fig. 24. Photomicrograph of a Chondrosarcoma Arising at the Base of the Anterior Fossa of the Skull.**

Path. No. 56038. Courtesy of Drs. Stevenson and Symmers, Bellevue Hospital, New York.

a tendency to destroy the tables of the skull as well as to produce a periosteal reaction and to invade the overlying soft parts (Figs. 21 and 22). Despite this destruction of bone, all but one of the patients in the present series died of metastases rather than of intracranial extension. In one patient there were metastases to the spine and ilium in the terminal stages of the disease. The roentgen diagnosis is based upon bone destruction in the tables and diploe, the periosteal reaction in the adjacent areas, and, in the late stages, extension of the tumor into the soft parts. In some instances, however, the nature of the lesion can be determined only by biopsy. Neither excision nor irradiation proved beneficial in the present series.

Microscopically, the predominant feature may be new bone formation with irregular spicules of new bone surrounded by proliferation of malignant osteoblasts. This is the sclerosing form (Fig. 23).
FIGS. 25 A AND B. Osteolytic Sarcoma Developing in Paget’s Disease of the Skull

FIG. 25 C. Photomicrograph of Sarcoma Shown in Figs. 25 A and B
the osteolytic tumors large malignant osteoblasts and small giant cells of the epulis type are predominant (Fig. 20). Adult and fetal cartilage may occur in osteogenic sarcoma at the occiput.

A case of typical chondrosarcoma (Fig. 24) involving the base of the skull was seen on the pathological service of Bellevue Hospital and is included here through the courtesy of Dr. Louis Stevenson and Dr. Douglas Symmers.

The patient was a white female aged sixty-four, who had noticed progressive exophthalmos for a period of three years. There was loss of vision in the left eye, with dimness of vision on the right and bilateral proptosis. The sense of smell was lost and breathing through the nose was difficult. Five months prior to operation convulsions first occurred. The roentgenogram showed destruction of the nasal portion of the right orbit extending into the ethmoid cells and the sphenoid bone. Numerous calcified deposits were seen in the brain tissue and anterior fossa near the midline.

![Fig. 26. Photomicrograph of Ewing's Sarcoma Arising in the Occipital Bone. Path. No. 49672](image)

On May 22, 1930, operation was performed for a tumor at the base of the anterior fossa, eroding through the palate and extending into the sinuses and pharynx. Excision was made above the eyebrows and the frontal bones rongeured away, down to the roof of the orbits. The frontal sinuses were enucleated. The bone at the back of the anterior fossa was soft and necrotic and adhered to the overlying tumor. The frontal lobes above the tumor tissue were thin and degenerated. The tumor was shiny and lobulated, invading the bone but not the brain substance. The ethmoidal and sphenoidal sinuses were exposed at the removal of the tumor.

The patient died postoperatively. At autopsy a portion of the tumor tissue left behind at operation was found attached to the region of the pituitary and the left middle fossa, bulging like a mushroom from the bone beneath. It was about the size of a lemon. The tumor was lobulated and pearly white, with translucent cartilaginous areas. It was firmly attached to the body of the sphenoid, obliterating the sella turcica. Its microscopic features are shown in Fig. 24. Anteriorly the mass removed at operation had eroded the frontal sinuses, the bony structures of the nose, the walls of both orbits, and the roof of the mouth.
Ewing's Sarcoma

There were 3 cases of Ewing's sarcoma in our series, 2 involving the mastoid process and 1 primary in the frontal bone. Two of the cases were in children and one in a young adult, aged twenty-two. The two tumors in the mastoid region (Fig. 26) were excised, and as far as could be determined both patients died of recurrence rather than metastases. The third tumor, which involved the frontal bone, destroyed practically the entire anterior half of the vault. The upper ribs, clavicle, and left tibia were also involved in this case. Despite the immense size of the cranial tumor, it is possible that the lesion of the skull was a metastasis from a primary focus in the tibia.

Fig. 27 A. Chordoma Invading the Occipital Bone. Path. No. 36943

Chordomas

Chordoma is a rare malignant neoplasm found at either extreme of the spinal axis. It is generally accepted as originating from embryonic remnants of the notochord. Although in the adult remnants of the notochord persist in the intervertebral discs in the form of the nucleus pulposus, the majority of chordomas arise either in the sphenoid-occipital or sacrococcygeal region. They have also been reported in the cervical spine and rarely in the dorsal or lumbar region. Owen and his associates estimate that approximately 100 cases have been recorded to date.

Coenen found 36 cases in the sphenoid-occipital region and 25 having a sacrococcygeal location. In this laboratory there are 5 cases recorded. One occurred in the occipital region, one in the cervical spine, and 3 in the sacrococcygeal region.

The cranial chordomas are most common at the sphenoid-occipital synchondrosis or at the clivus blumenbachii behind the sella turcica. They may destroy bone and produce symptoms of dyspituitarism and intracranial pressure. In only rare instances is the tumor encapsulated and found totally within the bone. Usually it extends to the soft
Fig. 27B. Photomicrograph of tumor shown in Fig. 27A

Fig. 28. High-power photomicrograph of the tumor shown in Figs. 27 A and B

The tumor cells are extremely variable in size and are undergoing vacuolization.
parts, both intracranially and extracranially. The one case in our series (Fig. 27) occurred in a male adult with signs of intracranial pressure following an injury to the back of his head. The tumor invaded the base of the occipital bone with intracranial and extracranial extensions. The tumor cells were variable in size, with the usual vacuolated cells and so-called physaliphorous nuclei. Adson has recently reported 2 cranial chordomas, one involving the sphenoid bone in a woman aged twenty-nine, suffering from diplopia and frontal headaches, and the other in a boy of eight with vomiting, indistinct speech, and listlessness. The child had suffered a blow to the occipital region seven months previously. The tumor passed through the foramen magnum.

These tumors are locally invasive and tend to recur following removal. In rare instances metastases occur. There is nothing characteristic in the bony erosion produced, and the diagnosis is made most frequently at autopsy.

Microscopically chordomas are distinguished with difficulty from atypical chondromas and mucoi d or signet-ring cell carcinomas arising in the gastro-intestinal tract. Myoblastomas originating in the sacrococcygeal region are usually interpreted as chordomas. The chordoma cells are more epithelial in appearance than cartilage and more variable in size. Evidences of calcification are absent, and there is a great tendency for vacuolization and variability in the size of the nuclei (Fig. 28).

Bibliography


