

Head: Fibrous Dysplasia

Sub: What is fibrous dysplasia?

Copy: Fibrous dysplasia is a congenital, non-hereditary, progressive skeletal disorder by which normal bone is replaced by a variable amount of structurally weak fibrous and osseous tissue. It is characterized by the presence of woven bone. In normal bone formation, woven bone appears first and later matures into lamellar bone. Under concentrated light, lamellar bone trabeculae (rows of cells bridging an intercellular space) show widely spaced parallel birefringent lines, which are rimmed with osteoblasts and osteoclasts. In fibrous dysplasia, where bone development ceases in the woven bone stage, the trabeculae show random irregular birefringence and are surrounded by abundant fibrous tissue.

Monostotic disease is the most common type of fibrous dysplasia, occurring in 70% of cases and most frequently on the long bones — femur, ribs and skull. Polyostotic disease, afflicting 30% of patients, occurs in two or more bones and involves the head and neck 50% of the time. The third type, McCune-Albright syndrome, occurs in 3% of cases and is characterized by polyostotic fibrous dysplasia, skin pigmentation and precocious puberty in females.

Skull involvement occurs in 27% of monostotic and up to 50% of polyostotic patients. “Leontiasis ossea” is the specific name for fibrous dysplasia involving the facial and cranial bones. In its common form, one or more bones progressively increase, encroaching on the cavities of the orbit, mouth, the nose and its sinuses. Abnormal protrusion of the eyeball (exophthalmos) may develop and eventually cause complete loss of sight because of compression of the optic nerve. There may also be interference of the nasal passage and with eating.

Fibrous dysplasia is a progressive disease, which typically occurs early in childhood and continues until skeletal growth ceases. It is first detected in young children, manifesting as a swelling of the jaw and a possible separation of teeth. It causes deformity and impingement, and if occurring in the frontal and sphenoid bones, it can eventually lead to deformation of facial features and skull shape.

Sub: Fibrous dysplasia of the skull base

Copy: Fibrous dysplasia of the frontal and sphenoid bones eventually leads to distortion of facial features and skull shape as a result of the proliferation of thick, dense bone. Craniofacial fibrous dysplasia is different in that it ignores suture lines; more than one bone is usually involved. It can also result in cranial nerve impingement. Disease of the temporal bone may present a patient with 80% hearing loss because the inner ear canal narrows. It may also cause facial nerve (VII) paralysis or vertigo. Although any of our 12 cranial nerves and their cranial foramina can be involved with fibrous dysplasia, resulting in cranial nerve deficits, visual and hearing loss represent the more common and debilitating clinical presentations.

Sub: How does fibrous dysplasia present in radiographic appearance?

Copy: The radiologic appearance of fibrous dysplasia can appear as a lucent area with a sclerotic rim. In the skull base and facial bones, fibrous dysplasia manifests as marked sclerosis and bone thickening.

[Slide 1]

[Slide 2]

Sub: Treatment

Copy: It is estimated that patients with fibrous dysplasia are 400 times more likely than the general population to develop a malignant bone tumor. Therefore radiation is not recommended. Treatment involves management through observation first, then conservative measures such as surgical excision and shaving. A more curable treatment by complete is bone resection.

Surgical intervention is generally intended for cosmetic facial deformities and cranial nerve compressions. The abnormal bone must be completely removed. Surgery is usually delayed until adolescence; however, if the progression of the disease comprises neurological function, a decompressive procedure should be considered early in childhood to preserve normal function.

Fibrous dysplasia is a non-hereditary, progressive and benign disease, in which treatment options are symptomatic. Some lesions are amenable to resection for a cure by a single procedure. More often, most lesions can be managed through staged procedures with overall very favorable outcomes and good long term prognosis.

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