MANAGEMENT OF FIBROUS DYSPLASIA

Essay

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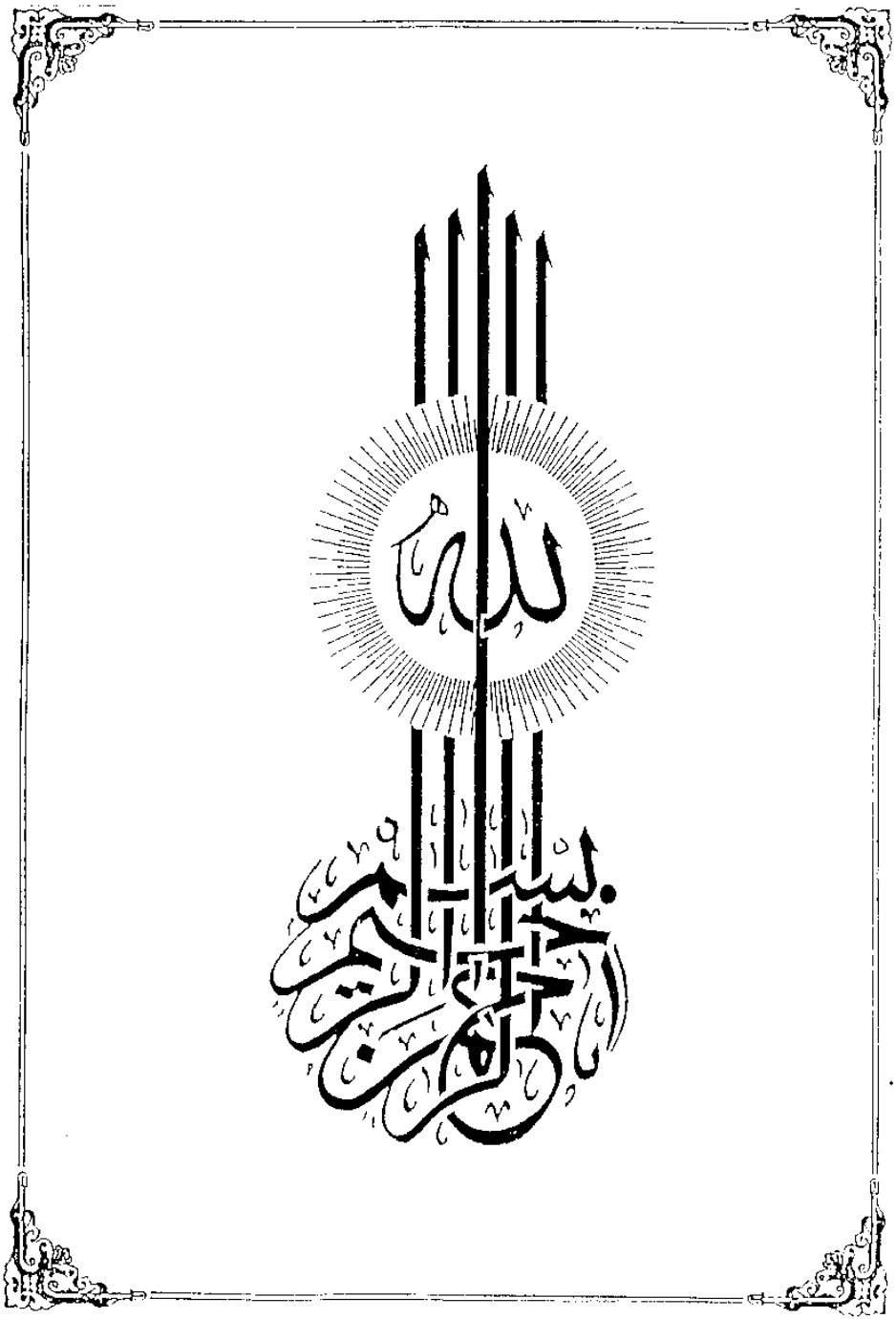
TO...

MY PARENTS



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Alaa El-Deen

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INTRODUCTION

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Historical view:

In 1891 Von Recklinghauwsen reported a group of benign fibro-osseous lesions of bone using the term "osteitis fibrosa". This group of patients was probably a heterogenous one and may have included cases that would now be classified as "Fibrous dysplasia". During the next several decades osteitis fibrosa was used to describe most benign fibro-osseous lesions, including multifocal lesion that occured in the absence of hyperparathyroidism. In 1937 Mc Cune and Bruch reported a case of osteodystrophia fibrosa in which multipule fibro-osseous lesion occured associated with precocious puberty and skin pigmentation. At about the same time, Albright and coworkers reported several similar cases and this combination of features was subsequently termed Albright's syndrome. Lichtenstein and Jaffe (1942) recognized that the bony lesions may occur in one or more sites in the absence of other clinical features and proposed the term "fibrous dysplasia". *(Marks & Bauer., 1989)*

DEFINITION:

Fibrous dysplasia is a skeletal developmental anomaly of bone-forming mesenchyme in which the osteoblast fails to undergo normal morphologic differentiation and maturation. *(Lichtenstein and Jaffe., 1942).*

CLASSIFICATION:

Its classification depends upon the number of bones involved and the extent of involvement of individual bone.

1- MONOSTOTIC FIBROUS DYSPLASIA:

When the disease affects one bone, it is called monostotic fibrous dysplasia, In this type of disease, there is swelling with or without pain, it may be associated with fractures. Cutaneous pigmentation is uncommon. Serum alkaline phosphatase, calcium and phosphorous are within normal limit.

2- POLYOSTOTIC FIBROUS DYSPLASIA:

When the disease affects more than one bone, it is called POLYOSTOTIC fibrous dysplasia.

In this type of disease, there is swelling, skeletal deformity and pain due to fracture. Abnormal cutaneous pigmentation is common.Precocious puberty and premature skeletal maturation are less common. Hyperthyroidism, diabetes mellitus, myxomas

of soft tissue and increase in serum alkaline phosphatase are unusual associated features. *(Mirra., 1980).*

NOMENCLATURE:

The number of different titles under which cases representing the fibrous dysplasia in degrees of severity have been discussed in the past is bewildering. They do fall into certain groups, however. Specifically, most of titles represent variation on the subject of "osteitis- fibrosa" sometimes qualified by such term as "focal", "unilateral," or "disseminated" and often also supplemented by references to some of the extraskeletal abnormalities. Most of the rest of the titles ring the changes on "fibrocystic disease of bone", or on "a form of Recklinghausen's disease of bone." These names are often likewise qualified by such term as "regional" and "unilateral" and sometimes likewise supplemented by reference to associated extraskeletal changes. In relation to fibrous dysplasia of skull bones and particularly of jaw bones, such terms as "fibrous osteoma" or "ossifying fibroma" have most often been employed. *(Jaffe, 1958).*

ETIOLOGY:

The etiology of fibrous dysplasia is unknown. *(Lichtenstein and Jaffe., 1942).* There is some theories to explain the

association of endocrinopathies with fibrous dysplasia, with or without a concomitant McCune-Albright syndrome.

Some authers postulate, the possibility of increased sensitivity of target organs (i.e. gonads and breasts) to normal or minimally elevated prepubertal circulating pituitary hormones. So premature vaginal bleeding has, therefore, been explained by early release of ovarian hypersensitivity to follicle - stimulating hormone.

Alternatively, another theory suggesting end-organ autonomy was postulated, in a 6 month old girl whose vaginal bleeding ceased after removal of a functioning luteinized ovarain cyst. She also had cushing's syndrome secondary to adrenal nodular hyperplasia. Biochemical evidence supported adrenal and gonadal autonomy and refuted hyperfunction mediation by the hypothalamic, pituitary axis. *(Danon and Crawford, 1974; Senior and Robboy, 1975).* Albright, noting a variety of associated endocrinopathies, initially stressed end-organ sensitivity as a cause, whereas others subsequently maintained that hypersecretion of hypothalamic releasing hormones was responsible. *(Hall and Warrick., 1972).*

Such hypersecretion, however, generally has not been substantiated neither in the hyperthyroidism with toxic nodular

goiter that occurs in 5-30% of the cases of fibrous dysplasia, nor in many of the early reported cases of reputed acromegaly or cushing syndrome that have fail to satisfy modern endocrinologic criteria. *(Scurry, Bicknell and Fajans, 1964; Hamilton and Maloof, 1973).* Some of these clinical diagnoses must also be regarded with caution because acromegalic facies, the exophthalmos of Graves' disease and visual defect suggesting a pituitary tumor which may be mimicked by facial and skeletal involvement in Fibrous dysplasia alone. Although pituitary modularity and hyperplasia have been said to exist in fibrous dysplasia frank tumors have not been documented consistently. The significance of recent demonstration by computed tomography and ultrasonography of presumed pituitary and ovarian structural abnormalities remains uncertain because computed tomographic standards for endocrinologically normal subjects have yet to be established. *(Rieth, Comite, Shawker and Culter, 1984).*

The observed pituitary prominence may in fact, represent physiologic enlargement of the normal prepubertal gland. Recent discoveries of single and multiple adenomas in the pituitary, adrenal, thyroid and parathyroid glands, in addition to hyperplasia of target organs, tend to support to the thesis of glandular autonomy and strengthen the hypothesis of an

exciting variant of multiple endocrine adenomatosis *(DiGeorge, 1975, D'Armiento et al, 1983)*

So, the endocrinopathies in fibrous dysplasia with or without Mc Cune. Albright syndrome can be explained by two major theories:

One that congential hypothalemic dysfunction with hypersecreation of releasing hormones affect the target organs.

The other theory postulates an underlying form of multiple endocrine neoplasia with autonomous function of the involved endocrine glands. *(Hall and Warrick, 1972; DiGeorge, 1975).*

PATHOLOGY

PATHOLOGY

RELATION TO AGE AND SEX:

The ages of patients with monostotic fibrous dysplasia have wide ranged from 10 to 70 years but recognition is most frequent in the second and third decades of life. The age distribution is considerably younger in polyostotic fibrous dysplasia as its more severe mainfestations lead to earlier clinical and radiographic recognition. Two thirds of patients are symptomatic before the age of 10 years. *(Harris et al., 1962).*

Both sexes are equally affected. *(Turek, 1984)* Some authers said that fibrous dysplasia is definitely more common in females than in males in the proportion of 2 or 3 to 1. *(Jaffe, 1958)*

Fibrous dysplasia usually become symptomtic during childhood but the adolescent patient who was discribed by Stevens-Simon and his coworkers (1991) is unusual because the patient was a girl and she was not known to suffer from fibrous dysplasia until she become pregnant and experienced a dramatic reactivation of the symptomes. So they postulate that there is exacerbation of fibrous dysplasia associated with adolescence pregnancy. *(Stevens et al., 1991).*

DISTRIBUTION:

Approximately 70-80% of cases are monostotic and 20-30% are polyostotic *(Gibson, 1971).*

Monostotic fibrous dysplasia is most frequent in ribs, femur, tibia, mandib le, clavicleand humerus in a decreasing order of frequency. Polyostotic fibrous dysplasia more frequently involves the skull, facial bones, pelvis, spine and shoulder girdle. *(Feldman, 1988).*

MACROSCOPIC FEATURES:

The gross features depend upon the amount of woven bone produced in the lesional fibrous tissue, the degree of degenerative changes(cysts)and the extent of bone involvement. Since the basic prolifrating process is fibrous, the tissue will be firm and white. Most cases produce significant quantities of woven bone so that the tissue will be gritty. A magnifying glass may be necessary to appreciate the very fine spicules of bone produced that are usually in the range of 0.1 mm in width by 0.5 mm in length. Normal bone trabeculae are at least 2-3 times thicker. It is common to find areas of cystic degeneration, the cysts may range from 1 mm to several centimeters in size and contain a mucoid serous or serosanguinous fluid. Areas of cartilage may be seen.