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Fibrous dysplasia in children – presentation of two cases

Fibrous dysplasia is localized, osteolytic, expansive disease and may involve a single bone or many bones. It imitates a neoplasm and is characterized by the presence of fibrous-osseous tissue. The etiology of the disease is not clear. The base of the disease is developmental disorder of mesenchyma and impaired differentiation of bone marrow into fibrous marrow and bone. Usually affected one side of the body can be explained by congenital developmental disorder. It starts during the first or second decade of life, more often in women. It is manifested by pains, swelling, dilation or malformation of bones, and the foci may involve single bones or many bones (femoral bone, tibiae bone, ribs, craniofacial bones). Abnormal skin pigmentation, accelerated growth termination or premature sexual development may be accompanied by osseous changes (1–5).

Radiological image demonstrates clearances clearing up and the change looks like a "frosted glass". With time there is an increase of the focus with medial atrophy of cortex of bone and simulataneously there is a broadening of marrow cavity without periosteal reaction (3). Thickened trabecula which separate the created cavities present the "soap bubbles" image. This disease is differentiated in children with adolescent bone cyst, non-ossifing fibroma and eosinophilic granuloma (3).

The microscopic picture reveals poor vascularisation, spirally twisted connective tissue, yellow or white-grey, taking place of the trabecula and bone marrow, with a small number of osteoclasts and osteoblasts which form osteoid tissue (2,3). Fibrous dysplasia occurs in about 7.5% of patients before 30 years of age and the cranial dysplasia are still more rare (2,3). This kind if disease is occasionally diagnosed in children and the authors decide to present two boys with this type of disorder.

PRESENTATION I

Fourteen-year-old boy (B.B.) in whom painless hump within left frontal sinus was noticed. An X-ray imaging of paranasal sinuses (Fig. 1) and then a CT of head were performed (Fig. 2, Fig. 3). Also a sample from the pathologically changed site was taken. The histopathological examination confirmed the diagnosis – fibrous dysplasia (Fig. 4), which was in compliance with the diagnosis made on the basis of the radiological test. The boy underwent neurosurgical intervention with complete removal of the lesion. The post-operated niche was treated with TachoComb and Codubix. During the two-years' observation we have not confirmed any pathological change in the site of operating or in any other bones. Malformation of face was the indication for the surgical operation in this case.

Fig. 1. B.B. – X-ray imaging of paranasal sinuses

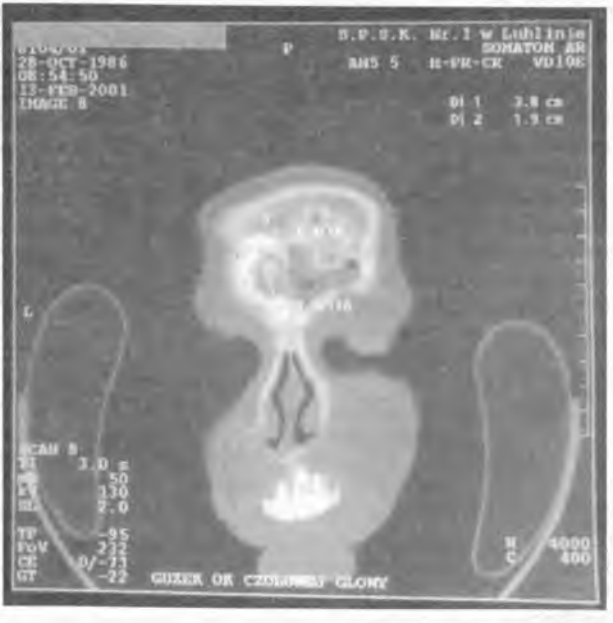

Fig. 2. B.B. – CT scan of skull bone

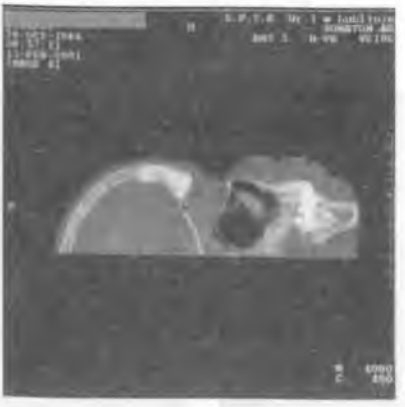

Fig. 3. B.B. – CT scan lateral projection

Fig. 4. B.B. – Irregularly shaped trabecula of bone lie in a moderately cellular fibrous stroma. (H+E. Magn. 250x)

PRESENTATION 2

Eleven-year-old boy (D.K.) was admitted to the department due to five-months' continuing swelling of the mandible arm on the left side. The swelling periodically got increased in size causing pain and face deformation. CT imaging was performed and next a sample was taken (Fig. 5, 6). The radiological image and histopathological tests (Fig. 7) confirmed fibrous dysplasia. In spite of face deformation and with regard to age. the dental surgeons did not qualify the patient for surgery at the moment. The indication for total removal of the lesion will be the risk of pathological breaking of the mandibula.

Fibrous dysplasia is a rare and tumour-imitating bone disease. It may cause diagnostic problems, particularly if the diagnosis is based only on the radiological tests and not supported by histopathological verification. Causative treatment is not known so the surgical intervention should be performed in a very non-mutilating way. Children with this type of diagnosis require prolonged

observation because after many years it may lead in 0.5% of patients to neoplastic transformation to osteosarcoma or fibrosarcoma (1, 3, 5).

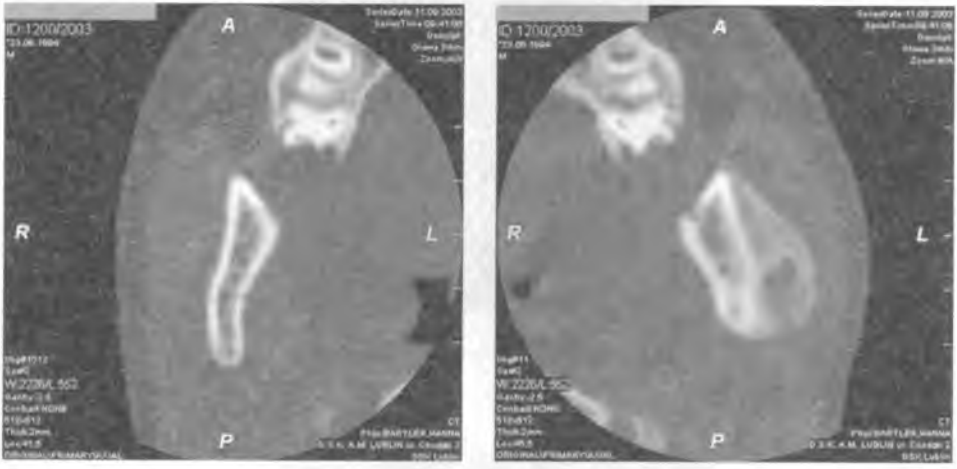

Fig. 5. CT scan: on the left side – fibrous dysplasia

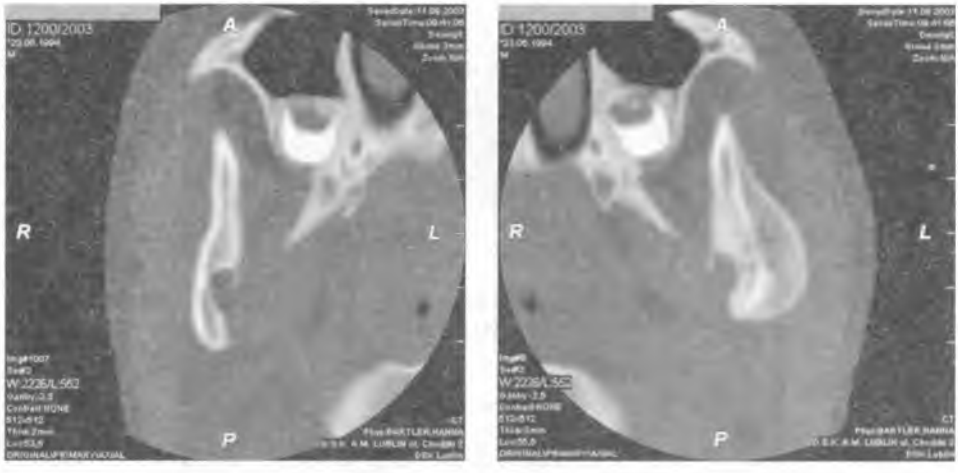

Fig. 6. CT scan – another projection

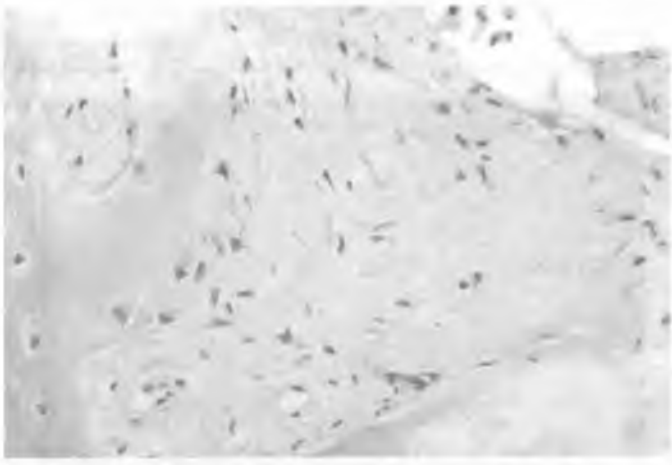

Fig. 7. Histopathological tests (H+E. Magn. 250x)

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SUMMARY

Fibrous dysplasia is an osteolytic, expansive disease. It is characterized by dilation, feeling of pain in the bones and bone deformation. It occasionally occurs in children, mostly in the first and second decade of life. Etiology of this disease has not been explained yet. It may occur in a single bone or in several bones. Due to its rare occurrence and diagnostic problems, the authors present two cases of disease recognized in boys. The results of radiological and histopathological examinations were revealed, which will enable the diagnosis and further treatment.

Dysplazje włókniste u dzieci – prezentacja przypadków

Dysplazja włóknista jest schorzeniem obejmującym kości i charakteryzuje się rozdęciem, bolesnością kości oraz jej deformacją. Występuje sporadycznie u dzieci, przeważnie w pierwszej–drugiej dekadzie życia. Etiologia tej jednostki chorobowej nie jest wyjaśniona. Może dotyczyć pojedynczej kości lub występować w kilku. W związku z rzadkim występowaniem, trudnościami diagnostycznymi, autorzy prezentują dwa przypadki rozpoznane u chłopców. Przedstawione zostały wyniki badań radiologicznych i histopatologicznych, które pozwoliły na postawienie rozpoznania oraz dalsze leczenie.