Cortical fibrous defects and non-ossifying fibromas in children and young adults: The analysis of radiological features in 28 cases and a review of literature

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Summary

Background: To assess and describe the variability of radiological presentations of fibrous cortical defects and non-ossifying fibromas in children and young adults.

Material/Methods: Medical records of 28 patients (15 males, 13 females, mean age of 17 years) with a radiological diagnosis of cortical fibrous defect or non-ossifying fibroma were reviewed retrospectively. The presentation of the lesion, its location and morphology according to Ritschl’s classification, as well as the number and types of imaging studies performed in the study group were assessed.

Results: Almost all lesions constituted an incidental finding discovered on plain films performed due to trauma. One lesion presented with a pathological fracture. There were 4 patients (mean age of 11 years) with stage A lesion, 9 patients (mean age of 16 years) with stage B lesion, 10 patients (mean age of 18 years) with stage C lesion, and 5 patients (mean age of 23 years) with stage D lesion. The lesions were located mostly in bones around the knee joint. In more than a half of the patients, further imaging was performed apart from plain films. Four lesions were biopsied (1 of stage B and 3 of stage C).

Conclusions: A considerable morphological variability of cortical fibrous defects and non-ossifying fibromas, especially of stage C, seems to be the main cause of unnecessary additional imaging and invasive diagnostic procedures in patients with this benign pathology. The knowledge of their age-related evolution and typical skeletal distribution should help in making a correct diagnosis.

Key words: fibrous bone tumors • imaging

PDF file: http://www.polradiol.com/fulltxt.php?ICID=882142

Received: 2011.06.08
Accepted: 2011.08.11

Background

Fibrous cortical defects and non-ossifying fibromas constitute the most common focal lesions found in bones [1,2]. Due to their benign character and typical radiological appearance, they belong to a group of lesions that do not require histopathological sampling for diagnosis (don’t touch lesions) [3]. Despite this fact, their detection in radiological examination with surprising frequency causes unjustified concern, and leads to consultations in orthopaedic oncology clinics [4]. It seems that this may be due to the variability of their presentations related to age and evolution of the lesions. The aim of this work is to assess and present radiological morphology of these lesions in older children and young adults.

Material and Methods

We assessed retrospectively cases consulted in our Unit from January 2006 to December 2009. The aim of research was to find medical records of patients:
1. who were referred for an evaluation of focal lesions in bones,
2. with diagnosed lesions fulfilling radiological criteria of a fibrous cortical defect or non-ossifying fibroma,
3. or with an equivocal radiological picture and available results of histopathological examination, confirming the nature of lesion.

The above presented criteria were met by 28 patients (15 males and 13 females, aged from 7 to 27 years, with a mean age of 17 years). In 26 patients, the lesions fulfilled radiological criteria completely; in 2 cases, the radiological image was equivocal and the type of lesion was confirmed with histopathological examination. Histopathological results were available also for 2 patients with a classic radiological image.

In all cases, the following was assessed: the circumstances of lesion detection, lesion location, morphology on X-rays with the use of classification proposed by Ritschl et al. (Figure 1), and the number and types of highly specialised imaging examinations performed [5].

**Results**

Radiographs were available for evaluation in all cases. In 27 cases, they were performed following trauma, and in 1 patient due to a chronic pain syndrome of knee joints. In one patient pathological fracture was diagnosed. Moreover, in 12 patients (43%) a CT examination was available for assessment, an MRI in 7 patients (25%), and a bone scan.
in 2 cases (7%). In total, highly specialised diagnostic imaging was performed in 16 patients (57%).

Stage A lesions were found in 4 patients (age of 7–14 years), stage B in 9 (12–19 years), stage C in 10 (13–24 years), and stage D in 5 (21–27 years). A mean age of patients with stage A lesions was 11 years, stage B – 16 years, stage C – 18 years, and stage D – 23 years. Epidemiological data of patients, morphological classification of lesions, and information concerning the performed highly specialised imaging examinations were all presented in Table 1.

The lesions were most commonly located within the knee joint (proximal tibia n=9, distal femur n=7) and in distal tibia (n=7). Other locations were less frequent (proximal femur n=2, one case of a lesion in femoral shaft, one in proximal humerus, and one in proximal fibula).
Discussion

Fibrous cortical defect and non-ossifying fibroma belong to the most common focal lesions in bones. It is estimated that they may be present in up to 30% of the asymptomatic population in the first and second decade of life [1,2]. They are not neoplasms and, according to WHO, belong to the group of developmental abnormalities. Similar to osteocartilaginous exostoses, they develop in the metaphysis, in the region of intensive bone growth [6]. According to some authors, an injury in the area of muscle attachment with a focal, subperiosteal haemorrhage may lay at the background of these lesions [1,5]. Both lesions are histopathologically identical, composed of benign spindle-shaped cells (fibroblasts) and histiocytes, with a scattering of xanthomatosic cells [7]. As they do not cause clinical symptoms, they are usually incidentally found in X-ray examinations performed because of an injury, which is in accordance with our findings. Only in one patient, the non-ossifying fibroma was manifested by a pathological fracture caused by a minor injury during football play. In the remaining cases, the pathology was discovered incidentally. Despite the proven benign nature of these lesions, finding them during diagnostic imaging still causes anxiety, making them some of the most common causes of referral for consultation at the clinics of orthopaedic oncology [4].

It is believed that an X-ray image is sufficient for the diagnosis of a fibrous cortical defect and non-ossifying fibroma and it is so typical, that extending the diagnostics or acquiring material for histopathological examination are unnecessary or even inadvisable (don’t touch lesions) [3]. A classic X-ray picture of a fibrous cortical defect shows a cystic lesion located in the cortical layer of a bone, usually oval, surrounded with a thin sclerotic rim, with a long axis parallel to the axis of the bone [8,9]. The external outline of the cortical layer at the level of the lesion may be poorly visible or completely invisible. Non-ossifying fibroma is a larger lesion, with polycyclic borders, protruding into the medullary cavity. However, this differentiation is a bit unnatural, due to the already mentioned fact that these lesions are histopathologically identical (Figure 2) [7,10]. The diagnosis is also facilitated by a typical location of the lesions, which was confirmed in our study. The lesions are usually found in the metaphyseal areas, mainly in bones constituting the knee joint [1,2].

Diagnosis may be complicated by the variability of radiological presentations of these lesions, which are biologically active, may grow and, in their involution phase, somewhat contrary to its name, gradually become filled with bone tissue [5]. Evolution of non-ossifying fibromas was described by Ritschl et al., who developed a classification distinguishing four stages of the lesion [5]. Stage A describes a small, oval lesion, adjacent to the growth plate. As the bone grows, the lesion moves toward the metaphysis and may increase in size, exhibiting more polycyclic, grape-shaped borders (stage B). Subsequently, it fills with bone tissue; mineralisation tends to start in the shaft and proceeds toward the growth plate (stage C). Stage D is a completely calcified lesion resembling a large bony islet (Figure 1). The pictures of the lesions may therefore vary. Our gathered material was dominated by stage C lesions (n=10), gradually filling with bone tissue, and stage B lesions (n=9). This is most likely associated with patients’ age, as our group consisted mainly of patients in their second and third decade of life. According to our results, individuals in their third decade of life are expected to have almost exclusively calcified stage
Figure 3. MR and scintigraphic appearance of non-ossifying fibroma. 
(A,B) MRI of the lesion in Figure 2B. The lesion reveals a heterogeneous, low signal intensity on T1-weighted images (A) and enhances after IV contrast medium administration (B). (C–E) A 21-year-old male. In the proximal femoral epiphysis, there is a well-delineated, non-uniformly sclerotic lesion (C,D). On bone scan, a slightly increased accumulation of a radioisotope within the lesion (E) (courtesy of Joanna Mażewska, MD, Department of Nuclear Medicine, Medical University of Warsaw). Biopsy: non-ossifying fibroma.
C and D lesions. Biological activity is reflected by contrast enhancement present in MRI and increased radiotracer accumulation in bone scan (Figure 3) [11,12]. These symptoms should not raise concern. Bone scan picture depends on the stage of evolution of a lesion. Accumulation of the tracer is usually mild or moderate, but it may be more intense in cases complicated by a fracture [12,13].

Multiple lesions, which can be found in about 8% of patients, should not cause concern either [14]. They tend to focus around knee joints. Sporadically, they may be manifestations of metabolic or developmental disorders. Their atypical location (e.g. in flat bones or shafts of long bones) may raise suspicion of hyperlipidaemia [15]. In the Jaffe-Campanacci syndrome, multiple non-ossifying fibromas coexist with neurofibromatosis and café-au-lait spots on the skin [16].

In more than a half of presented cases, following the finding of the lesions on X-ray, the diagnostic process was
extended and included CT, MRI or a bone scan. It seems unjustified, both from the medical, and economic point of view. CT provides us with exactly the same information as plain radiography, but exposes the patient to much higher doses of ionising radiation (Figure 4). Changes in MRI signal are also not characteristic [11]. As mentioned previously, finding of contrast enhancement in MRI or an increased radiotracer accumulation in bone scan is typical and does not point to an aggressive or malignant nature of lesion. Extension of imaging diagnostics seems justified only in cases of lesions qualified for surgery. This concerns large lesions located in weight-bearing bones that are at risk of fracture (Figure 5). The method of choice in such cases is computed tomography, allowing for a detailed evaluation of the size of a bone defect, together with a volumetric assessment. Another indication for surgery is a sporadic rickets/osteomalacia syndrome associated with the tumour [17]. In rare cases, non-ossifying fibroma may produce endocrine substances blocking the activity of vitamin D and only resection of the lesion may cure vitamin
D-resistant rickets/osteomalacia. In our material, imaging diagnostics was extended mainly in case of stage C lesions, although it happened a few times that highly specialised imaging examinations, including CT, were used for absolutely typical stage A and B lesions (Table 1). Biopsy was carried out in one patient with stage B lesion (in our centre) and in three patients with stage C lesions (outside of our centre in all cases) (Table 1). In case of the patient with stage B lesion, the collection of histopathological material was justified by a very large size of the lesion, which required differentiation with fibrous displasia (Figure 6). In case of the patient presented in Figure 3 biopsy could be justified by an atypical location of the lesion in the shaft of a long bone. In the remaining two cases classic radiological presentations of stage C lesions were found (Figure 7).

As mentioned before, radiological appearance of fibrous cortical defect and non-ossifying fibroma is so typical in the majority of cases, that differential diagnoses should not be included in the examination report [3,7]. A cortical desmoid is a similar lesion, that may be found in boys and young men. It is located at the attachment of the medial head of the gastrocnemius muscle to the medial femoral epicondyle. The lesion may present a more aggressive appearance than the fibrous cortical defect, with irregular, interrupted borders of the cortical layer. In case of particularly large, especially in craniocaudal dimension, non-ossifying fibromas, the differential diagnosis should include fibrous displasia. Long lesions located in the anterior cortical layer of the tibia require more caution and they should be differentiated with ossifying fibroma and ameloblastoma, which is a malignant tumor.

Conclusions

A high variability of radiological presentation of fibrous cortical defects and non-ossifying fibromas, especially in case of stage C lesion, of mixed cystic and osteosclerotic type, seems to be the cause of performing unnecessary additional diagnostic imaging and even invasive diagnostic procedures in patients with benign lesions. The knowledge of age-related evolution of the radiological picture of the lesions, their typical distribution, as well as patients’ history pointing to the incidental detection of the lesions should be helpful in stating a correct diagnosis.

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