FIBROUS DYSPLASIA OF THE SKULL

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GENERAL ASPECTS OF FIBROUS DYSPLASIA

Within recent years, largely as a result of the contributions of Albright and his associates,2,4 of McCune and Bruch,34 Lichtenstein,31 and of Lichtenstein and Jaffe,32 the concept of fibrous dysplasia of bone as a disease entity has become established. The clinical features of this disorder were clearly epitomized in the subtitle of a paper published by Lichtenstein and Jaffe32 in 1942, as “A condition affecting one, several or many bones, the graver cases of which may present abnormal pigmentation of the skin, premature sexual development, hyperthyroidism or still other extraskeletal abnormalities.” The term, Albright’s syndrome, has been applied to those cases in which disseminated bone lesions are associated with cutaneous pigmentation and sexual precocity in females.

Judging from the literature and from our own experience, it is evident that involvement of the skull is frequently observed in this condition and may, in fact, constitute its only manifestation. It is with this aspect of the disease and its neurosurgical implications that we are primarily concerned. Detailed accounts of a more general nature are available in a number of publications.2,3,5,8,10,16,17,21,23,26,31,32,46

Pathology. Involvement of osseous structures may occur alone or in association with the endocrine and cutaneous manifestations of Albright’s syndrome. The skeletal lesions may be confined to a single bone (monostotic), or may involve a number of bones (polyostotic). Regardless of the clinical form assumed by the disease, the basic histopathologic changes are fundamentally identical in all cases. Within an affected bone, either flat or tubular, there takes place an accumulation of fibrous connective tissue. This may be cellular, consisting of spindle cells arranged in whorls, or densely collagenous. Dispersed throughout the connective tissue, trabeculae of immature bone may be frequently observed. This new bone is poorly formed, imperfectly calcified and may show evidence of osteoclastic resorption. In some cases the amount of bone laid down may be considerable, so that the connective-tissue element appears relatively insignificant. Islands of cartilage may also be observed within the fibrous tissue and occasionally small cysts, areas of hemorrhage, giant cells or foam cells are to be found. With the ter-

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mination of the active phase of the disease, the fibrodysplastic tissue may become increasingly ossified. In some bones, especially those of the skull, sclerotization of the fibrodysplastic tissue may eventually become extremely marked. As will be indicated later, the roentgenologic manifestations of this disorder are variable, depending on the relative amounts and distribution of fibrous tissue and bone.

Grossly such dysplastic tissue appears yellow or greyish-white, is of variable consistency, though usually firm and rubbery, and often feels gritty owing to the presence of many small bone spicules. It may occasionally contain small cysts filled with brown fluid. The skeletal structures affected by this disease may either retain their shape or become widened and deformed, or even fractured as a result of the expansile nature of the pathologic process.

Etiology. The etiology of this disorder is unknown. As indicated by Albright and his associates, the regional distribution of the lesions would tend to exclude a metabolic or endocrine cause. They suggest that the responsible factor may be either a neurologic disturbance or an embryologic defect. A congenital anomaly of development that may account for both skeletal and extraskeletal features is advocated by Lichtenstein and Jaffe. In keeping with this hypothesis, abnormal activity of the specific bone-forming mesenchyme would be the cause of the osseous lesions. The embryonic defect theory is also favored by Neller. The possibility that the monostotic form of fibrous dysplasia may represent a disturbance of the normal reparative processes following injury is proposed by Schlumberger. Derangement of liver function, hormonal disturbances, hyperemia caused by anomalies of the sympathetic nervous system, and chronic hyperparathyroidism due to a hyperphosphatemia have also been suggested as etiologic factors.

Snapper formerly was of the opinion that fibrous dysplasia represented a variant of Hand-Schüller-Christian's disease. In the latest edition of his book, however, he has modified this view and concedes that in most cases this interpretation does not appear to have been substantiated. Thannhauser's thesis that fibrous dysplasia is a form of neurofibromatosis of von Recklinghausen has been critically analyzed by Jaffe and Albright. Both reject this concept. The clinical and anatomic evidence they submit leaves little doubt that the two conditions are indeed separate entities.

Clinical Aspects. Fibrous dysplasia is primarily a disease of childhood, its progress frequently becoming retarded or arrested when adult life is reached and skeletal growth ceases. It is believed that in cases in which the disorder is first encountered later in life, the skeletal lesions originated during childhood. Females are more commonly affected than males. The usual clinical manifestations are limp, deformity, pain and pathologic fracture. In cases with advanced skull lesions, the degree of disfigurement may be considerable. This will be discussed in greater detail subsequently. When more than one bone is affected, the involvement is frequently exclusively or predominantly unilateral. It may be monomelic. Long bones are mainly affected, the proximal parts more than the distal. Thus the upper half of the