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Usefulness of CT and MRI in diagnostics of cranial fibrous dysplasia lesions

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Summary

Background:

Fibrous dysplasia (FD) is a developmental anomaly of unknown pathogenesis in which normal bone is replaced by an abnormal fibrous tissue. The process may affect a single bone (monoostic form of fibrous dysplasia) or multiple bones (polyostic form). The skull is the second most common site of FD and comprises 25% of cases.

The purpose of this article is to evaluate the usefulness of CT and MRI in diagnostics of cranial fibrous dysplasia lesions.

Material/Methods:

We analyzed 13 CT and 6 MR examinations performed in 9 females aged 40 to 73 years (mean age 54.7 years). Four patients were referred to the examinations with a diagnosis of fibrous dysplasia, 3 others –with a suspicion of neoplastic disease and 2 patients - with no preliminary diagnosis. Clinical symptoms included: headache, deformity and asymmetry of craniofacial region.

Results:

Fibrous dysplasia lesions were unilateral in 8 patients; in one patient they were bilateral. The sphenoid and ethmoid bones were the most common single bones involved by FD lesions. Among 9 cases of FD, orbital involvement was found in 8, nasal sinuses and cavity involvement in 6 and intracranial extension in 3 patients.

Conclusions:

CT and MR are useful in confirming the diagnosis of FD and especially in evaluating the extent of the disease.

Key words:

fibrous dysplasia • cranial • CT • MRI

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Background

In literature the fibrous dysplasia (FD) is referred to as the Jaffe-Lichtenstein disease, *osteitis fibrosa juvenilis* or *osteitis fibrosa disseminata* and represents developmental disorder characterized by fibrocystic structure of bones and replacement of osseous tissue by fibrous and osteoid tissue of variable consistency. The bone is deformed – thickened or thinned, due to uneven development of abnormal osseous tissue [1, 2].

The disease usually begins in the first two decades of life, rarely in adult age. It occurs in two forms: more frequent-monoostic (70-80% of cases) and less frequent – polyostic

form (20–30%) which often affects one half of the body. The latter can coexist with endocrine dysfunctions and pigment skin lesions and in such case is defined as the Albright-McCune-Sternberg syndrome [3, 4].

The incidence of FD lesions in cranial and craniofacial bones is estimated for 10-20% of monoostic cases and 50% of polyostic cases [5, 6].

Lesions in the course of FD can be asymptomatic or manifest as bone deformity (most often visible in craniofacial bones), bone pain, compression of soft-tissue structures of orbit, as well as disturbances of hearing and equilibrium [7].

Table 1. Location of the lesions in the skull.

Location of the FD lesions	Number of patients (n=9)
Sphenoid bone	7
Ethmoid bone	5
Maxillary bone	2
Zygomatic bone	1

Although the radiographic appearance of cranium affected by FD is well known and described, hardly any reports in Polish literature mention the use of computed tomography (CT) and magnetic resonance (MR) in this disease. Therefore, the aim of this work is to discuss the use of MR and CT in diagnostics of cranial fibrous dysplasia on the basis of the own material.

Materials and methods

The study comprised 9 female patients aged 40–73 (mean age 54.7). Examinations were performed in the Department of Radiology of the Marciniak's Lower-Silesian Specialist Hospital in Wrocław. Altogether 13 CT and 6 MR examinations were carried out. Two patients underwent 3 CTs each, 2 patients – 1 MR each. All in all, one or more CT examinations were performed in 7 patients and one or more MR examinations – in 5 of them.

Four patients were referred to the examination with clinical diagnosis of fibrous dysplasia, three with suspicion of neoplastic process, while 2 other patients – with non-defined preliminary diagnosis. Clinical symptoms observed in patients included pain, deformity and craniofacial asymmetry.

CT examinations were carried out using 5 mm thick slices (craniofacial area) and 8 mm (cerebrocranial area), in all cases with the use of bone and soft-tissue windows. The scope included cranium and upper part of craniofacial area including orbits. In some cases multiplanar reconstructions and three dimensional (3D) CT were also applied.

The MR examinations were carried out in sequences FSE (T2-weighted images), FE (T1-weighted images) and FLAIR sequences, before and after the contrast medium administration, in frontal, sagittal and transverse planes.

Results

In most patients the dysplastic lesions of bones occurred on one side of the body. Only in one patient they were bilateral.

The most common locations of lesions considering the single bones were sphenoid and ethmoid bones (table 1) (fig. 1).

In 8 of 9 patients the lesions affected the structures of orbit by protruding into its lumen. In 5 cases it caused compression on the orbit muscles, in 4 – protrusion of an eyeball, and in 3 – compression of optic nerve (table 2) (fig. 2).

Lesions within paranasal sinuses and nasal cavity were found in 6 patients – for layout see table 3.

Protrusion into the intracranial space with compression of cerebral structures was observed in 3 patients (table 4).

Dysplastic lesions were visible on CT images as areas of high density corresponding with sclerotic density of osseous

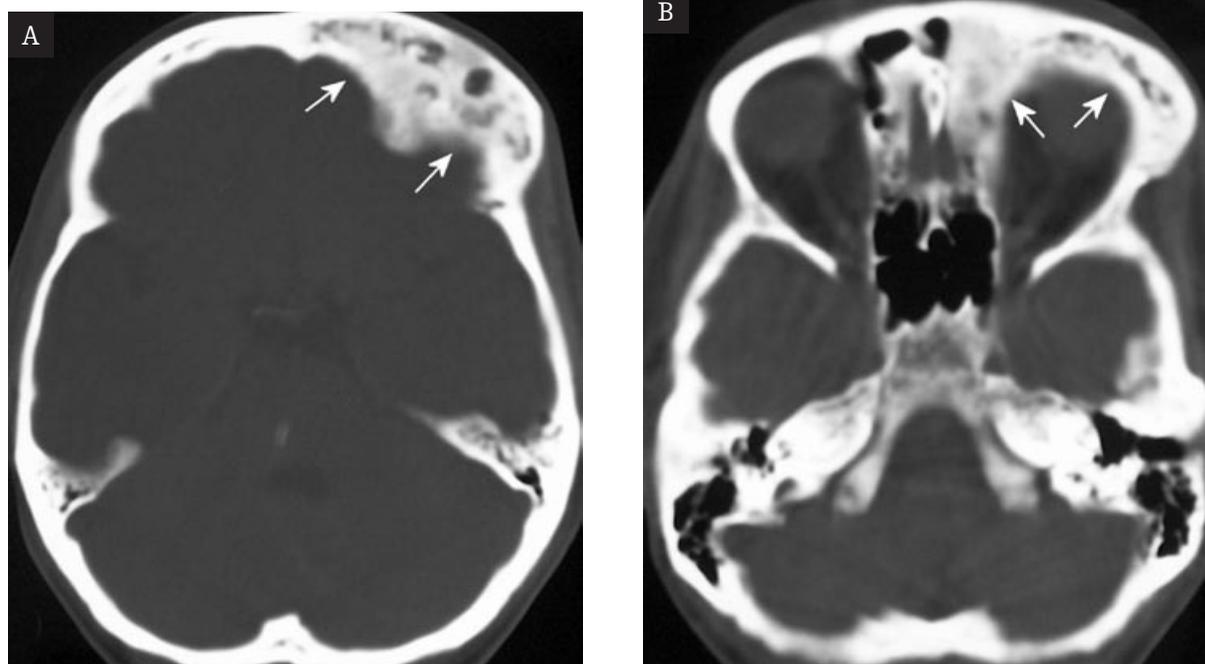


Figure 1. Patient Z.T., 43 y.o. Axial CT images. There is a large area of inhomogeneous density involving the lower part of the frontal bone, frontal sinus and ethmoid cells on the left side (arrows).

Table 2. Characteristics of the lesions in the orbital region.

Type of lesion	Number of patients (n=8)
Thickening of contours of the orbital bones with protrusion into the lumen of orbit	8
Compression and modellation of orbital muscles	5
– lateral rectus muscle	4
– inferior rectus muscle	1
Protrusion of an eyeball	4
Compression and displacement of optical nerve	3

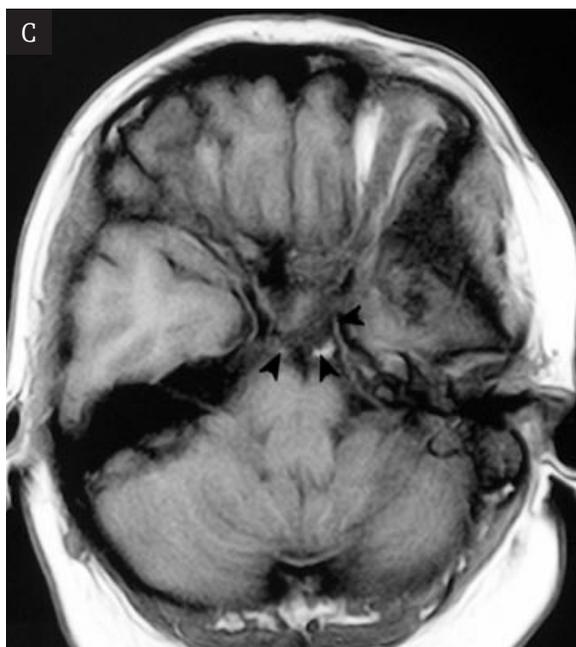
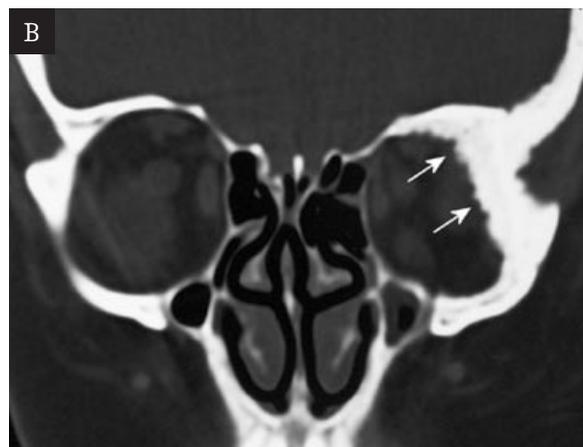
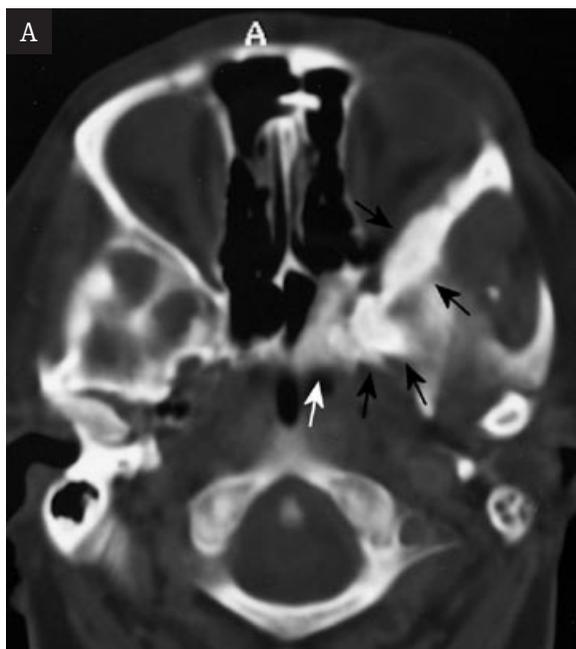


Figure 2. Patient D.B., 51 y.o. Axial (A) and coronal (B) CT images. Thickening and sclerosis of lateral and upper walls of the left orbit and the walls of the left sphenoid sinus with protrusion of lesions into the left orbit and left sphenoid sinus (arrows). MR, axial T1-weighted pre- (C) and post contrast (D) images. Lesions which were visible on CT enhanced after intravenous injection of Gadolinium (arrowheads).

Table 3. Lesions in the nasal cavity and sinuses.

Lesions within sinuses	Number of patients (n=6)
Sphenoid sinus	4
Ethmoid sinus	4
Maxillary sinus	3
Frontal sinus	2
Protrusion of abnormal tissue into the nasal cavity	3

tissue (fig. 3, 4). Although on MR they had low signal in all sequences, after intravenous contrast agent administration they were slightly enhanced (fig. 5).

Follow-up examinations of 2 patients revealed progression of the lesions. Both patients underwent surgical treatment; diagnosis of FD in these cases was confirmed by histopathologic examination. The follow-up CT and MR examinations of other patients showed no significant differences compared to the previous studies.

Discussion

Conventional radiography is the basic examination in radiological diagnostics of FD but due to the limitations of this method (visualization of the bone lesions only, limited number of views) it does not allow complete evaluation of the dysplastic lesions.

Daffner et al. emphasize the significance of assessment of FD lesions in CT examination in patients in whom the lesions lead to compression of the orbital soft-tissue structures and protrusion of the eyeball, what concerned about 35% of FD patients in their material [8]. Therefore, as far as FD diagnostics is concerned, assessment in both – bone and soft tissue window is obligatory. Evaluation of the bones enables precise estimation of the extent of the process: borders, location and size of pathologic foci as well as assessment of continuity of cortical bone what can be considered as the feature distinguishing from neoplastic processes. The aim of the soft-tissue window examination is to evaluate state whether the lesions involve extraosseous structures (orbits, paranasal cavities, intracranial space) or not.

The possibility of 3D reconstruction is an additional advantage of the CT examination, what is especially useful in planning surgical procedures.

Brown et al. mention 3 forms of FD cranial lesions observed on CT images – lesions of homogeneously higher density-osteosclerotic (23%), lesions of lower density-cystic (21%) and mixed, non-homogenous lesions, the so-called “ground glass” appearance (56%) [9]. In our material FD lesions in examined patients were in most cases the “ground glass” type.

However, the aforementioned forms do not have a characteristic CT pattern and require differentiation from other pathologic lesions such as: the Paget’s disease, inflammatory lesions of bones or neoplastic process. Tehranzadeh

Table 4. Intracranial lesions.

Intracranial lesions	Number of patients (n=3)
compression of brain, including:	3
Frontal lobes	2
Temporal lobes	1

et al. made an attempt to define the distinctive features of FD lesions and the Paget’s disease on CT images. According to them the FD is characterized by “ground glass” lesion, asymmetry of cranial bone infiltration and involvement of paranasal sinuses. Another signs of FD include: involvement of the sphenoid bone, orbit and nasal cavity, presence of soft-tissue mass, involvement of maxillary bone and associated cystic lesions in clivus [10]. Our material confirms the asymmetrical localization of FD lesions within the cranial bones, frequent involvement of sphenoid bone, orbital structures and paranasal cavities.

In cases of differentiation between FD and neoplastic processes the aforementioned assessment of signs of bone destruction is important and the CT examination needs to be extended with an exam with contrast medium administration in order to show possible pathologic enhancement within the lesion. Kumar et al. claim that the FD lesions in craniofacial area to be well-vascularized, can be strongly enhanced with contrast medium and cause spontaneous recurrent bleedings [11]. In one of our patients the lesions led to bone destruction and were intensively enhanced after contrast medium administration. The patient underwent surgical procedure due to fast progression of changes and symptoms caused by compression of intraorbital structures. In case of that patient, the FD diagnosis was confirmed with histopathologic examination.

The MR examination is usually used to define the lesions within the soft tissues – to evaluate their compression and displacement caused by pathologic dysplastic tissue, also in case of suspicion of other etiology of lesions and their atypical location [3, 5, 12, 13].

The MR signal of dysplastic lesions is usually hypointense on T1 and T2-weighted images, as in the cases we examined. With such pattern, the interpretation of the lesions does not cause problems, especially in cases of coexistence of sclerotic lesions on plain films or CT examinations. The assessment of etiology of the lesions is complicated when their signal differs from the mentioned above and the location of lesions is not typical for FD.

One can find reports on FD cases in which the signal was hypo- or isointense on T1-weighted images and hypo-, iso- or hyperintense on T2-weighted images [5, 6, 14, 15]. Jee et al. compared atypical manifestation of FD with the result of histopathologic examination and found that the lesions with stronger T2 signal are built of smaller number of bone trabeculae, collagen and cellular elements [6]. Chong et al. also claim that the hyperintense signal of FD lesions on T2-weighted images corresponds to non-mineralized areas and cystic lesions found in CT. They also suggest

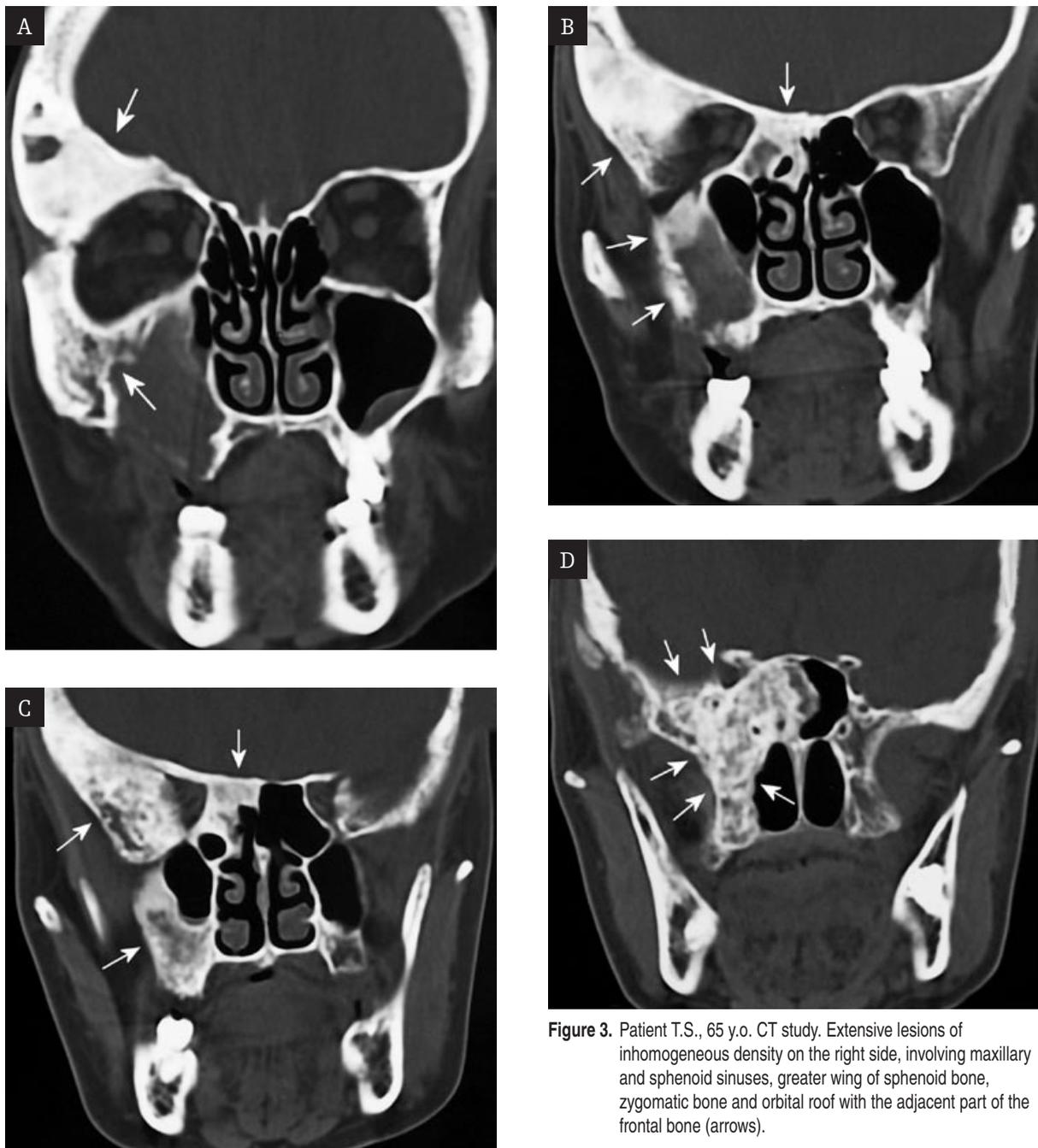


Figure 3. Patient T.S., 65 y.o. CT study. Extensive lesions of inhomogeneous density on the right side, involving maxillary and sphenoid sinuses, greater wing of sphenoid bone, zygomatic bone and orbital roof with the adjacent part of the frontal bone (arrows).

verification of this finding with fat saturation sequence in case of higher signal on T1-weighted images, as these regions usually correspond to bone marrow [5].

Resnick emphasizes the correlation between the FD lesions signal intensity and the proportion of fibrous tissue to calcified matrix. According to his experience the hypointense lesions on T1-weighted images are histopathologically built mainly of the richly calcified matrix, while the isointense lesions contain a lot of fibrous tissue. The image of lesions in T2 sequence shows higher variation of signal. He observed lower signal in well-mineralized lesions and increased signal in lesions rich in fibrous tissue and cystic changes [16].

FD lesions of atypical MR image and unusual location can be misdiagnosed as inflammatory or neoplastic process, especially if they visibly enhance after contrast medium administration [17]. However, Resnick emphasizes that, unlike the scar tissue, the fibrous tissue in FD is metabolically active and well-vascularized, what explains why it is relatively sensitive to contrast enhancement [16]. The MR of our patients showed moderate contrast enhancement.

Chong et al. believe that the certainty of diagnosis is high when the lesions show low signal on T1 and T2-weighted images, even after contrast medium administration [5]. In case of different, atypical signal and contrast enhancement, the verification of CT appearance of lesions ought to be decisive [3, 5].

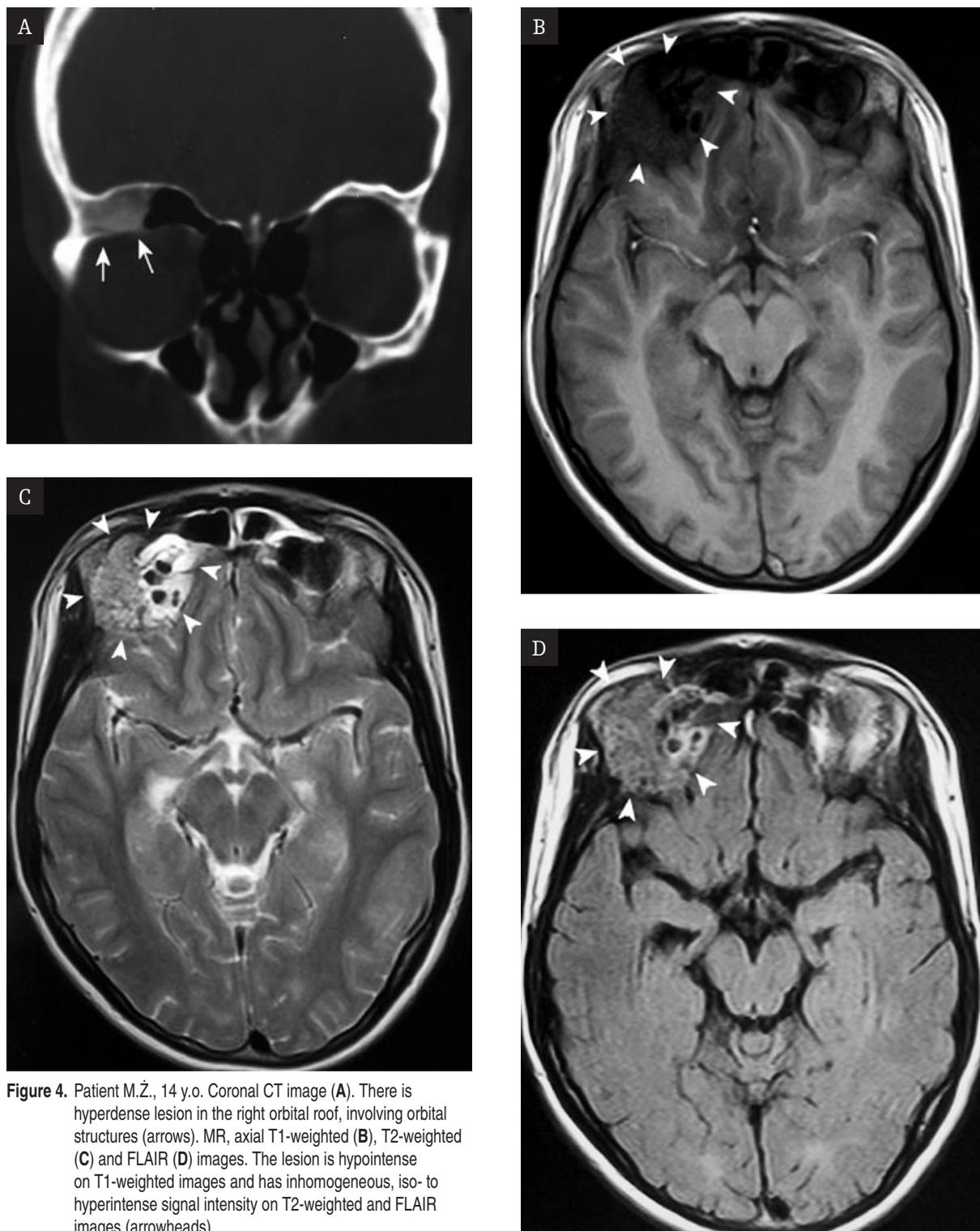


Figure 4. Patient M.Ž., 14 y.o. Coronal CT image (A). There is hyperdense lesion in the right orbital roof, involving orbital structures (arrows). MR, axial T1-weighted (B), T2-weighted (C) and FLAIR (D) images. The lesion is hypointense on T1-weighted images and has inhomogeneous, iso- to hyperintense signal intensity on T2-weighted and FLAIR images (arrowheads).

Sometimes it is necessary to follow-up the changes and evaluate the possible progression of the disease – in such a case the advantage of MR over plain films or CT is its lack of radiation and the possibility to study the soft-tissue structures. Monitoring the orbital lesions is particularly significant because the compression of optical nerve leads to sight disturbance, while the displacement of external eye muscles and protrusion of the eyeball causes disorders of its position and settle-

ment, what can lead to double vision etc. Lesions diagnosed too late can result in irreversible unilateral blindness. In cases of advanced FD lesions within the cranial bones it is necessary to visualize the compression of brain which can cause neurological focal symptoms. Moreover, the MR shows precisely the protrusion of abnormal tissue into the nasal cavity and paranasal sinuses what is important as it can cause airlessness, patency and secretion outflow disturbances of the involved sinuses.

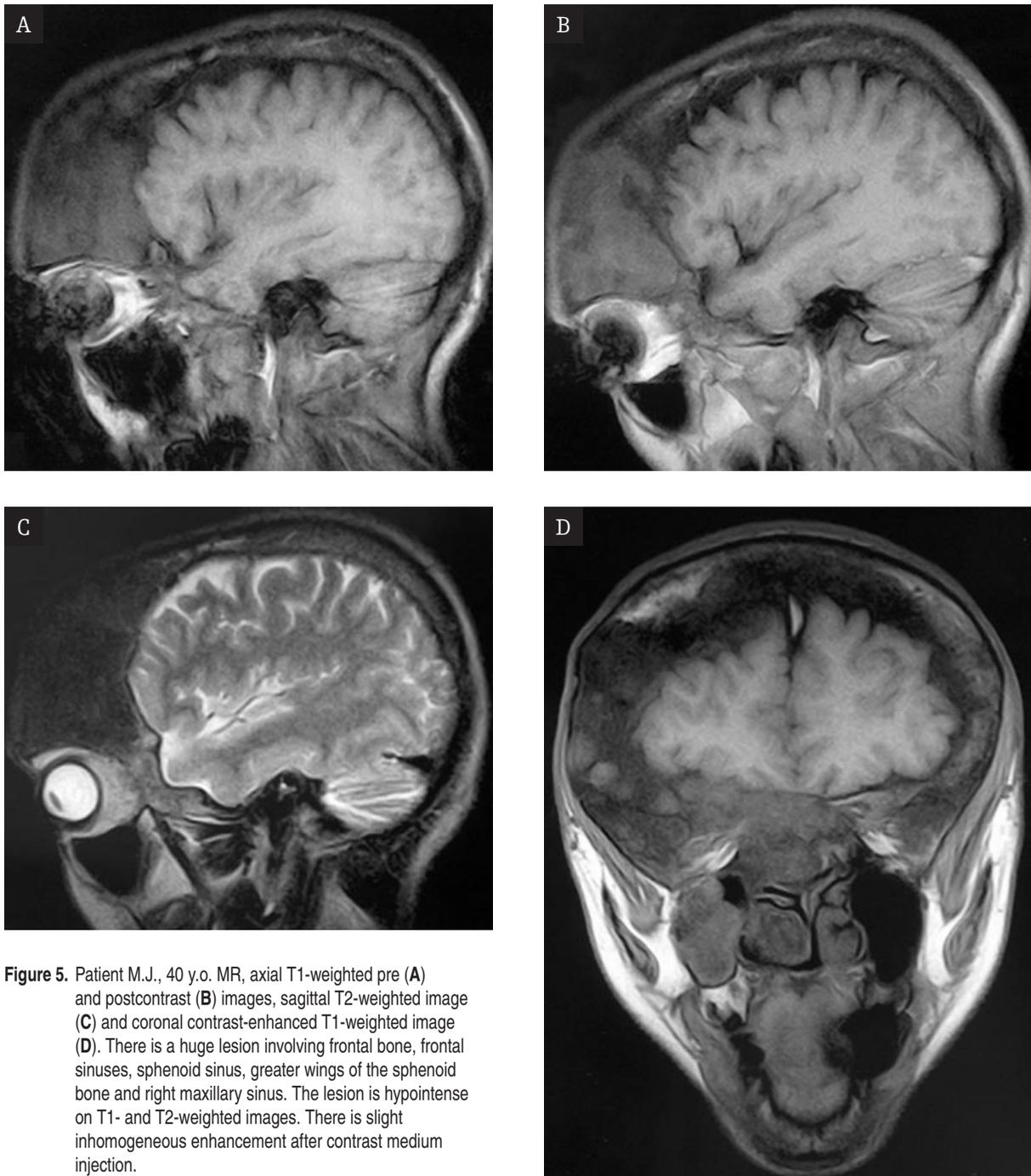


Figure 5. Patient M.J., 40 y.o. MR, axial T1-weighted pre (A) and postcontrast (B) images, sagittal T2-weighted image (C) and coronal contrast-enhanced T1-weighted image (D). There is a huge lesion involving frontal bone, frontal sinuses, sphenoid sinus, greater wings of the sphenoid bone and right maxillary sinus. The lesion is hypointense on T1- and T2-weighted images. There is slight inhomogeneous enhancement after contrast medium injection.

The treatment of FD patients is usually conservative. Surgical treatment is applied only in cases of increasing deformation, symptomatic compression of nerves and intracranial structures or malignant transformation of the lesion [3, 7].

CT and MR played an important role in our study – in diagnosing as well as in evaluation of the extent of lesions in course of FD. Among 9 patients only 4 were referred to examinations with FD diagnosis, while the remaining were diagnosed on the basis of CT, MR or both of them. In 3 cases malignant infiltration was suspected clinically but imaging examinations allowed ruling out such diagnosis.

Our study corroborates high frequency of orbital structures being involved with dysplastic lesions as they were observed in 8 of 9 patients. CT examinations, especially the MR, enabled a precise evaluation of the compression of the soft-tissue orbital structures found in most of our patients.

Moreover, CT and MR examinations enabled us to precisely define the involvement of particular craniofacial bones and paranasal sinuses, what can be significant when planning the possible reconstruction operations. It is important to visualize the compression of the brain structures which was observed in 3 of 9 patients and to exclude the coexistence of other lesions within the intracranial space.

Conclusions

On the basis of our material it can be stated that CT and MR studies provide much better visualization of lesions in the course of FD than the plain films, better estimation of the extent of lesions and their relation to orbital structures and intracranial space. They are also significant for

differential diagnosis in order to exclude other pathological processes which can occur within the craniofacial area and clinically can resemble the FD (neoplastic process, Paget's disease, bone inflammation). The CT and MR ought to be considered as complementary methods which play crucial role in assessment and monitoring the lesions as well as in qualification to the surgical treatment.

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