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CHONDROMYXOID FIBROMA OF THE FEMUR IN 9-YEAR OLD BOY (CLINICAL CASE)

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The article reflects the experience in the diagnosis and surgical treatment of chondromyxoid fibroma in the proximal metadiaphysis of a large femoral bone in a child in the first decade of life. We presented the family and medical history of the child complaints, orthopedic/local status, and the results of radiological methods and emphasized difficulties of nosologic identification and differential diagnosis of the bone destructive process at the stage before biopsy and surgery. Macroscopic findings during surgery were identified, and the amount of resection of the affected bone was determined. A detailed description of the results of pathological and morphological (macro- and microscopic) studies is given, which could confirm the tumor process. Late results with the absence of oncological re-occurrence (7 years) and good anatomical and functional results from orthopedic surgery are shown.

Keywords: children, chondromyxoid fibroma, diagnosis and treatment.

Chondromyxoid fibroma of the bone is a rare tumor, accounting for less than 1% of all bone tissue neoplasms and less than 2% of all benign bone tumors [1, 2]. Chondromyxoid fibroma was differentiated from chondrosarcoma by American morphologists Jaffe and Lichtenstein in 1948 [3]. Although called "chondromyxoid fibroma," this entity is not entirely fibroid in its histological structure. In 1972, WHO [4] classified it under benign chondrogenic neoplasms. The synonyms that most objectively reflect the morphological structure of the tumor, and its cartilage genesis, are "fibromyxoid chondroma" and "myxoid chondroma."

Although this tumor usually presents in the second and third decades of life cases of affected children, including smaller children, have been reported [1, 2, 5]. The most common sites of the tumor is the metaepiphyseal segment of the long bones of the lower extremities (tibia, femur, and fibula), but the tumor can arise in the pelvis, ribs, vertebrae, short tubular bones of the hands and feet, as well as tarsal bones.

In both domestic and foreign guidelines for bone disease management, chondromyxoid fibroma is presented clinically and radiologically as a progressive bone destructive process that can not only recur but also become malignant in case of incomplete surgical resection [1, 5, 6, 7]. However, some studies [8, 9] reported the possibility of a less aggressive disease course, and indicated that in that the absence of persistent pain, the radiological findings does not always suggest chondromyxoid fibroma as part of the differential diagnosis. As a consequence, identification of the tumor before biopsy is very difficult, particularly in cases of subperiosteal localization [8, 10, 11].

Knowledge of tumor histology allows pathologists to verify bone destructive processes accurately. The cartilaginous nature of chondromyxoid fibroma is indicated by the following specific morphological features: lobulated structure; a certain polymorphism of the basic cellular composition, organized in one histogenetic row; and characteristic interstitial substance of myxoid type [1, 2, 5, 12, 13]. A histological feature of chondromyxoid fibroma that distinguishes it from chondrosarcoma is presence of a zoned structure in each tumor tissue slice, where the central part of a slice is relatively hypocellular and the periferal parts are distincly hypercellular. Immunohistochemical studies on chondromyxoid fibroma revealed the presence of high activity of the protein S-100, thereby confirming the cartilaginous/chondroid immunophenotype [1, 2, 13].

There is a consensus regarding the treatment of this tumor: radical surgery, which eliminates con-

tinuous growth of tumor tissue [5, 6, 7, 9, 10]. This is achieved by carrying out boundary or segmental resection of the affected bone. The usage of curettage (excochleation or so called intralesional resection) of the tumor is known to ensure the occurrence of relapse in 15–25% of the cases [1, 2, 9].

In this article, we share our experience in the diagnosis and surgical treatment of a large femoral chondromyxoid fibroma in a 9-year-old child, whom we were able to follow-up postoperatively for 7 years and who has been disease-free since surgery.

Clinical case

On August 21, 2007, a 9-year old boy was brought to the diagnostic department of FGBI "Research Pediatric Orthopedic Institute of G.I. Turner" by his parents because of pain in his left thigh radiating to the knee joint. There was swelling in the upper third of the left thigh, and he was limping on his left leg. The pain was primarily on exertion and rarely at rest; active pain was absent at night. After the onset, pain usually disappeared on its own, and the patient did not often require analgesics. The pain had started in March 2007. Over time, the frequency and intensity of pain continued to increase. In May, the patient started presenting a limp on the left lower limb; and in July, the parents noticed that local swelling appeared at the upper third of the left thigh. The pain intensity clearly increased with exertion.

Prosthetic status at the time of treatment. The boy had an asthenic constitutional type and a proportionate physique. He walked without support, but limped on the left leg. The spine, chest, and upper extremities were normal. The axes of the lower extremities were physiological, and the length was the same. A mass involving the soft tissues of the left thigh and leg were obvious upon physical examination. The difference in the circumferential length (D > S) at the level of the medium third of the thighs and shins was 4.5 cm and 2.0 cm, respectively. Clinically, marked functional limitations were observed in the left hip: mainly, limited flexion (90°), abduction (15°), and external rotation (10°); internal rotation was absent. We attempted to perform a passive motion of the left hip joint beyond these limits, but the maneuver caused the pain in the left hip. Full range of motion of the right hip, knee, and ankle joints was observed. Vascular and neurological disorders in the lower limbs were not observed.

Status localis. Visually, at the level of the upper third of the left thigh, in the projection of the greater trochanteric and subtrochanteric regions, mainly on the anterior outer surface, there was local swelling of the soft tissues without changes or vascular patterns on the skin. The dimensions of the swollen area were on anterior external surface of the upper third of the left femur were ~ 13.0 cm × 8.0 cm. The surface area of the hip was painless upon palpation. Upon deep palpation, the mass was palpable, non-mobile, and dense in consistency. It extended around the entire circumference of the affected femur. Deep palpation in some areas was accompanied by tenderness. We did not palpate any regional lymph nodes with increased size.

Radiographic findings

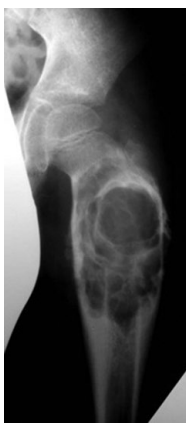
On radiographs of the hip joints, including the upper third of the thighs in AP view and in the position on Lowenstein dated September 6, 2007 (Fig. 1), a focus of the lytic destruction with clear sclerotic borders, 9.0 × 7.0 × 6.5 cm in size, was seen at the level of the proximal left femur metadiaphysis. It was accompanied by fusiform thickening of the bone and in some places, solution of continuity of its cortical layer. The internal structure of the pathological site on the radiograph in frontal projection was predominantly coarse, and in the position of Lowenstein, it was somewhat "blurred" or feathery, with clearly defined areas of cortical layer destruction. Neither periosteal spicular reaction, "bulbous" periostitis, nor soft tissue reactions were detected. On the border between the pathological site and diaphyseal bone, the periosteal overlay was a reflection of the previous stage of active tumor growth.

Computed tomography findings

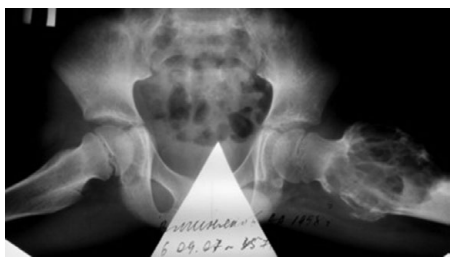
On computed tomography of the hip joints, including the upper third of the thighs (dated September 06, 2007) (Fig. 2), a high-intensity regional of osteoporosis was seen in the proximal left femur, 9.0 × 7.0 × 6.5 cm in size—considered as the focal point of the lytic lesion. It occupied the base

of the femoral neck and intertrochanteric and subtrochanteric regions, spreading to the proximal part of the diaphysis of the bone. Marked growth of the bone was noticeable with a sharp thinning of the cortical layer and local areas of destruction. Directly under the pathological site, on the border of the unaltered diaphyseal part of the bone, there was a significant thickening of the cortical layer secondary to the assimilated periosteal strata. The densitometric content of the pathological site was uneven: the distribution of the relative density of the contents of the pathological site ranged from 3.9 to 39.0 HU, including fat density.

On the basis of the patient's symptoms, anamnestic information, and radiological findings, the patient was diagnosed with an aneurysmal bone cyst of the proximal left femur metaepiphysis. The boy underwent segmental resection of the affected part of the left femur and combined autologous and alloplastic bone graft reconstruction with a metalosteosynthesis angled plate. Here is a brief excerpt from the minutes of the surgery: the proximal femoral metaphysis had a fusiform enlargement and uneven/lumpy cortical layer circuit. Upon subperiosteal detachment of the disease site, we identified the destruction of the outer part of the cortical layer with prolapse through its chondroid masses (Fig. 3). Similar areas were identified in the cortex and the anterior–posterior parts of the outer proximal metadiaphysis. Given the nature of bone destruction, it was decided to perform a segmental resection of the affected part of the bone. The length of the bone defect after segmental resection was 12.0 cm. We found thickened and infiltrated areas of the periosteum. A sample of a portion of the iliac crest was taken. The homolateral side was approached (3.5 × 1.8 × 0.5 cm in size) without separating the bone from the corresponding portions of the gluteus medius muscle. The resulting musculoskeletal complex was mobilized over sufficient length for adduction to the proximal fragment of the femur. The "beak" of the angled plate was intromitted to the femoral neck. The homologous graft (17 × 1.8 cm) was inserted in the medullary cavity of the distal part of the femur and was adapted to its extramedullary part ("leg"). The musculoskeletal autocomplex was inserted under the stump of the femoral neck. The plate and the aforementioned osteoplastic material were fixed with screws. In the post-resection defect, the allogeneic osteoplastic material was further adapted in the form of cortical alloplastic shafts and mixtures of crushed and demineralized cortical allografts (Fig. 4).



A



B

Fig. 1 Radiographs of the left hip joint in AP view and of both hips in the Lowenstein position. The radiographs (dated September 06, 2007) clearly showed the bone destruction at the level of the proximal left femur metadiaphysis.

A fragment of the left femur was resected en bloc (Fig. 5) and examined histopathologically. On cutting medially (Fig. 6), the lesion consisted of solid structures that appeared to be firmly packed in the affected part of bone.

Pathomorphological examination findings

Gross examination The surgical material was a one block fragment resected from the femur (overall femur dimensions 12.0 × 7.5–1.8 × 5.5 cm) (Fig. 5). There was a fusiform thickening on the metaphyseal end of the fragment, which then narrowed down concentrically to the normal diameter and shape of the diaphyseal femur up to the diaphyseal end. On the surfaces of the sample cut medially (Fig. 6), the tumors tissue is uneven, with a bluish-grayish color. The boundary between the tumor tissue and bone is clear, and the edges of the tumor site are jagged. The uneven grayish tumor tissue appears to be lobed. In some places, there are semi-translucent areas. Necrotic changes and/or melting portions and cystic structures are absent. On certain parts of the surface of the tumor, a subtle grayish membrane ("pseudocapsule"?) can be distinguished.

Histologic pattern. In the benign tumor (chondromyxoid fibroma) sections, lobular tumor (chondroid) tissue contains elongated and stellate dendritic cells (Fig. 7), with a clearly discernible oxyphilic cytoplasm and middle-sized, irregularly rounded or oval nuclei (Fig. 8). The cells are arranged in a bright, slightly basophilic matrix. In some lobules, these cells are located quite uniformly (Fig. 7). In other

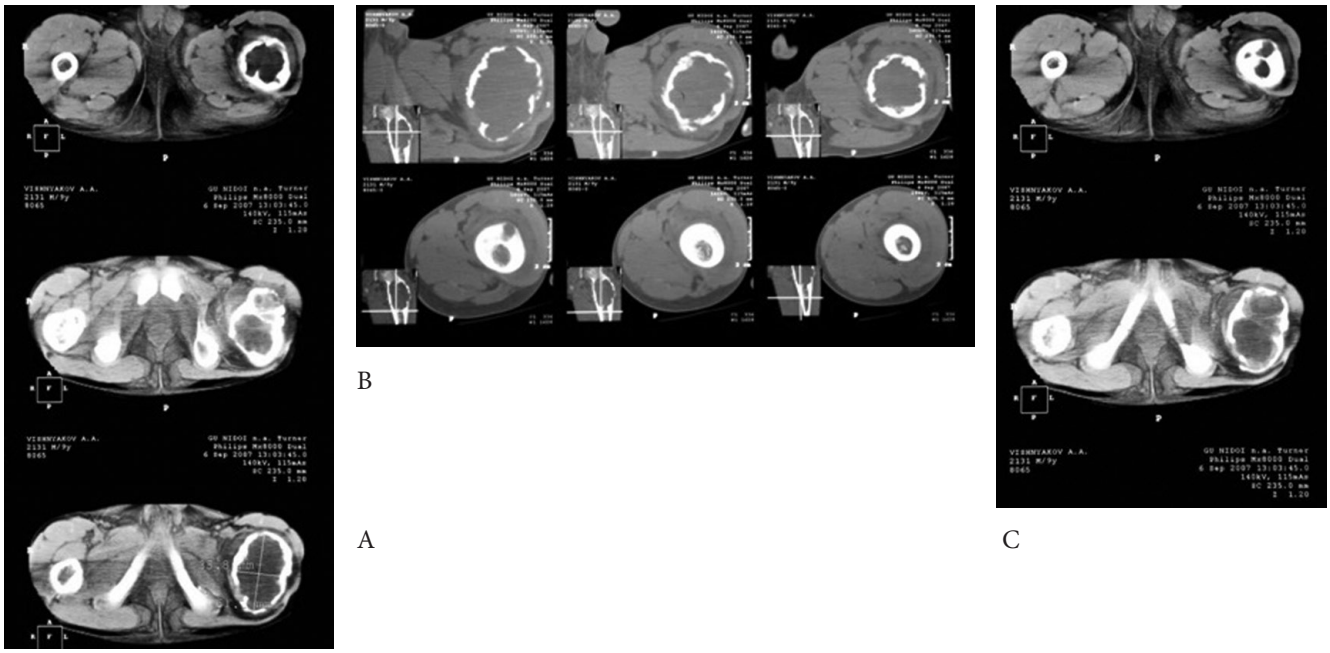


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Fig. 2 Computed tomography images including multi-level axial slices of the affected part of the left femur

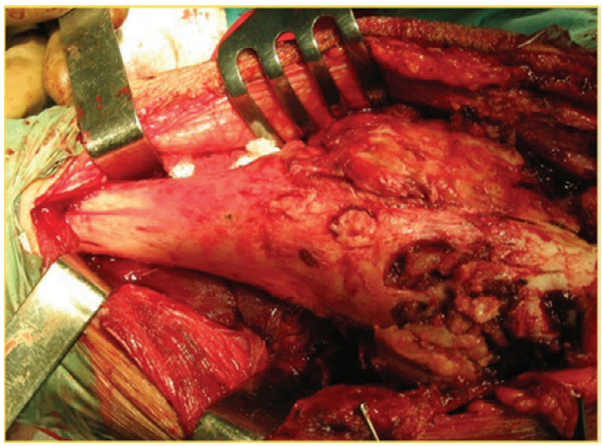

Fig. 3 Intraoperative photo shows the local destruction of the cortical layer of the proximal femur metadiaphysis with prolapse of the contents of the pathological site beyond the anatomical contour of the bone

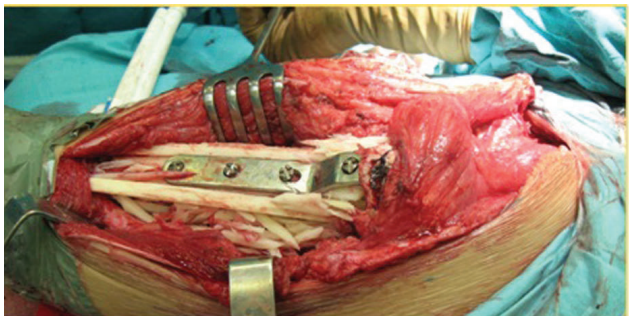

Fig. 4 Intraoperative photo that reflects the final stage of bone plastic surgery

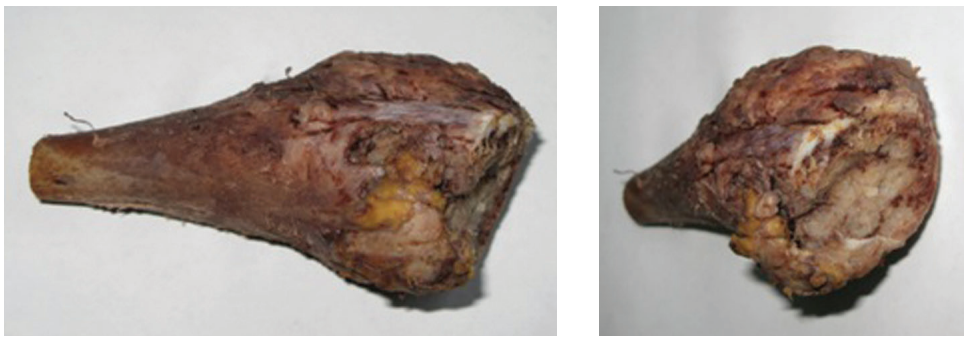

Fig. 5 Photo of gross specimen (resected en bloc from the affected femur)

lobules, multiple dendritic cells are located mainly around the periphery, whereas the central sections of the lobules are hypocellular with myxoid matrix, as well as focuses of dilution and/or melting (Fig. 9). Figures of mitosis (normal and/or pathological) could not be found in mononuclear cells of the studied material.

Often, variable number of multinucleated giant osteoclast-type cells can be seen distributed unevenly on the periphery of the lobules (Fig. 10).

Chondroid lobules are separated by fibrous layers formed in bundles of elongated oval and spindle-shaped cells with irregular presence of inflammatory cells (mainly lymphocytes). Often, small osteoid ravines of different shapes can be seen in fibrous tissue (Fig. 11).

The tumor is clearly separated from the unchanged bone by a thin fibrous pseudocapsule (Fig. 12).

Long-term results

Radiographs of the left hip joint (Fig. 13), taken 4 years and 4 months after reconstructive surgery on the proximal left femur affected by a benign tumor process. The image shows the results of the restructuring of osteoplastic material and the restoration of organotypic bone structure in the surgical site.

Dynamic observation of the patient ascertained growth retardation of the left femur. The quantity of shortening the left lower limb 6 years after the surgery (when the patient was 16 years of age) was 5.0 cm (Fig. 14).

This was considered to be indication for restoration of the length of the left thigh by distraction osteosynthesis. Therefore, on October 27, 2014, the teenager was admitted to our institution (case № 14/5769). At the time of the admission, the functional limitations of the left hip joint were minimal. The range of motion in the joint was as follows: flexion/extension, 90°/5°; abduction/ adduction, 35°/45°; and internal/external rotation, 30°/35°. On November 05, 2014, the patient underwent imposition of external fixation rod on the left thigh and left femur osteotomy at the level of the middle third of the diaphyseal part. The length of the segment was restored by the method of distraction osteosynthesis (Fig. 15). Currently, the patient is at the stage of "consolidation"/"maturation" of distraction regeneration of the femur.

Conclusion

Chondromyxoid fibroma is a rare, bone-destructive, benign tumor. The cases presented in this article demonstrate the absence of specific clinical and radiological symptoms, thus indicating the low probability of identifying the tumor before biopsy or surgery. We consider that the following factors allowed us to achieve stable relapse-free results as well as satisfactory anatomical and functional outcomes: radical resection, rational combination of autologous and alloplastic methods applied to the post-resection bone defect, stable metalosteosynthesis, and pronounced regenerative potential of bone during childhood.

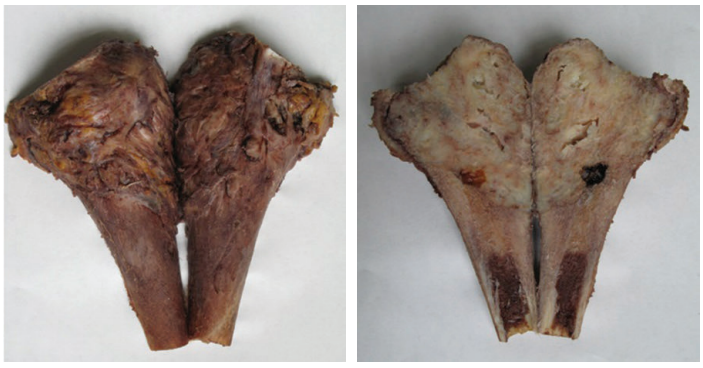

Fig. 6 Photo of gross specimen after it was cut medially

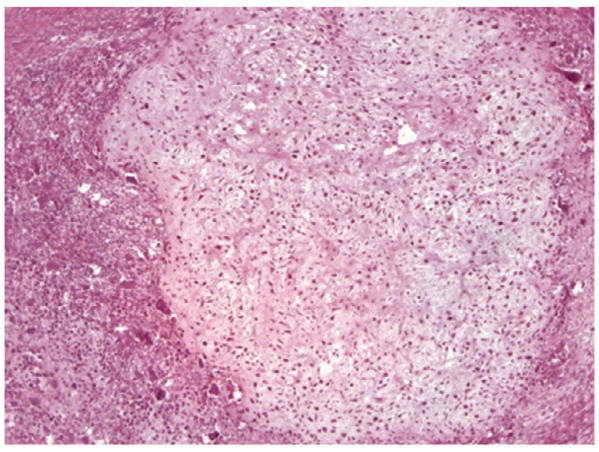

Fig. 7 A smaller chondromyxoid fibroma slice with relatively uniformly distributed dendritic cells in the intercellular matrix. Stain with hematoxylin and eosin (HE). Magnification ×150

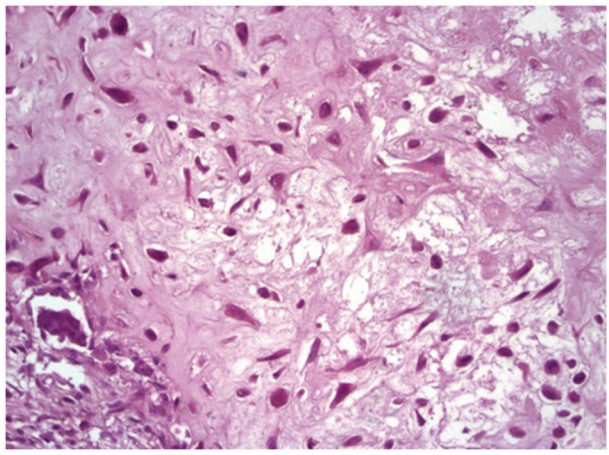

Fig. 8 Dendritic fusiform and stellate cells in a non-uniformly edematous (myxoid) extracellular matrix. Stain: HE. Magnification ×600

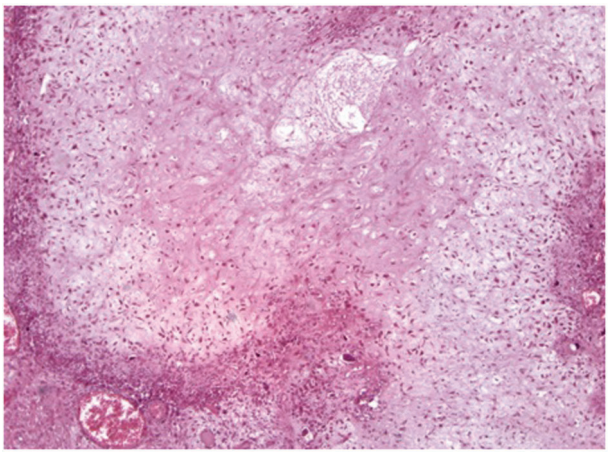

Fig. 9 Hypocellular central part of slice and hypercellular in its periphery. In the sample, there are irregularly pronounced myxoid changes and tissue dilution in the center of the slice. Stain: HE. Magnification ×150

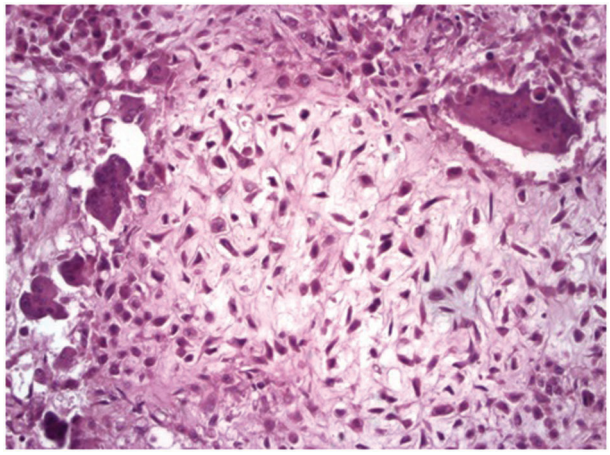

Fig. 10 Unevenly distributed multinucleated giant osteoclast-type cells of on the periphery of the lobules. Stain: HE. Magnification ×600

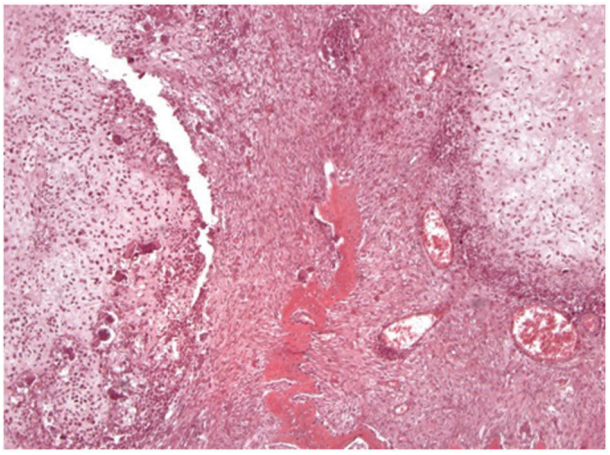

Fig. 11 Osteoid ravine in the fibrous tissue layer between the lobules of the chondromyxoid fibroma. Stain: HE. Magnification ×150

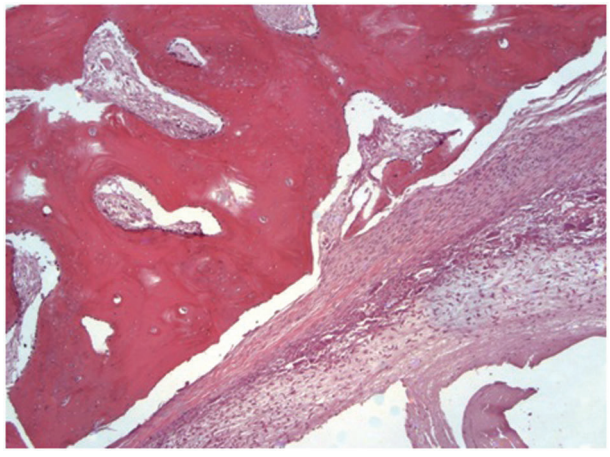

Fig. 12 Fibrous pseudocapsule, which separates tissue of chondromyxoid fibroma from unchanged bone tissue. Stain: HE. Magnification ×150

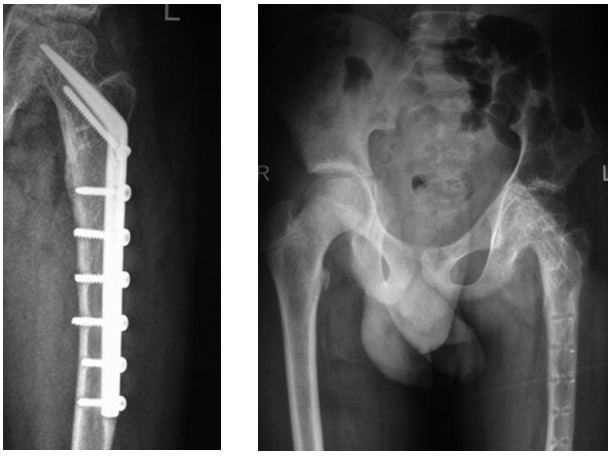

Fig. 13 Radiographs of the left proximal part of the femur before April 19, 2012 and after May 12, 2012. The deletion of the angled plate is observed (4 years and 4 months after segmental resection of the affected portion of the bone tumor)

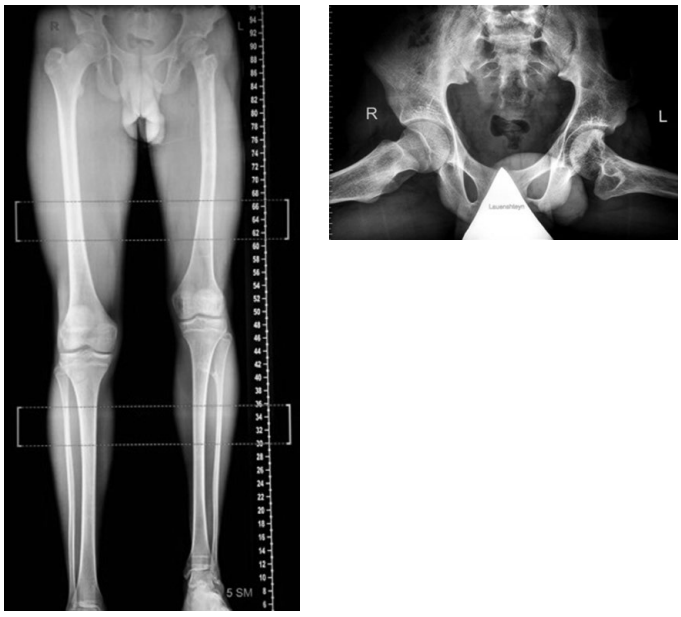

Fig. 14 Radiographs of patient B. on October 29, 2014, showing the full restoration of the organotypic structure of left femur in the area of surgery and shortening of the left lower limb owing to femoral segment shortening

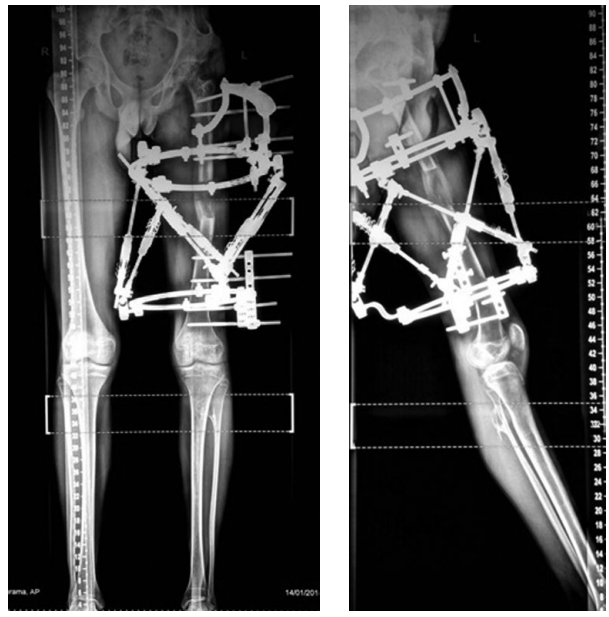

Fig. 15 Radiographs of patient B taken on January 14, 2015, at the final stage of distraction and correction of the axis of the left lower limb

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ХОНДРОМИКСОИДНАЯ ФИБРОМА БЕДРЕННОЙ КОСТИ У РЕБЕНКА 9 ЛЕТ (КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ)

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В статье отражен опыт диагностики и хирургического лечения хондромиксоидной фибромы проксимального метадиафиза бедренной кости больших размеров у ребенка первого десятилетия жизни. Подробно представлены анамнез заболевания с детализацией жалоб ребенка, ортопедический/локальный статусы, результаты лучевых методов исследования, подчеркивающие трудности нозологической идентификации и дифференциальной диагностики костно-деструктивного процесса на добиопсийном/дооперационном этапе. Запротоколированы макроскопические находки во время оперативного вмешательства, определившие объем резекции пораженного отдела кости. Дано подробное описание результатов патоморфологического (макро- и микроскопического) исследования, позволившего верифицировать опухолевый процесс. Продемонстрированы отдаленные (семилетний) безрецидивный онкологический и хороший анатомо-функциональный результаты ортопедохирургического лечения.

Ключевые слова: дети, фиброма хондромиксоидная, диагностика, лечение.

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