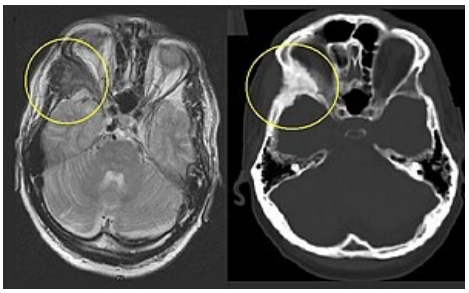
Fibrous bone dysplasia

Fibrous bone dysplasia (or *Jaffé-Lichtensteinova disease*) is a more common, non-hereditary disease characterized by the presence of ***expansive connective-bone tissue inside the diaphyses and metaphyses of growing bones*** (and possibly other symptoms).



Fibrous bone dysplasia of the cheekbone (os zygomaticum)

Pathological anatomy

- **replacement of spongiosis** of (mainly long) bones by **high-cell fibrous tissue** (dif. dg. fibrosarcoma) in young people
- The connective tissue is sometimes arranged in a nodular manner, contains trabecular meshwork without osteoblastic borders
- it is probably **a bone tissue development disorder** (stopped at the mesh bone stage, does not transform into lamellar bone)

Clinical picture

- bones susceptible to pressure, tend to be deformed (**shepherd's stick-shaped femur**), tendency to pathological fractures
- painful swelling, lameness, shortness of limbs, genu varum nebo valgum
- occurrence in the form of monoostotic and polyostotic
- **McCune-Albright syndrome**: association of the polyostotic form of fibrous dysplasia with foci of hyperpigmentation in the lumbar region, endocrine disorders (mainly pubertas praecox) and pathological changes in the hip joints (only in women)
- familial fibrous dysplasia in the jaws leads to **cherubism**

X-ray image

- irregular cystic confluent clearing within the bone ("milk glass")
- thinning of the cortex

Therapy

- excochleation, spongioplasty
- corrective osteotomy, prolongation with an external fixator

Links

Related articles

- Ossifying fibroma
- Cherubism

Used literature

- DUNGL, P., et al. *Ortopedie.* 1. edition. Praha : Grada Publishing, 2005. ISBN 80-247-0550-8.

Related articles

- Congenital multiple exostoses
- Enchondromatosis (Ollier's disease))
- Osteogenesis imperfecta
- Morbus Albers-Schönberg (bone marbling, osteosclerosis, osteopetrosis)
- Osteopoikilosis