Surgical Pathology of the Nervous System and Its Coverings

Second Edition

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Preface to the Second Edition

We have been gratified by the response to the first edition of this book and are especially appreciative of favorable personal comments and constructive criticisms. Accordingly, we have retained the anatomic approach to the diagnosis of neurologic disease. In the second edition, we have focused our efforts on updating and refining the existing text in light of recent knowledge, while adding descriptions of entities not previously discussed. Throughout, additional emphasis has been placed on differential diagnosis, including that used at the time of frozen section. The illustrations have been evaluated carefully; some have been deleted and many new ones acquired. Among the latter are electron micrographs that have been added in accord with their practical and conceptual contributions.

The preparation of this second edition has been a pleasure. Our efforts will be doubly rewarded if the text continues to find acceptance as a practical diagnostic aid.

PETER C. BURGER
F. STEPHEN VOGEL

Preface to the First Edition

This book has been prepared for pathologists, clinicians, radiologists, and medical students who are confronted by the diversity of neurological disorders which are diagnosed and/or treated by surgical biopsy or excision. To assist in the recognition and understanding of the specific entities within this formidable array of diseases, we have given preeminence to geographical considerations and devoted chapters to individual anatomic regions.

We hope this text will assist the pathologist in his evaluation of surgical specimens by the compilation in one volume of the major pathologic entities that arise in or encroach upon the nervous system. Hopefully, it will also enable the clinician to better visualize the anatomic relationships of those lesions that have expressed themselves clinically, and for the radiologist to give substance to the images which he has carefully studied before surgery. Lastly, we hope that the book will sufficiently illuminate neurologic disease so that the medical student will not be intimidated by, but rather encouraged to study, a system so elegant in its normal structure and so complex in its expressions of disease.

Our efforts will be fully rewarded if this book is as instructive and pleasurable to the reader as its preparation has been to the authors.

PETER C. BURGER
F. STEPHEN VOGEL

Acknowledgments

The usefulness of a pathology text rests largely upon the quality of its illustrations, and, for this reason, we greatly appreciate the personal attention given the illustrations in this second edition by Mr. Bill Boyarsky, the departmental photographer.

The preparation of histologic material has been directed by Mr. Bernard Lloyd. We are indebted to him for his continued dedication to quality.

Once again, the exacting chore of proofreading has been willingly undertaken by Dr. Eleanor Branch, whose attention to detail and constructive suggestions have been invaluable.

To Mrs. Bonnie Lynch has fallen the laborious task of preparing the manuscript. Her patience, good humor, and skill are acknowledged with the deepest gratitude.

Facilities for the radiographic study of specimens were kindly provided by Dr. Donald B. Hackel. Cytologic specimens were skillfully prepared in the laboratory of Dr. William W. Johnston.

P.C.B.
F.S.V.

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1.I. INTRODUCTION

The diversity of anatomic structures of the head, and their juxtaposition to the central nervous system, provides the setting for a heterogeneous group of pathologic lesions of neurologic interest. In spite of the nonneural character of most of these, they fall within the bailiwick of neuropathology—underscoring the catholic scope of the latter discipline and its inseparable relationship to general anatomic pathology.

1.II. TUMEFACTIONS AND DESTRUCTIVE PROCESSES IN THE SKULL

1.II.A. Aneurysmal Bone Cyst

Aneurysmal bone cysts are expansile, tender, benign lesions of uncertain genesis that occur in almost any bone of the body including, on rare occasion, the skull. In this latter site, a scalp mass resulting from outward expansion is usually responsible for their discovery (2,6) (Figs. 1.1 and 1.2), while, less commonly, centripetal enlargement results in compression of the brain or cranial nerves with subsequent neurologic symptoms (4–6,8). Most aneurysmal bone cysts in the skull have occurred in the first three decades of life, a feature in common with their extracalvarial counterparts (9,12). The occipital bone (6,8,10) has been more frequently affected than the temporal (5), parietal (2), frontal (1,7) or orbital (2), but the small number of reported cases would caution against any generalization about preferred sites (2).

Macroscopically, the lesion is notable for its vascular channels, which impart a sponge-like appearance and are responsible for the effusion of blood during surgical excision (Fig. 1.3). Soft tissue and bone form compact masses or are inserted as delicate trabeculae between the vascular channels. The content of new bone is not sufficient to offset the overall osteolytic nature, although a thin rim of subperiosteal bone may be present (3,5,6).

Microscopically, the vascular channels are delineated by septa of variable thickness and composition. Some are mere delicate strands of fibrous connective tissue; others are thickened by granulation tissue, proliferating fibroblasts, osteoid, bone, or giant cells—all components that may also be formed into solid masses near the periphery (Fig. 1.4). Leakage of blood may be recent or old, as evidenced by extravasated erythrocytes or hemosiderin-laden macrophages. The vascular channels are noted for the variable presence of endothelium and for the lack of smooth muscle. The latter feature is confirmed by electron microscopy (11).

The diagnosis of aneurysmal bone cyst may be suggested by the radiographic and gross features, especially when the latter can be evaluated in an intact specimen. However, the curet, which may be employed in excision, can mutilate the lesion and obscure its vascular nature. In these fragments the giant cell component may be misinterpreted as a giant cell tumor, or the osteoid and proliferating spindle cells as an osteosarcoma. A kin of the aneurysmal bone

cyst, the hemangioma, shares a content of vascular spaces but is associated with radiating spicules and does not demonstrate the exuberant response of fibroblasts, giant cells, and osteoblasts.

The aneurysmal bone cyst is a benign lesion cured by total excision, or, more commonly, curettage. Recurrence may follow subtotal resection (9).

1.II.B. Chordoma

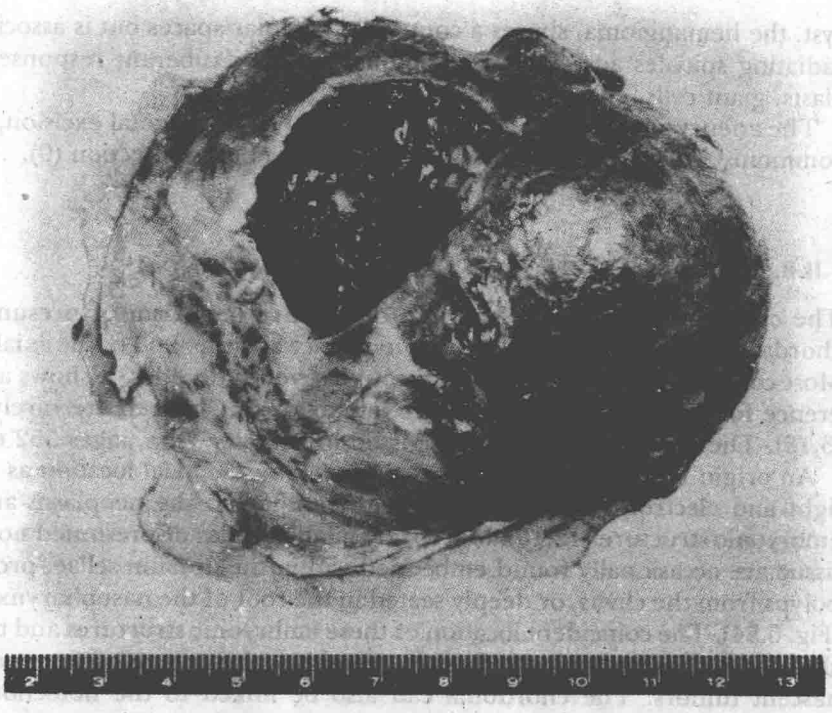
The chordoma is a slowly growing, destructive neoplasm of presumed notochordal origin that occurs in greatest incidence at either end of the axial skeleton. Most commonly seen in the third through seventh decades, it shows a sex preference for males in a ratio of approximately 2:1. Children are rarely affected (3,18). The spinal varieties are discussed in Chapter 7 on pages 552 to 553.

An origin from the notochord is suggested by the axial location as well as by light and electron microscopic features common to the neoplasm and to this embryonic structure (12,16). Moreover, small nodules of presumed notochordal tissue are occasionally found embedded within the dorsum sellae, projecting as polyps from the clivus, or deeply seated in the roof of the nasopharynx (4,20,22) (Fig. 6.54). The coincident location of these embryonic structures and the cranial chordoma suggests that the former are either the anlage of the neoplasm or nascent tumors. The chordoma can also be linked to the notochord by the morphologic similarity of the neoplasm's physaliphorous component to notochord-derived nucleus pulposus cells proliferating in response to puncture of the intervertebral disc in rabbits (6). A notochordal origin leaves unexplained the enigmatic rarity of chordomas along most of the vertebral column where the notochordal derivatives are abundant in the form of the intervertebral discs.

The cranial neoplasms, which comprise about 40% of all chordomas, occur at the base of the skull in the region of the spheno-occipital synchondrosis (the clivus) (Fig. 1.5). Growth of this neoplasm, accompanied by destruction of any restraining bone, may extend the lesion rostrally to involve the sella turcica, optic chiasm, and adjacent cavernous and sphenoid sinuses; dorsally to displace the pons; and laterally to appear in the cerebellopontine angle. Common symptoms therefore include headache, gaze palsies (especially lateral), visual field defects, and long tract signs (1,3,7,10,13,17,18). Although the anlage of the neoplasm is midline, the growth is often asymmetrical and cranial nerve signs therefore are sometimes unilateral (7). Antero-inferior extension creates a nasopharyngeal mass (2,21).

Grossly, the chordoma is a lobulated mass of translucent, gray tissue; it is usually firm and friable but may be soft and sometimes almost mucinous, appearances that belie its tendency to bleed profusely at surgery.

Microscopically, most of the neoplastic cells are contained within lobules that vary in size and are embedded in a fibrous connective tissue stroma (Fig. 1.6). The neoplastic cells vary in configuration and alignment and are both surrounded by and contain variable amounts of mucin. Some, which have well-defined, homogenous, eosinophilic cytoplasm distinct from the surrounding intercellular material, are arranged in solid sheets or strung out into isolated or interconnecting strands (Fig. 1.7). Others demonstrate a spectrum of vacuolation



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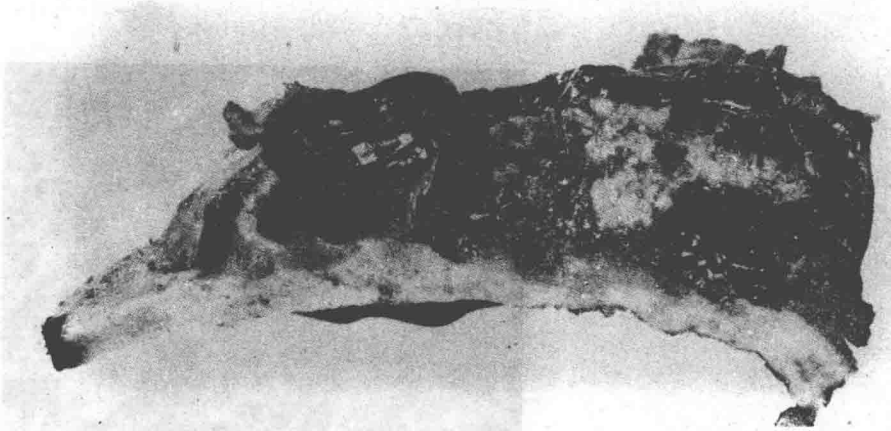

1.3

Figures 1.1, 1.2, and 1.3. ANEURYSMAL BONE CYST

This patient was a 77-year-old female who had noted a protrusion of the forehead for more than 20 years. In the six months before excision the mass rapidly enlarged and became tender. Characteristically, the hemorrhagic aneurysmal bone cyst expands through the outer table and elevates the galea (Figs. 1.1 and 1.3), whereas involvement of the inner table is minimal (Figs. 1.2 and 1.3).

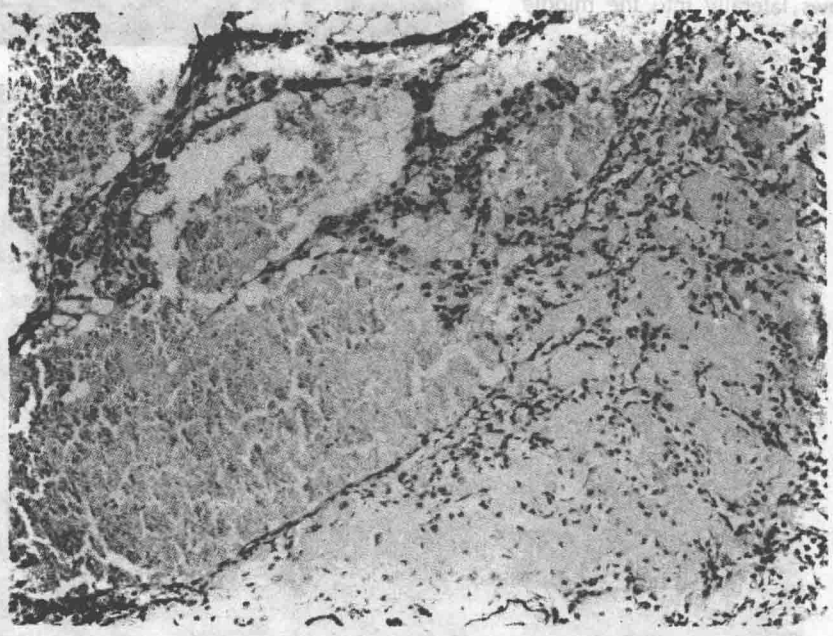

Figure 1.4. ANEURYSMAL BONE CYST

Prominent histologic features of the aneurysmal bone cyst are multiple thin-walled vascular channels associated with considerable osteoid and fibrous tissue (H&E, ×100).

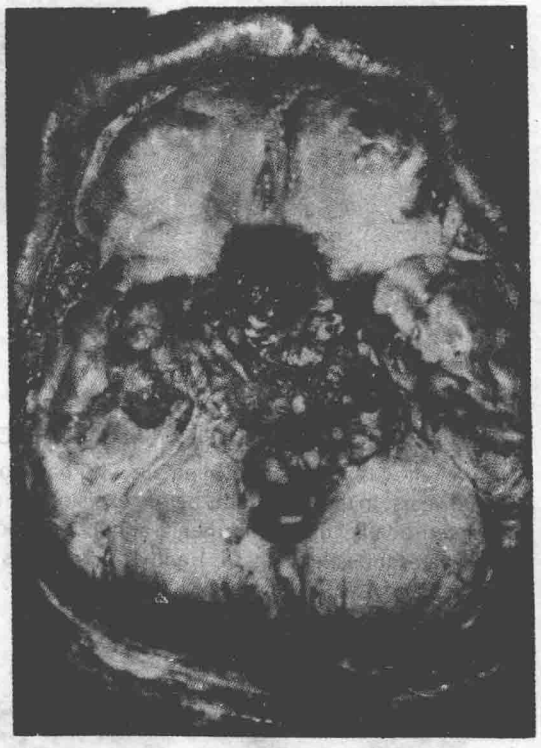

Figure 1.5. CHORDOMA
This neoplasm destroys the clivus, extends rostrally into the sella turcica, and flows laterally into the middle cranial fossae.

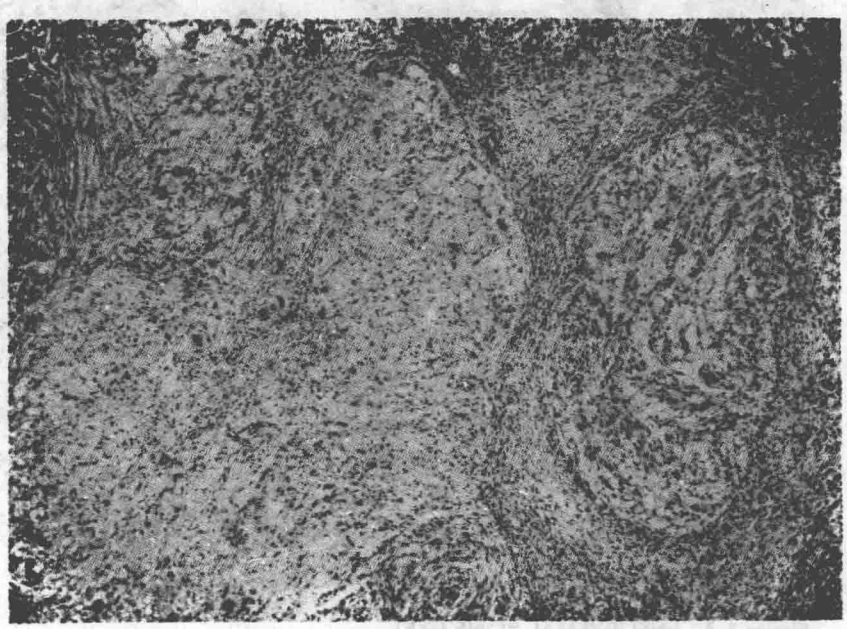

Figure 1.6. CHORDOMA
The characteristic lobularity of the chordoma is readily appreciated at low magnification (H&E, ×40).

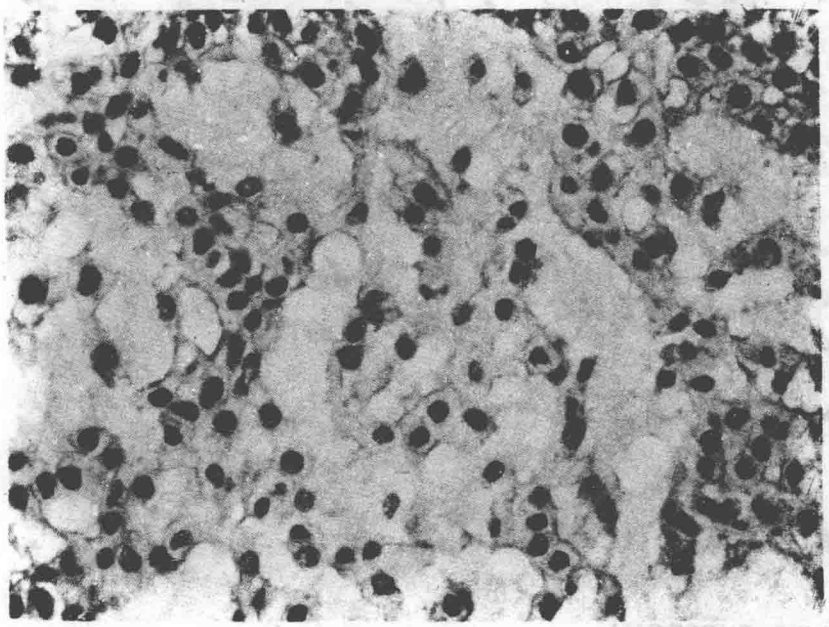

Figure 1.7. CHORDOMA
In some regions the cells have well-defined borders and are arranged into cords separated by extracellular mucinous material (H&E, ×250).

which terminates in cells so altered that the distinction between intra- and extracellular material cannot be defined. These latter elements are known as physaliphorous (Gr. bubble-bearing) cells and are considered characteristic of the chordoma (Fig. 1.8). These two cell types do not exist as distinct populations, however, because of the presence of transitional forms. It has been suggested that, similar to the developing notochord, the vacuolated cells sequentially evolve from those with solid cytoplasm (11,16). Electron microscopic studies have shown that the extracellular material is probably formed within the endoplasmic reticulum and that the presence of this mucinous substance, either within these dilated cisternae or as extracellular invaginations into the cytoplasm, imparts the vacuolar appearance seen by light microscopy (8,11,14). It is of interest that the notochordal cells and those of the chordoma share the characteristic of desmosomal connection (5,14). The nuclei of the chordoma cells may show marked pleomorphism and rare mitotic figures.

In addition to the typical chordoma cells, the neoplasm may contain areas of differentiation into conventional mesenchymal tissues such as bone (Fig. 1.9) and especially cartilage (Fig. 1.10). The latter, in the form of chondrosarcoma, may be represented focally or may be so extensive as to dominate the histologic picture. Usually, the bony tissue is benign and the fibrous component reactive or supportive, rather than neoplastic, although one osteosarcoma has been identified in a sacral lesion (9) and a fibrosarcoma in a spheno-occipital neoplasm (9).

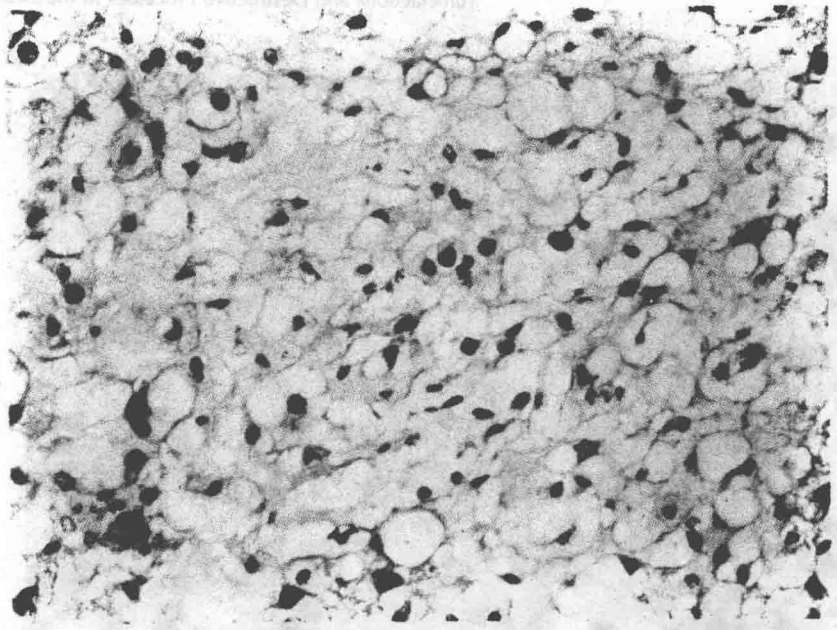

Figure 1.8. CHORDOMA
Elsewhere in the neoplasm of Fig. 1.7, the cells are severely distorted by vacuolization and their boundaries are indistinct. These are the classic physaliphorous cells of the chordoma (H&E, ×250).

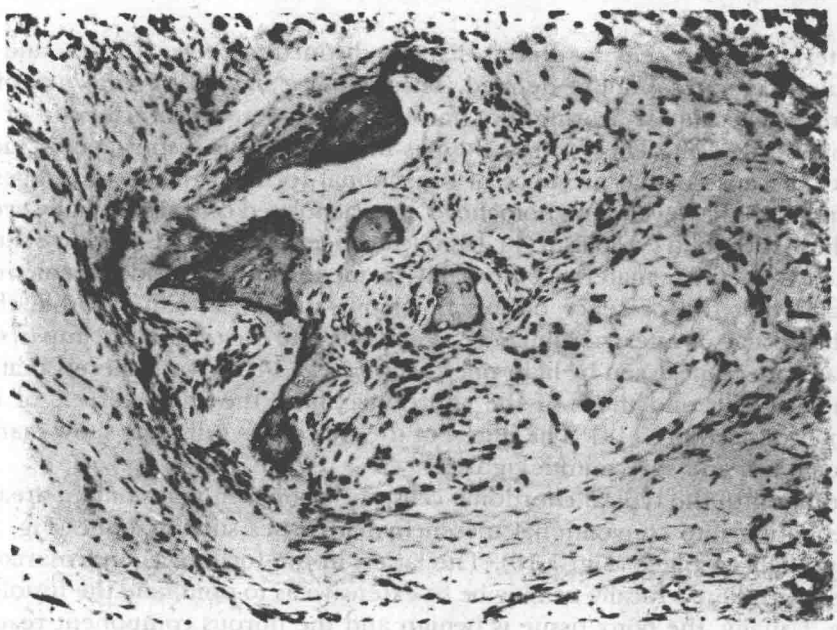

Figure 1.9. CHORDOMA
Bone formation may be observed in some chordomas. Here, it is adjacent to a lobule of typical physaliphorous cells (H&E, ×120).

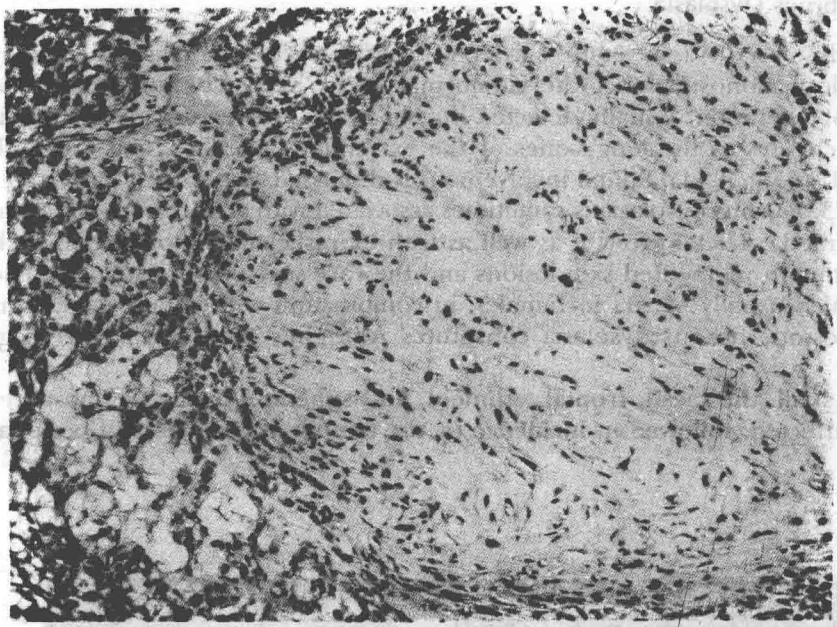

Figure 1.10. CHORDOMA
Foci of chondrosarcoma may be encountered in the chordoma (H&E, ×100).

The differential diagnosis of the chordoma in the skull is usually simplified by its characteristic location as well as its distinctive gross and microscopic features. However, the latter aspects may be obscured by the presence of a predominating chondrosarcoma and it then becomes difficult to distinguish the chordoma from a chondrosarcoma. The property of mucin production in chordomas and adenocarcinomas creates histologic similarities. However, the adenocarcinoma is not likely to produce a large, slowly growing, exophytic mass, nor one that at low magnification shows formation of lobules lacking central necrosis. While nuclear pleomorphism may be shared by both neoplasms, mitotic figures are much less common in the chordoma.

Although the cranial chordoma is not usually malignant in the sense of breeding distant metastases, dissemination to liver, lungs, and spinal meninges has been recorded (1,4,18,19). The location and extent of the lesion at the time of diagnosis generally make surgical extirpation impossible and subtotal resection hazardous. Radiation therapy following partial removal may extend the patient's useful life. In a study where both of these modalities were employed, the average survival for patients with a typical chordoma was 4.1 years in one series (9) and 3.6 in another (13). Interestingly, chordomas containing foci of chondrosarcoma in another group of patients in the same study were associated with a longer survival, 15.8 years.

1.II.C. Fibrous Dysplasia

Fibrous dysplasia is a skeletal disorder of unknown etiology characterized by polyostotic or monostotic foci wherein normal bone is altered by abnormal fibro-osseous proliferation. Usually detected during the first three decades of life, it favors such sites as the long bones of the extremities, the ribs, and the skull (1,11,12,18) (Fig. 1.11). Spinal involvement is discussed in Chapter 7 on pages 571 to 573. Fibrous dysplasia is sometimes associated with accelerated, but eventually stunted, skeletal growth, as well as with extraskeletal abnormalities such as the common pigmented skin lesions and the rare sexual precocity (12). The latter, which usually occurs in females, in combination with cutaneous lesions and polyostotic fibrous dysplasia constitutes Albright's syndrome (2,14) (Fig. 1.12).

In the skull, the facial, frontal, ethmoid, and sphenoid bones are most frequently affected, followed in incidence by the temporal, parietal, and occipital

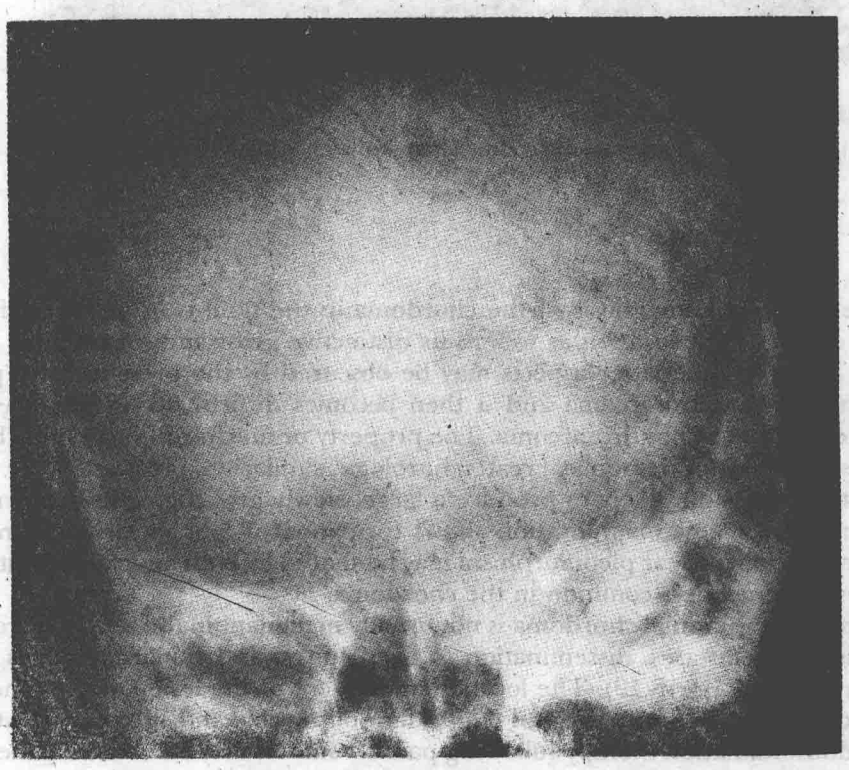

Figure 1.11. FIBROUS DYSPLASIA
In this case of advanced fibrous dysplasia there is both destruction and formation of bone. In contrast to Paget's disease, the process is unilateral and facial bones are involved. (Courtesy of Dr. John A. Goree, Durham, NC)