THE COEXISTENCE OF GIANT CELL TUMOR, PARA-ARTICULAR OSTEOCHONDROMA AND IDIOPATHIC THROMBOCYTOPENIC PURPURA (ITP) IN ONE PATIENT: A CASE REPORT

Nurettin HEYBELİ, Müjdat ÖK**, Fahri ERDOĞAN****

*Muharrem BABACAN*****

SUMMARY

Giant-cell tumor (GCT) of bone is a primary bone neoplasm which is most commonly seen around the knee joint. Curettage and bone grafting is the mainstay of treatment in GCT. However, local recurrence is a frequent complication. Osteochondroma is a common benign tumor, which arises from a developmental defect in the growth plate. A para-articular osteochondroma is a rare lesion with only a few reported cases in the literature. This case is presented as, in the literature survey, we were not able to find any case where a GCT and para-articular osteochondroma occurred in the same patient. Furthermore, a superimposed Idiopathic Thrombocytopenic Purpura was detected and treated before her second "primary" operation.

Key Words: *Giant-cell Tumor, Para-articular Osteochondroma, Idiopathic Thrombocytopenic Purpura.*

ÖZET

DEV HÜCRELİ TÜMÖR, PARAARTİKÜLER OSTEOKONEDROMA VE İDİOPATİK TROMBSİTOPENİK PURPURANIN (ITP) BİRLİKTE GÖRÜLMESİ: OLGU SUNUMU

Dev hücreli kemik tümörü sıklıkla diz çevresinde görülen primer bir neoplasmdır. Dev hücreli tümörün tedavisinde küretaj ve kemik grefti uygulaması yapılmaktadır. Bununla birlikte, lokal nüksler sık görülmektedir. Osteokondrom ise büyüme kıkırdağındaki bir gelişme defekti sonucu ortaya çıkan ve sık görülen selim tümörlerdendir. Fakat, literatürde sadece az sayıda olgu sunusu olarak bildirilmiş olan eklem çevresi osteokondromlarına nadiren rastlanmaktadır. Bu olgu, literatür incelememizde aynı hastada hem Dev Hücreli Tümör hem de osteokondrom lezyonlarına rastlamamız nedeniyle sunulmuştur. Bunun yanında, hastanın ikinci "primer" tümörünün ameliyatından önce İdiyopatik Trombositopenik Purpura tespit edilerek tedavisi gerçekleştirilmiştir.

Anahtar Kelimeler: *Dev Hücreli Tümör, Para-artiküler Osteokondrom, İdiyopatik Trombositopenik Purpura.*

INTRODUCTION

The giant-cell tumor (GCT) of bone is a primary bone neoplasm. It was first described macroscopically by Cooper in 1818 and later by Lebert using microscopy in 1845. An important progress was made by Jaffe who in 1940 defined this tumor as a separate clinical and radiological entity[1]. GCT is a monostotic condition probably originating from histio-fibroblastic elements in the metaphyseoepiphyseal areas of long bones[2]. It is constituted by round, ovoid or spindle-shaped basic cells that tend to fuse into multinucleate giant cells. It occurs relatively frequently and the transformation to sarcoma is around 5%. There is a slight predilection for the female sex. It occurs rarely before puberty and after 50 years of age. The most common localization is around the knee joint; more than 25%of all cases occurring in the proximal tibia[3,4]. Though curettage with bone grafting is the mainstay of treatment. A variety of adjuvant methods have been developed to reduce local recurrence rates which can be as high as 40%-60%. These include liquid nitrogen cryotherapy, phenol and alcohol chemical cautery and heat cauterization with saline or Polymethylmethacrylate (PMMA). Follow-up should include a quarterly history and physical examination as well as roentgenograms of the chest and involved extremities during the first two years[1].

* Assist. Prof., Department of Orthopaedics and Traumatology School of Medicine Süleyman Demirel University Isparta Turkey.
** Orth., Surgeon. Department of Orthopaedics and Traumatology Cerrahpaşa School of Medicine İstanbul University İstanbul Turkey.
*** Assoc. Prof., Department of Orthopaedics and Traumatology Cerrahpaşa School of Medicine İstanbul University İstanbul Turkey.
**** Prof., Department of Orthopaedics and Traumatology Cerrahpaşa School of Medicine İstanbul University İstanbul Turkey.

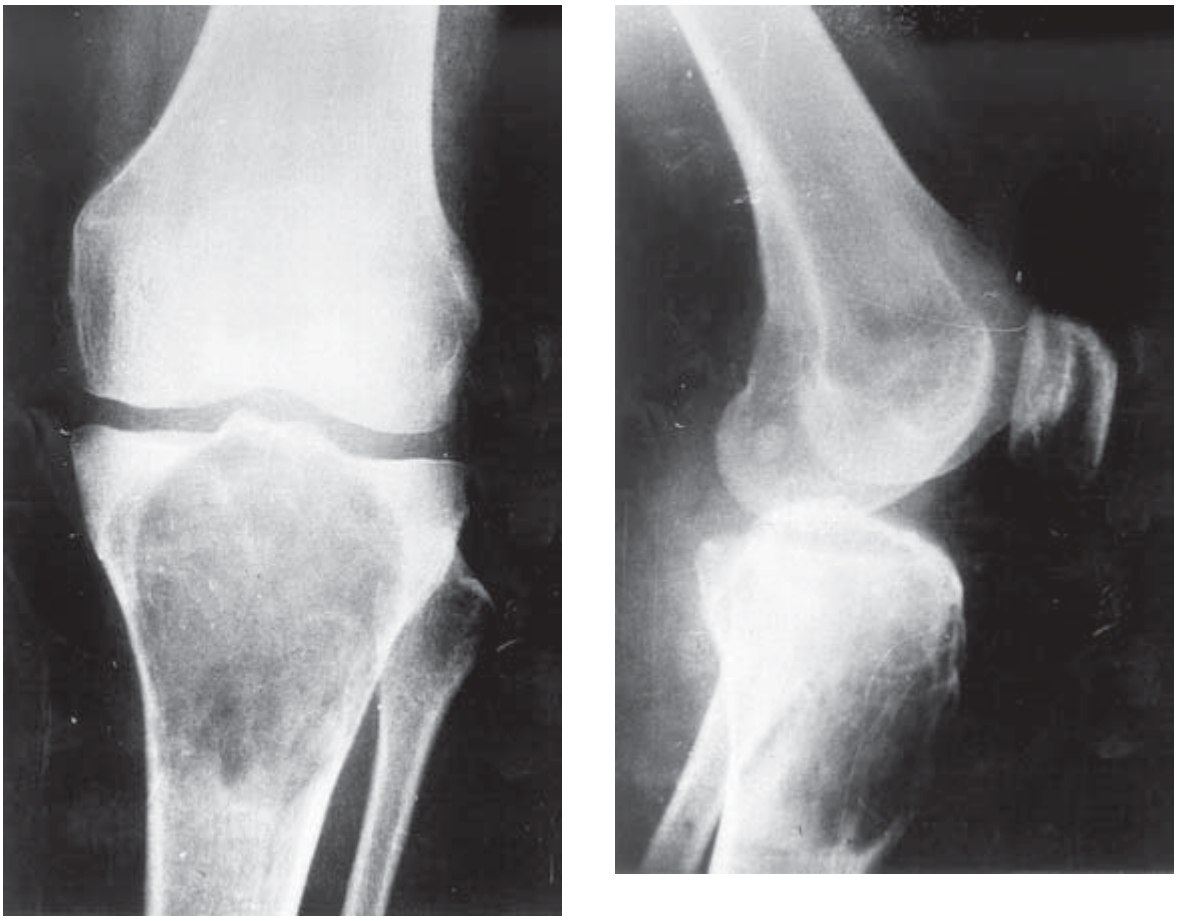
Osteochondromas develop from an aberrant subperiosteal germ of foetal cartilage. Para-articular or juxta-articular chondromas arise from the capsule or the extracapsular connective tissue of a large joint[5]. The concept of para-articular osteochondroma was first introduced in 1958 by Jaffe who used the terms para-articular chondroma and intracapsular chondroma to describe osteochondral metaplasia occurring in the joint capsule or soft tissues adjacent to a joint. Malignant transformation has not been reported though the histologic features may be suggestive of a malignant tumor including hypercellularity of the cartilaginous component and atypia of individual chondrocytes. Because marginal excision is adequate for these tumors it is important to distinguish them from synovial chondromatosis and chondrosarcoma[6].

Idiopathic (Immunologic) Thrombocytopenic Purpura (ITP) is in most adults a chronic disorder with no apparent predisposing course. In childhood ITP, a viral antigen is thought to trigger the synthesis of antibodies that may react with virus coated platelets in contrast to adults where the antibodies are directed against a structural platelet antigen (glycoprotein IIb-IIIa or glycoprotein complex Ib-IX). Acute ITP is generally seen in the paediatric population constituting only about 10% of adult cases where antibodies produced against a viral antigen cross-react with platelets accelerating their clearance. Chronic ITP mostly affects women in their 20-40s. The female to male ratio is 3:1[7].

CASE REPORT

A 31-year old female patient was first seen by us in January 1992 because of left knee pain of three months duration. The pain became worse with long distance walking. No significant personal or family history was elicited. A slight expansion of the proximal tibia was found on physical examination. There were no abnormal findings in the blood tests. An osteolytic lesion in the proximal metaphyseal region of the left tibia was seen on the plain radiographs (Fig. 1). Subsequently, the lesion was biopsied which revealed a GCT of bone (Fig. 2). A formal curettage and washout of the cavity with hot saline (60 °C) was performed as a definitive treatment. The resulting cavity was then filled with autogenous iliac bone graft and cement (Fig. 3). The postoperative course passed uneventfully and the patient was discharged with restricted weight-bearing on the involved extremity. She came to monthly follow-up visits and was allowed full weight-bearing at three months postoperatively when graft incorporation was observed.

Two and-a-half years had passed since the operation when the patient presented with pain in her right hip radiating to the knee. The physical examination revealed severe restriction of the Range of Motion (ROM) of the right hip. A proliferative lesion showing non-homogenous calcifications in the subcapital region of the right femur was seen on anteroposteriror and lateral views (Fig. 4). The patient was hospitalized again for surgery. Incidentally, it was found that the preoperative blood count showed marked thrombocytopenia (48,000/mm^3), so a haematological survey was started. The Prothrombin Time (PT) was normal and the activated Partial Thromboplastin Time (aPTT) was slightly delayed being 38 seconds (Normal: 26-34 sec.). Anti-nuclear antibodies were tested negative. The peripheral blood-smear revealed scarcely clustered thrombocytes and the bone-marrow aspirate showed slight erythroid hyperplasia, and abundant megakaryocytes with normal morphology. The patient was diagnosed as ITP and was placed on oral corticosteroid (16 mg. methylprednisolone, tid) therapy. She showed marked improvement with thrombocyte counts rising to 105,000/mm^3 at the end of the first week and to 147,000/mm^3 at the end of the second week of treatment. She is accepted as in remission and was taken to follow-up by haematologists. A thru-cut needle biopsy was done but could not be interpreted due to insufficient biopsy material. Then, a proximal femoral resection and prosthetic reconstruction was done. Macroscopic and microscopic evaluation of the resected mass revealed a para-articular osteochondroma (Figs. 5 and 6). The defect was managed with modular endoprosthetic reconstruction (prosthesis designed by Dr. Kotz) (Fig. 7). After the operation, there was postoperative bleeding from the incision, which could not be controlled by compressive bandages. The haematological parameters were checked again and the case was consulted with the haematologists. The blood tests were normal so a surgical cause of bleeding was contemplated. A second-look exploration was performed with evacuation of the haematoma and search for possible bleeding vessels, but none was found. There were recurrent bleeding episodes with intervals in-between. A catheter embolisation under fluoroscopic guidance was done at the end of the first postoperative month which stopped the bleeding instantly. A subcutaneous infection developed secondary to haematoma contamination. The bacteriological study revealed pseudomonas mirabilis and alpha-haemolytic streptococci as the infecting microorganisms. The infection resolved within one



(a) (b)

Fig. 1 a and b: Anteroposterior and lateral radiographs of the left knee.

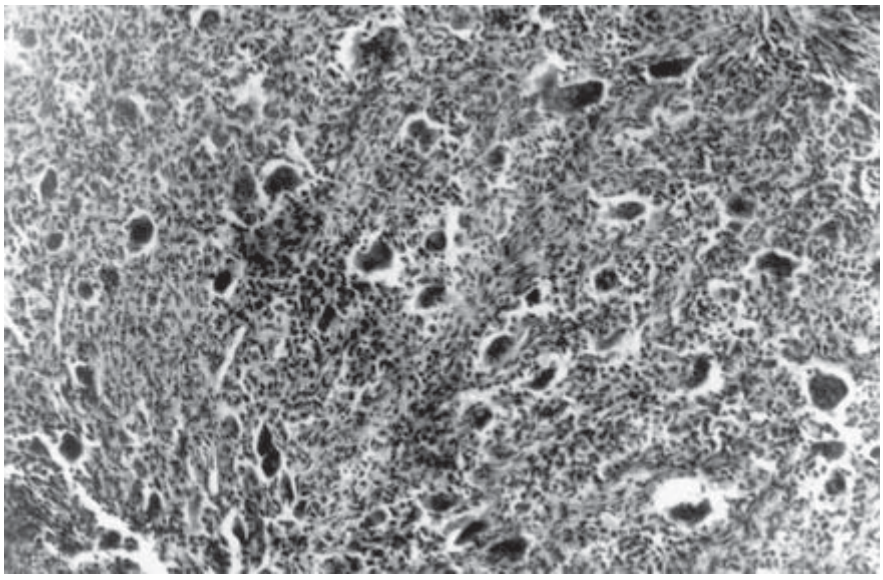


Fig. 2: The histologic specimen revealing multinucleated giant cells typical of GCT. H&E stain (Magnification: x 80).

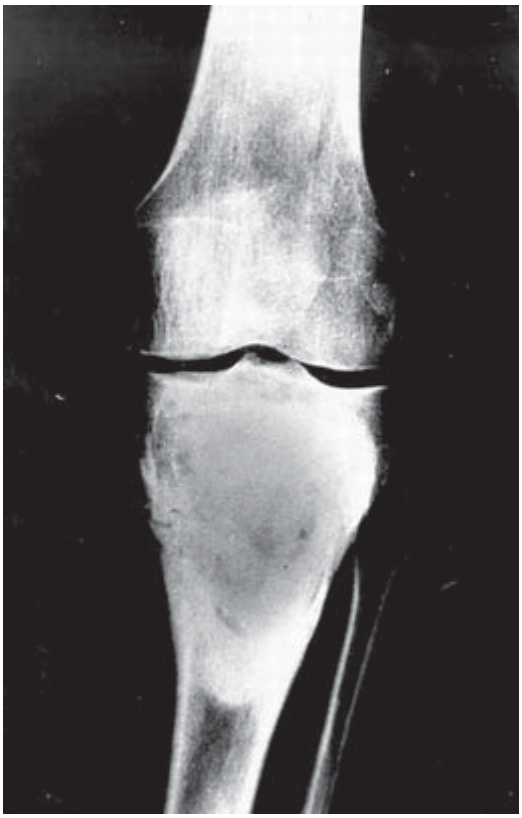


Fig. 3: Postoperative views after curettage and filling with autogenous graft and cement.

Fig. 4 a: Anteroposterior view of pelvis and lateral view of the right hip showing the para-articular mass.

Fig. 4 b: Anteroposterior view of pelvis and lateral view of the right hip showing the para-articular mass.

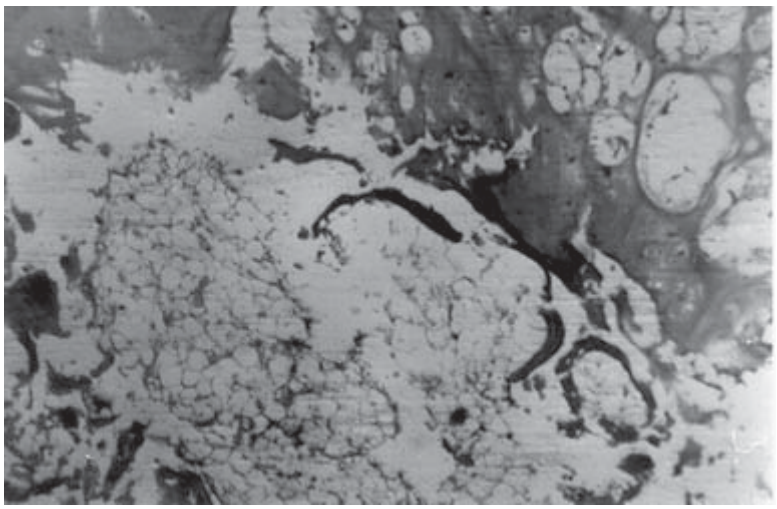


Fig. 5: Microscopic view of the resected tumor. H&E stain (Magnification: x 32).

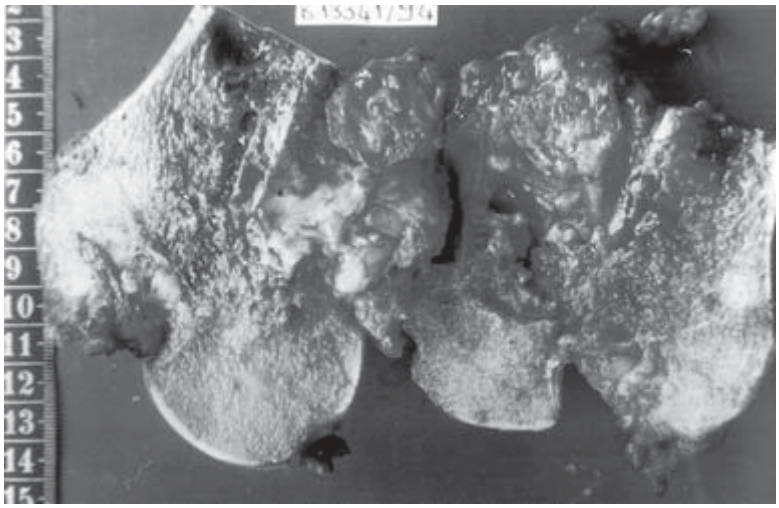


Fig. 6: The resected specimen macroscopically.

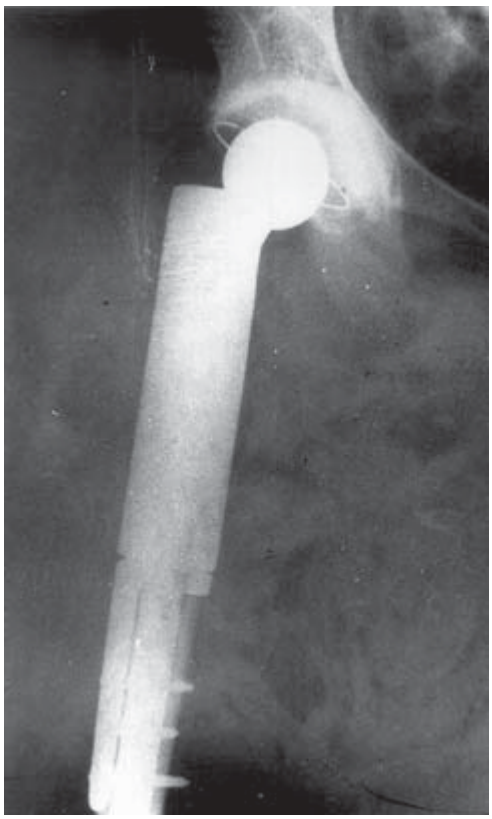


Fig. 7: Postoperative radiograph showing the reconstruction with a Kotz-type modular resection prosthesis in-situ.

month under appropriate antibiotic therapy. The patient was discharged from the hospital at 4.5 months postoperatively. She ambulates instantly and is still under follow-up with no local recurrence. Also, Relapse of ITP was not detected.

DISCUSSION

A variety of osteocartilaginous outgrowths are found near joints. Osteophytes occurring in degenerative arthritis are the most frequent. Proper osteochondromas, although near to the joint, are attached by a stalk to the cortical surface of the "parent" bone and so are metaphyseal non-articular lesions. However, they may occasionally protrude into the joint cavity. Differentiating between conventional osteochondromas, synovial chondromatosis, para-articular osteochondromas and even parosteal chondrosarcomas and osteosarcomas can be difficult. A conventional osteochondroma arises from a developmental defect in the growth plate and grows away from the nearest joint. Synovial chondromatosis, in contrast, is characterised by numerous cartilaginous nodules arising from a synovial membrane such as in a joint or a bursa[8]. Although their cause remains unknown, some authors have proposed that there is cartilaginous metaplasia of the connective tissue with subsequent ossification. The pathogenesis and classification of para-articular osteochondromas are controversial. However histological features can make the distinction from chondrosarcoma difficult on morphological basis alone[6].

An osteochondroma that continues to grow after skeletal maturity should be strongly considered to have undergone malignant degeneration. Features suggesting malignancy such as pain and growth after skeletal maturity should be recognized early. The risk of malignant degeneration is said to be approximately 1% per osteochondroma[9]. Imaging studies such as Magnetic Resonance Imaging (MRI) or Computerised Tomography are useful for evaluating adult osteochondroma. A cartilage cap greater than 0.5 cm may suggest a malignant transformation[10]. Radionuclide bone scanning may also be performed and it may show increased uptake at the site of the lesion[11]. If malignant degeneration is suggested by imaging studies, biopsy is usually not advised to be done before surgery as there is a significant risk of a non-representative biopsy and seeding of the biopsy track[9]. In our patient, sophisticated imaging techniques could not be obtained because of social and paramedical problems. The treatment algorithm of such a lesion should include a bone scanning and MRI of the lesion, in order to obtain adequate data. Although local resection of the lesion preserving the joint is the preferred method of treatment of such a lesion, proximal femoral excision and total joint replacement had to be performed because of the accompanying coxafemoral arthrosis and limited ROM.

Para-articular osteochondromas are extremely rare lesions. When literature is searched, only case reports of this lesion could be detected[12-14]. The most favoured sites of occurrence are the knee and the foot[6]. In our review of the relevant literature we could not find any case where a giant-cell tumor and a para-articular osteochondroma occurred in the same patient. Furthermore, our patient had a haematologic disorder superimposed that made the surgical management quite difficult. The patient was more than satisfied with the procedure that had been performed for the GCT of the left proximal tibia and there had been no remarkable postoperative occurrence. Unlike the first one, the second hospitalization was quite lengthy (4.5 months) because of intercurrent complications. Nevertheless, all the problems had been resolved and the patient was walking with crutches at the time of the discharge. At the latest follow-up; she was able to perform her daily activities without any problem.

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