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CASE REPORT

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Case report: rheumatoid arthritis occurring during observation after surgery for localized pigmented villonodular synovitis

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Abstract In the rare case presented here, arthroscopic resection was performed after a diagnosis of localized pigmented villonodular synovitis (LPVS) of the knee, but the disorder recurred after about 1 year as the diffuse form, and synovectomy was performed through a medial parapatella incision. Subsequently, rheumatoid arthritis (RA) occurred during the postoperative observation period. The course of this case is interesting because of the combination of the two diseases. In cases of PVS, the possibility of RA should always be considered.

Key words Pigmented villonodular synovitis (PVS) · Rare case · Rheumatoid arthritis (RA)

Introduction

Pigmerted villonodular synovitis (PVS) is a disorder that shows localized tumorous proliferation of the synovial membrane, often occurring in a knee. It is a rare disease, with the annual incidence reported to be 1.8 in every 1 000 000 of the population.[1] Inflammation,[2–5] tumor,[6,7] abnormal lipid metabolism,[8] and trauma[9–13] have been suggested as etiological factors of the disease, but details are still unknown. In this report, a rare case of a patient who developed rheumatoid arthritis (RA) during observation after surgery for localized PVS (LPVS) is presented.

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Case report

The patient was a 38-year-old woman. Her primary complaint was pain in the right knee. She had no particular past history and no familial history. Pain in the right knee occurred with no particular cause about December 1996, but was left untreated. In March 1997, the patient began to note pain in the right knee in flexion, and consulted another hospital because of difficulties in climbing stairs and kneeling down. Thirty cubic centimetres of yellow fluid was aspirated by puncture of the knee. Swelling recurred a few days after the puncture, and as the symptoms had not been alleviated after a total of five punctures, the patient consulted our hospital on April 23. Swelling, balloning of the patella, and tenderness on the lateral side of the femorotibial joint were noted at the initial examination, and the range of motion was 50° flexion and 0° extension, with a restriction in flexion. No abnormal findings were noted on X-ray films. Puncture of the knee yielded 10 cc transparent yellowish fluid. In blood tests, only mild increases were noted in C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and CH50 (Table 1). MRI revealed the retention of synovial fluid, and a mass with a relatively low signal intensity in the suprapatellar bursa in T1-weighted (Fig. 1a) and T2-weighted (Fig. 1b) images. Arthroscopy performed on June 17 showed a tongue-shaped tumorous node, with a pedicle of about 1 cm, in the lateral joint capsule in the suprapatellar bursa (Fig. 2). The pathological diagnosis by biopsy was PVS. The node was resected arthroscopically at the base of the pedicle on August 8. Histological examinations showed capillary proliferation, marked infiltration of inflammatory cells, which were mainly small lymphocytes, and occasional multinucleated giant cells (Fig. 3a,b). Villous proliferation of the synovial membrane and infiltration of small round cells, which were mainly lymphocytes, were noted (Fig. 3c). On August 20, 1998, about 1 year after arthroscopic surgery, MRI again showed the retention of articular fluid. After 30 cc of bloody synovial fluid was aspirated by puncture on September 1, bloody synovial fluid was aspirated repeatedly. MRI performed on April 8, 1999,

Table 1. Laboratory findings at the initial examination

Urinalysis		Blood chemistry		Serology		Plasma proteins	
Protein	(−)	TP	7.4 g/dl	ESR	57 mm/h	Alb	62%
Sugar	(−)	Alb	4.5 g/dl	CRP	0.8 mg/dl	α1-Glb	2.4%
Ketone	(−)	Glu	102 mg/dl	IgG	1490 mg/dl	α2-Glb	8.6%
Occult blood	(1+)	BUN	9.6 mg/dl	IgM	238 mg/dl	β-Glb	9.6%
		Cr	0.5 mg/dl	IgA	191 mg/dl	γ-Glb	18%
		Na	139 mEq/l	IgD	107 mg/dl		
		Cl	104 mEq/l	CH50	59.1 IU/ml		
Hematology		K	3.7 mEq/l	RF	<10.9 IU/ml		
WBC	3500/μl	T-Bil	0.6 mg/dl	RAPA	Negative		
Neutro	60%	AST	31 U/l	RF-IgG	Negative	Blood coagulation	
Lym	35%	ALT	34 U/l	LE	Negative	PT	87%
Mono	2.0%	LDH	333 U/l	ANA	Negative	APTT	84%
Eosino	2.5%	ALP	252 U/l	Anti-RNP	Negative	Fibrinogen	307 mg/dl
Baso	0.3%	γ-GTP	57 U/l	Anti-Sm	Negative		
RBC	431×10^4/μl	ChE	342 U/l	Anti-SS-A	Negative		
Hb	12.1 g/dl	T-CHO	201 mg/dl	Anti-SS-B	Negative		
Ht	36.5%			Anti-Scl-70	Negative		
Plt	30×10^4/μl			Anti-Jo-1	Negative		

BUN, blood urea nitrogen; AST, aspartate aminotransferase; ALT, alanine aminotransferase; LDH, lactic dehydrogenase; ALP, alkaline leukocyte phosphatase; γ-GTP, γ-guanosire triphosphate; ChE, cholinesterase; ESR, erythrocyte sedimentation rate; CRP, C-reactive protein; RF, rheumatoid factor; LE, lupus erythematosus; ANA, antinuclear antibody; RNP, RNA polymerase; PT, prothrombin time; APTT, activated partial thromboplastin time

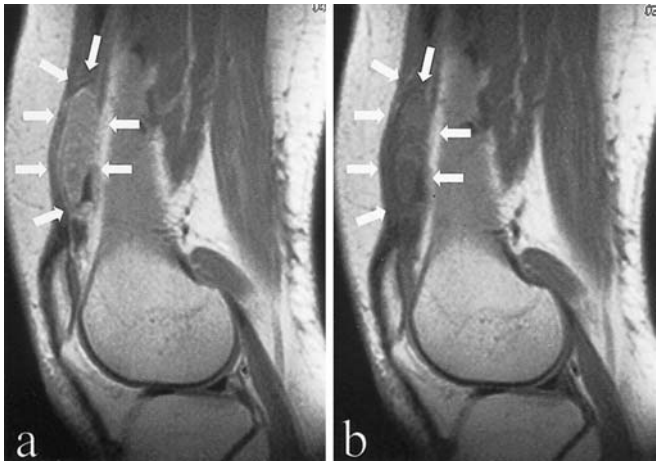

Fig. 1. MRI of the right knee showing a mass with a relatively low signal intensity in the suprapatellar bursa in **a** T1-weighted and **b** T2-weighted images

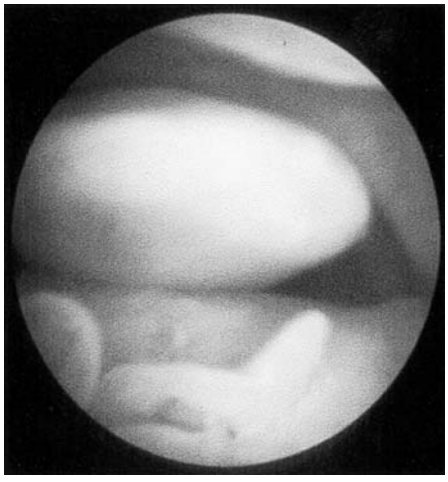

Fig. 2. Arthroscopic view showing a tongue-shaped tumorous node in the suprapatellar bursa

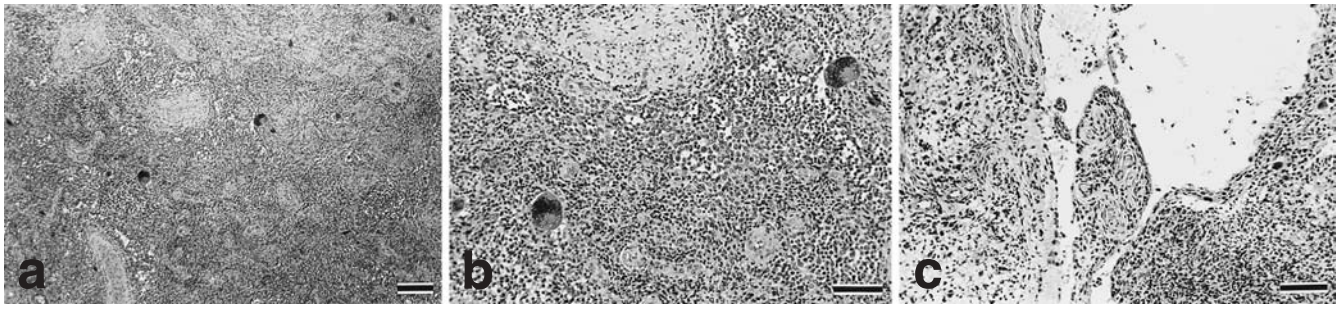

Fig. 3. **a** Histological findings of the node showing capillary proliferation, marked infiltration of inflammatory cells, which were mainly lymphocytes, and occasional multinucleated giant cells (hematoxylin–eosin; *bar* 200 μm). **b** A high-power field of (**a**) (hematoxylin–eosin; *bar* 100 μm). **c** Another region in the specimen showing villous proliferation of the synovial membrane, and infiltration of small round cells (hematoxylin–eosin; *bar* 100 μm)

suggested a recurrence of PVS in a wide area of the anterior and posterior parts of the knee (Fig. 4). Therefore, synovectomy was carried out through a medial parapatella incision on August 31. On gross observation, the synovial membrane was reddish brown and proliferated as a tumor over the entire knee joint. Histological findings were nearly the same as those at the initial surgery, i.e., capillary proliferation, marked infiltration of inflammatory cells, which were predominantly small lymphocytes, and a large number of multinucleated giant cells (Fig. 5a). Hemosiderin deposits were also noted occasionally (Fig. 5b). Radiation therapy was performed after surgery. The dosage applied was a cumulative focal dose of 48 Gy in 40 fractions.

Although the patient's subsequent course was uneventful, mild swellig was noted in the right knee at an examination on May 16, 2000, about 8 months after synovectomy, and mild swelling was also noted in the left knee on June 8. On September 12, morning stiffness lasting for 1 h, and swelling and pain of the bilateral wrists and the second and third MP joints of the left hand were noted. ESR was 82 mm/60 min, CRP was 2.7 mg/dl, and rheumatoid factor (RF) was 15 IU/ml. Radiographic findings of the bilateral wrists showed osteoporosis and joint-space narrowing of the carpal bones (Fig. 6). A diagnosis of RA was made, and drug therapy was initiated. On April 14, 2002, swelling and pain were observed in the bilateral wrists and the left ankle. Radiography of the bilateral hands showed destructive changes of the carpal bones, with a carpal height ratio (CHR) of 0.38 (Fig. 7).

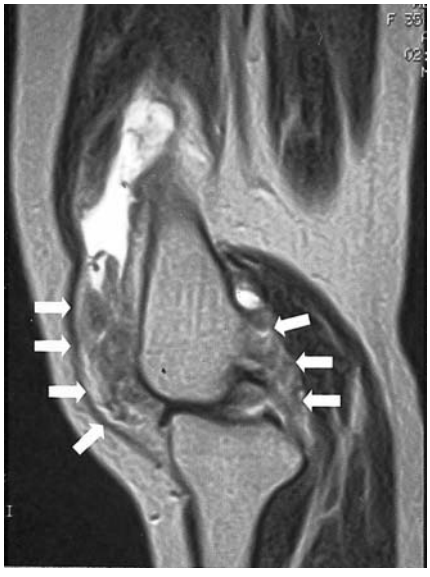

Fig. 4. MRI of the right knee 1 year and 8 months after arthroscopic resection, showing areas of low intensity mixed with an area of intermediate intensity, which is the multinodular mass of the anterior and posterior portion of the knee in a sagital T1-weighted image

Discussion

In this patient, localized PVS was diagnosed at the initial episode based on clinical findings, arthroscopy, and MRI. Arthroscopic resection was carried out. However, PVS recurred in the diffuse form after about 1 year. Histologically, no marked differences were observed between the initial and recurrent episodes. This course supports Hirohata's speculation that the localized form is an early phase of the diffuse form of PVS.[14]

This patient subsequently developed RA about 2 years and 6 months after the onset of PVS. According to our

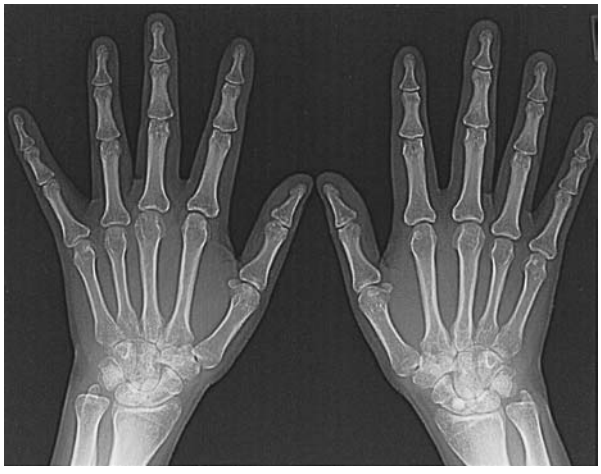

Fig. 6. Anteroposterior radiograph of the bilateral hands, showing osteoporosis and joint-space narrowing of the carpal bones on September 12, 2000

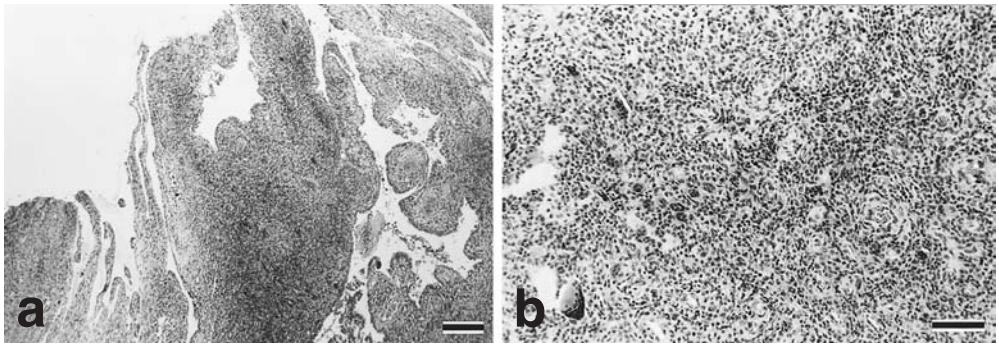

Fig. 5. **a** Histological findings of the recurrence of PVS showing capillary proliferation, marked infiltration of inflammatory cells, and a large number of multinucleated giant cells (hematoxylin–eosin; *bar* 200 μm). **b** In a high-power field, hemosiderin was noted (hematoxylin–eosin; *bar* 100 μm)

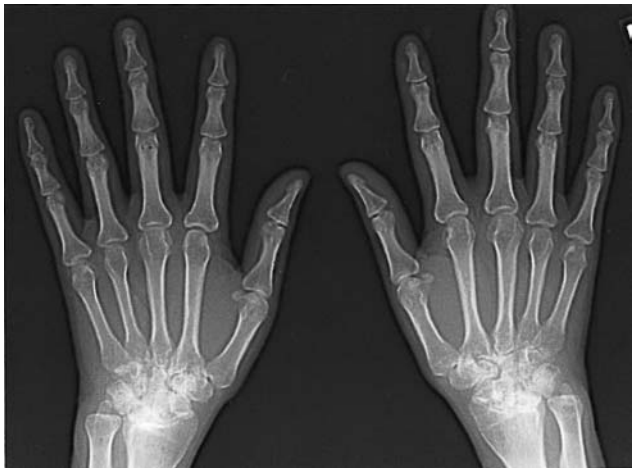

Fig. 7. Anteroposterior radiograph of the bilateral hands showing destructive changes in the carpal bones, March 14, 2002

review of the literature, there is no other report of the occurrence of the two diseases in this sequence. Basically, there are two components of the histological appearance of PVS: (1) villous projections of the synovial membrane due to a proliferation of surface synoviocytes and chronic inflammatory thickening of the synovial membrane, and (2) many pigment-laden and scattered lipid-laden histiocytes, occasional multinucleate giant cells, sometimes also bearing hemosiderin pigments, and a nonspecific chronic inflammatory infiltrate.[15] In this patient, the histological findings did not differ markedly between the initial and recurrent episodes, and no particular sign of RA was noted. According to the old diagnostic criteria published by the ARA in 1958,[16] RA is characterized by three or more of the following histological changes in synovial membrane: marked villous hypertrophy; proliferation of superficial synovial cells, often with palisading; marked infiltration of chronic inflammatory cells (with lymphocytes or plasma celle predominating) with a tendency to form "lymphoid nodules"; deposition of compact fibrin, either on the surface or interstitially; foci of cell necrosis. However, there is no specific pathological finding that decisively indicates a diagnosis of RA. Torisu and Watanabe[17] encountered a case in which PVS occurred in RA, indicating the possibility of the concurrence of the two conditions, although the order of their occurrence was the opposite of that in our case. There is one other similar report.[18] This patient could not be classified as having RA with the criteria of the American College of Rheumatology (ACR) at the initial examination. However, increased CRP and ESR may have suggested early RA. The case presented here, in which monoarthritis, diagnosed to be PVS, was followed by polyarthritis (RA), is interesting because of the combination of the two diseases. In all cases of PVS, the passibility of rheumatoid arthritis should be considered.

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