Case report
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Pigmented villonodular synovitis of the knee in a child - case report

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Pigmented villonodular synovitis (PVS) is a destructive fibrohistiocytic proliferation with production of villus and nodular protrusions in synovial membrane. A boy, aged 12 years, had a knee injury during a football game with severe pane, swelling, decreased range of motion, and presence of hemarthrosis. Magnetic resonance imaging revealed, in the projection of the suprapattelar recessus, tumefaction of irregular shape; the largest diameter 11x5 cm. Upper pole of tumefaction was partly impressed in m. vastus intermedius. In general endotracheal anesthesia partial synovectomy was done with complete extirpation of tumefaction. The diagnosis of pigmented villonodular synovitis was established histologically. This type of tumefaction is rare in children, but the adequate treatment is necessary because of its aggressive growth with possible bone destruction and propensity of recidivism. The suspicion of malignant neoplasm is often present. Due to various clinical presentation of villonodular synovitis, early detection and adequate treatment is necessary.

KEY WORDS: *Synovitis, Pigmented Villonodular; Knee; Child*

INTRODUCTION

Pigmented villonodular synovitis (PVS) is destructive fibrohistiocytic proliferation with protrusions of villus and nodular protrusions in synovial membrane. Jaffe et al. (1) presented a unique clinicopathological concept of pigmented villonodular synovitis and tenosynovitis that enclosed different groups of fibrohistiocytic proliferations that can originate from different conditions and diseases. The process can involve articulations, bursas, tendon sheaths and surrounding fascias and ligaments, or their different combinations. Depending on the location of synovial membrane PVS is divided in intra-articular and extra-articular form. In intra-articular form lesion can be diffuse involving whole synovial membrane of articulation, or localized when only a part of synovia is involved (nodular or polypoid form) (1). In extra-articular form PVS can also be either diffuse with multifocal lesions of synovia, or nodular type with localized nodular changes that are frequently called gigantocellular tumor of tendon sheaths. It is widely accepted that the process is of reactive inflammatory nature. However, because of its aggressive growth with bone destruction or recidivism, low malignant local aggressive neoplasm is frequently suspected (3,4). If bones are not affected, the differential diagnosis includes trauma of meniscus, juvenile rheumatoid arthritis, synovitis hemosiderotic, hemophilia, synoviosarcoma, synovial hemangioma of fibromatosis (5). The aim of this work was to point the possible misdiagnosis between synovial sarcoma and PVS. In pediatric patients they have the same clinical presentation, similar behavior, and growth potential.

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CASE REPORT

A boy, aged 12 years, had a knee injury during football game-contusion in supracondylar region of right supracondylar femoral region with severe pain, swelling, decreased range of motion, and presence of hemarthrosis. At clinical examination, in motionless state, the tumefaction dimensions 2x4 cm, soft-elastic consisten-

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cy with wide base attached to the bottom, without of possibilities of wide movements was found. Native radiography of the knee region was normal for the patient of this age. Ten ml of fresh blood was evacuated by kneecentesis. Cryotherapy and application of elastic bandage with physical activity restriction was advised. Three months later, at the next control examination, swelling was completely regressed but tumefaction in suprapatellar recessus in lateral portion even after the physical therapy was still persisting. The kneecentesis is done again and 3 ml fresh blood was obtained.

Because of persisting tumefaction the ultrasound examination of the affected knee was done. The presence of cystic tumefaction dimensions 4x5x5 cm of irregular shape with calcification in the center was verified clearly separated from medially placed muscular tissue.

Magnetic resonance (MR) imaging revealed a tumefaction of irregular shape, the largest diameter 11x5 cm, in the projection of the suprapattelar recessus. Upper pole of the tumefaction was partly impressed in m. vastus intermedius. At the lower pole of tumefaction, a wall separating the tumor from articulation was detected. Formation was mostly cystic in the part of the lower pole, with soft-tissue infiltrative mass, which was predominantly located in the upper pole, as well as in the medial aspect. The structure of soft-tissue component was very heterogeneous. Inhomogeneity of T2 signal, partly with detected septa, and hypodensity of T1 signal with focuses of lost signals (can match hemorrhagic products) were presented. After contrast medium administration there was inhomogeneous and intense contrast enhancement. Contrast enhancement was evident also in the wall of described formation. In perilesional zones inside m. quadriceps femoris there was no sign of edema or detectable contrast enhancement but muscle infiltration could not be excluded. The bone was intact. Above the popliteal fossa, in the paravascular region a lymph node of 1 cm in diameter, probably reactively changed, was detected (Figure 1).

Based on these findings the conclusion was established: intraperiarticular tumefaction inside the suprapatellar recessus and m.quadriceps femoris, predominantly cystic with soft tissue proliferation. The lesion had aggressive characteristics. Differential diagnosis included synovial sarcoma, synovial hemangioma, inflammatory arthropathy, synovial chondromatosis, and pigmented villonodular synovitis. Seven months after the injury, in general endotracheal anesthesia partial synovectomy with complete extirpation of tumefaction and vacuum drainages was done (Figure 2).

At gross pathology, the formation was described as brown, irregular papillary surface with singular papillary protrusions. On the section, the tissue was of different color - reddish brown and yellow blending into each other. Under the microscopic examination existence of fibroblasts, histiocytes and inflammatory cell infiltration in groups, with gigantic, multinuclear cell type of foreign body among them was verified. Deposits of hemosiderin pigment, which gave macroscopically reddish brown color, were prominent. Numerous histiocytes had foamy cytoplasm and they were found solitary or in groups. Papillary surface of tumefaction was covered with multiplied histiocytes unclearly separated from inflammatory infiltration. Cellularity inside the lesion was irregular, with existing zones of hypocellularity and multiplied connective tissue. Based on pathologic analysis the diagnosis was established: synovitis villonodularis pigmentosa (Figure 3).

Histopathological differential diagnosis considered reactive inflammatory changes, posttraumatic reactive synovitis, rheumatoid arthritis, and synovial changes in hemophilia. Plasma cellular inflammatory infiltration and lymphoid follicles, which predominate in rheumatoid arthritis, do not exist in PVS. In posttraumatic

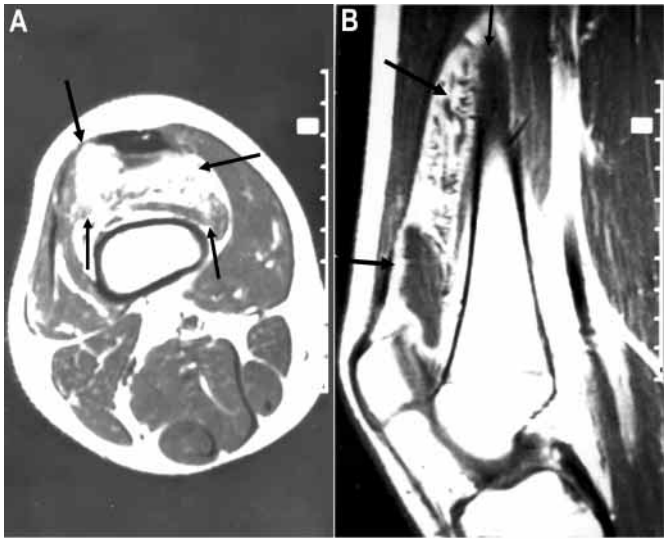

Figure 1. MR imaging of the tumefaction
In the projection of the supra-pattelar recessus, tumefaction of irregular shape, the largest diameter 11x5 cm, was detected (B). Upper pole of tumefaction was partly impressed in m. vastus intermedius. At the lower pole the wall separating the tumor from articulation is found (A, black arrow)

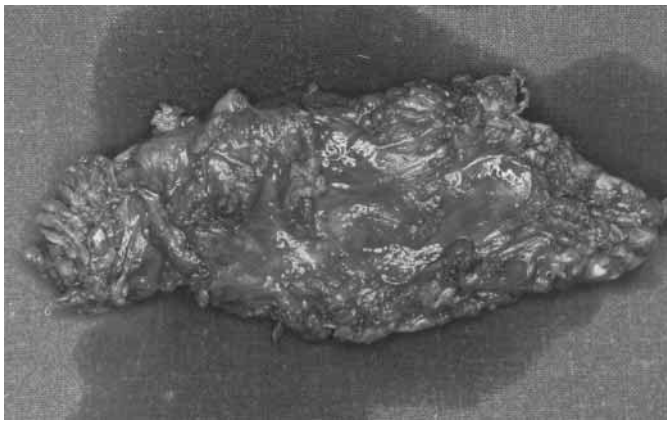

Figure 2. Exstirpated tumor immediately after operation

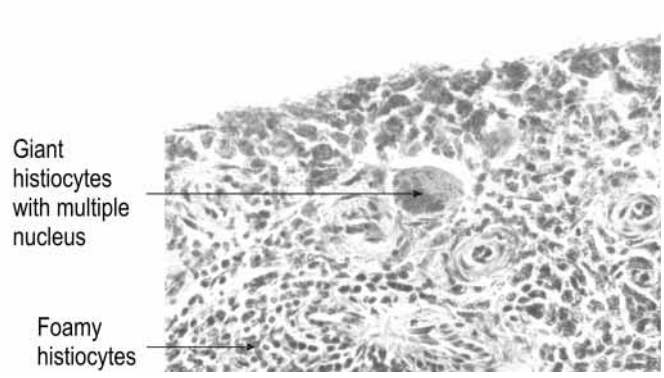

Figure 3. Histopathological analysis of PVS
Numerous histiocytes have foamy cytoplasm and they can be found solitary or in groups. Cellularity inside the lesion is irregular, with existing zones of hipocelularity with multiplicated connective tissue.

synovitis acute or chronic inflammatory infiltrate can be found but it is different from that found in PVS. The presence of foreign body in joint can be verified with the polarization light. In case of extra-articular form of PVS histological differentiation includes epithelioid sarcoma. Postoperative period was normal. In post hospitalization period, the applied physical therapy gave satisfactory results. The patient had no pain on day 10 of physical therapy. After four weeks of rehabilitation, a full range of motion of the affected knee was achieved. Control examination, 5 months after the operation, showed normal physical status with full range of motion and full physical activity, without pain, swelling or recidivism.

DISCUSSION

PVS diagnosis is frequently delayed due to nonspecific symptoms. Blood fluid aspiration and MRI are valuable tools for early diagnosis (4). For the successful treatment, it is very important to analyze the form and place of appearance of villonodular synovitis. It is considered that in 30% of cases recidivism appears after the surgical treatment. In patient with reoccurring recidivism one should think about the serious prosthetic procedure (5). In our case the patient underwent partial synovectomy without signs of recurrence 5 months after the surgery. Numerous authors report excellent or very good results after the local excision (6-8). Synovectomy can also be done with arthroscopy procedure or arthrotomy (9). Techniques used in different cases must have a complete synovectomy as the main principle of treatment (10). Sometimes, recidivism can occur after total synovectomy in diffuse forms of PVS and for that reason surgical treatment can be improved with radiotherapy. In children, radiotherapy as an additional therapy is not recommended because of potential damage of epiphyseal growth plate and postradiation sarcomas.
This type of tumefaction is rare in children, but the adequate treatment is necessary because of its aggressive growth with possible bone destruction and propensity of recidivism and malignant alteration. Secondary degenerative lesions that can affect joint may result in the prolongation of illness. Recognition of different clinical presentations of villonodular synovitis is a necessary precondition for early detection and adequate treatment of PVC.

Aknowledgement
Figure 1. was made at the Institute of Oncology Sremska Kamenica. Figure 2. was made at the University Clinic for Pediatric Surgery Novi Sad. Figure 3. was made at the Institute for pathology and histology Clinical Centre Novi Sad.

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