

## CASE REPORT

# SWEET'S SYNDROME ASSOCIATED WITH PIGMENTED VILLONODULAR SYNOVITIS

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Sweet's syndrome (acute febrile neutrophilic dermatosis) was first described in 1964. The typical symptoms of Sweet's syndrome are high temperature, peripheral leucocytosis, painful cutaneous rashes (papules, plaques) and arthralgia. Sweet's syndrome has particularly been described in association with neoplastic, infectious and immunological diseases. The pathogenesis of Sweet's syndrome can be explained by a reaction to an antigenic structure with accumulation of immunological complexes and liberation of inflammatory mediators. For the first time we report on a patient with Sweet's syndrome and pigmented villonodular synovitis, which is believed to play the antigenic role in the Sweet syndrome.

**Keywords** : pigmented villonodular synovitis ; Sweet's syndrome ; acute febrile neutrophilic dermatosis.

**Mots-clés** : synovite villo-nodulaire pigmentée ; syndrome de Sweet ; dermatose aiguë fébrile avec neutrophilie.

## INTRODUCTION

Sweet's syndrome (acute febrile neutrophilic dermatosis) was first described by Sweet in 1964 (10). The syndrome is clinically characterized by pyrexia, peripheral neutrophilia, painful erythematous papules and plaques and arthralgia.

Acute febrile neutrophilic dermatosis has most often been reported in association with neoplastic and infectious diseases, predominantly myeloproliferative diseases on the one hand, and infections of the respiratory, gastrointestinal and urogenital system on the other hand. We report on a patient with Sweet's syndrome who also had pigmented

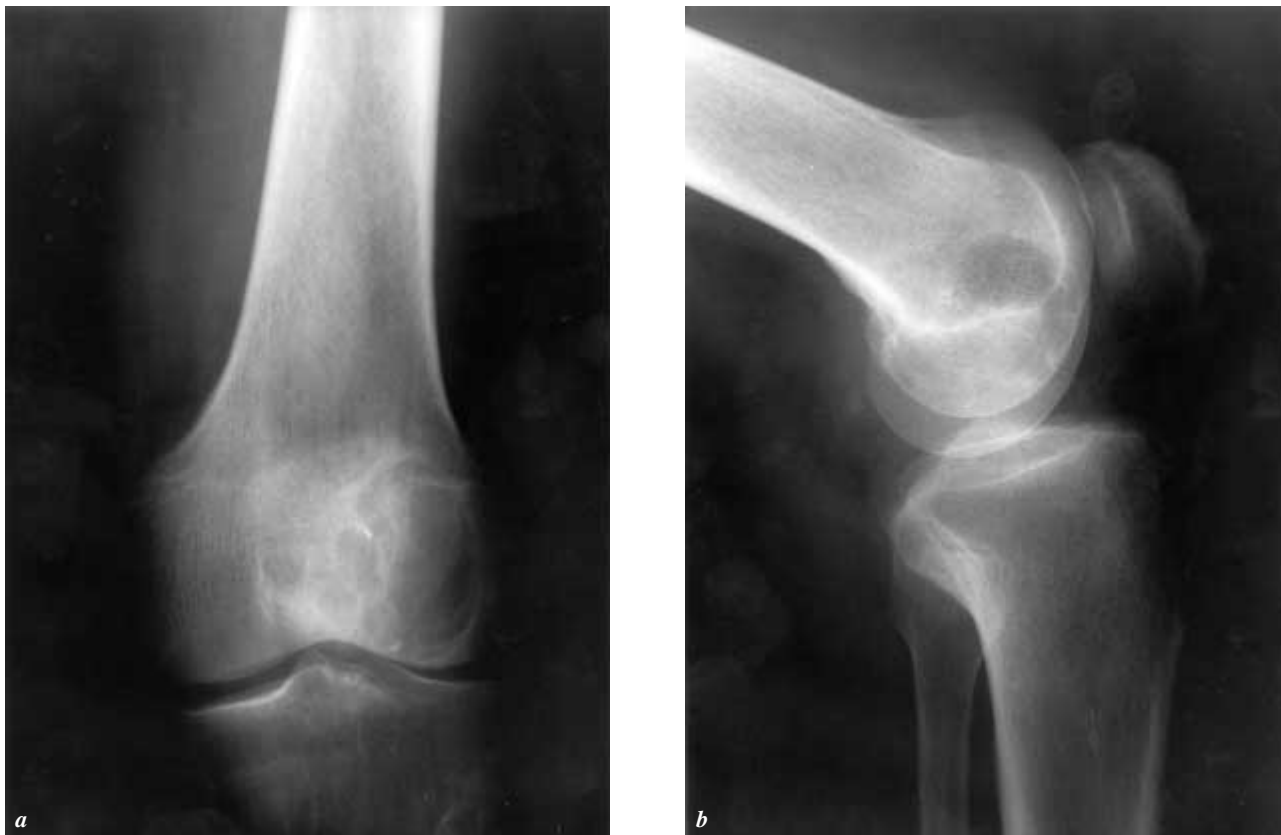
villonodular synovitis. To our knowledge this is the first case described as such.

## CASE REPORT

A 53-year-old man, who had resettled from Russia to Germany one year previously, had a temperature of 39° C and presented with painful disciform erythematous plaques and papules particularly on the back and on the extremities. In addition to this the patient complained of general weakness and athralgia in the large joints of the extremities. During a treatment in the dermatology clinic, the diagnosis of Sweet's syndrome was made. The patient complained of pain and swelling of the right knee, which were present for seven years and were treated conservatively. Laboratory examination showed neutrophilia, elevated erythrocyte sedimentation rate and positive Bence-Jones proteinuria, but a normal serum electrophoresis. Bone marrow biopsy and an intensive xray search did not indicate multiple myeloma. Therapy with steroids led to normalization of temperature, improvement of his general condition and reduction of arthralgias. The swelling and limitation of joint mobility of the right knee were resistant to therapy ; a repeat

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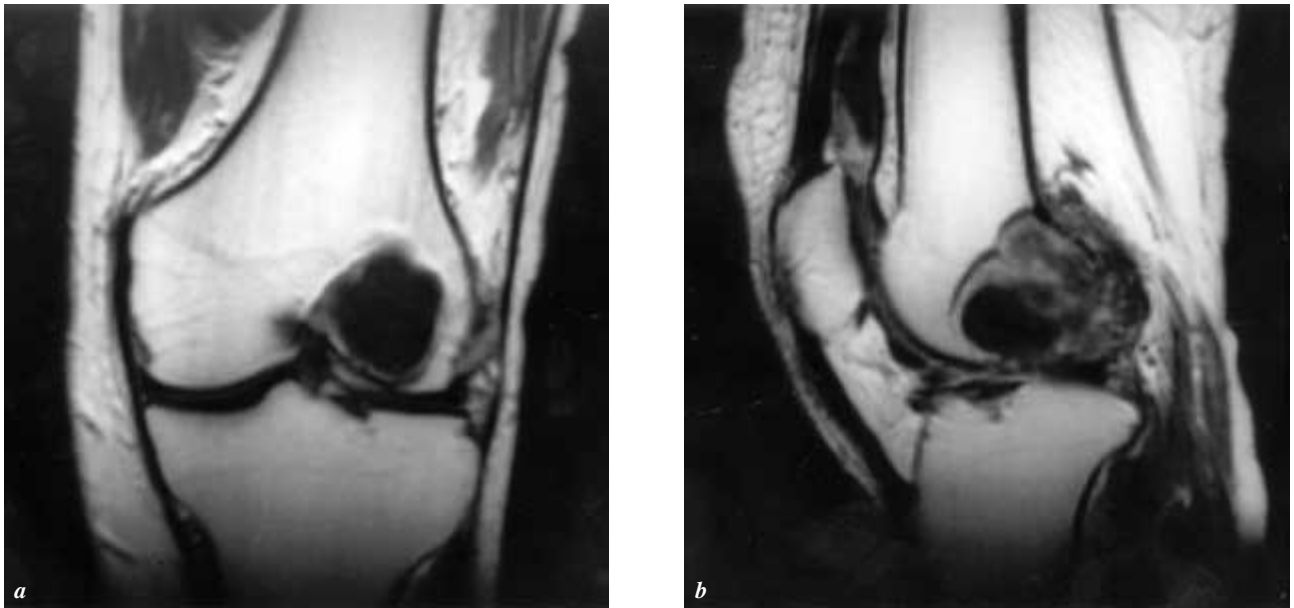
*Fig. 1.* — Xray of the right knee *a.* A.P. and *b.* lateral showing the osteolytic changes in the lateral femoral condyle

xray was therefore performed. In the xray (fig. 1) an osteolytic area was noted in the the lateral femoral condyle. Magnetic resonance imaging (fig. 2) showed a cystic lesion in contact with the joint.

Preoperatively the following differential diagnoses were discussed: osteoclastoma, aneurysmatic bone cyst and pigmented villonodular synovitis. At operation fresh frozen sections gave the diagnosis of pigmented villonodular synovitis, therefore the lesion and the synovia were resected. The definitive histological result confirmed the diagnosis of pigmented villonodular synovitis (PVS). After the operation the patient underwent further treatment in the dermatology clinic. The subsequent xray of the knee showed no abnormalities.

## DISCUSSION

Sweet's syndrome has been described in association with different diseases. A marked heterogeneity of diseases which are associated with Sweet's syndrome can be found in literature: Neoplastic diseases have been encountered, especially myeloproliferative diseases and solid malignant tumors (2). In addition Sweet's syndrome was described in association with infections of the respiratory, gastrointestinal and urogenital system. In some cases Sweet's syndrome was observed in patients with immunological diseases (Sjögren syndrome, *lupus erythematosus disseminatus*, Crohn's disease and ulcerative colitis) (1). Muster *et al.* (6) even found a coincidence of cardiovascular diseases and Sweet's syndrome. Some authors observed mani-



**Fig. 2.** — Magnetic resonance imaging of the right knee (TR 450 ms, TE 15 ms) showing the cystic lesion in contact with the joint corresponding to fig. 1. a) frontal plane b) sagittal plane.

festations of Sweet's syndrome during therapy with granulocyte colony-stimulating factor (3).

As far as we know Sweet's syndrome has rarely been described in association with diseases of the locomotor system : Majeed *et al.* (5) found Sweet's syndrome in a patient with recurrent multifocal osteomyelitis. Smolle and Kresbach (9) found Sweet's syndrome in a patient with rheumatoid arthritis. Furthermore one case was described in association with an osteosarcoma (8).

With respect to its pathogenesis, Sweet's syndrome is seen by various authors as an immunological reaction to an antigen (infection, neoplasia, autoantibody) with reactive formation of immunological complexes and accumulation of these complexes in tissues. Furthermore as a reaction to an antigen, activation of inflammatory cells can be observed, especially neutrophilic granulocytes. In addition to this a liberation of inflammatory mediators can be seen. The classical symptoms are pyrexia, arthralgia, peripheral leukocytosis and skin rashes ; they were all present in our patient.

PVS is a tumor-like change of the synovia of the tendon sheaths, bursae and of the synovium of the joints. The etiology of pigmented villonodular synovitis is also unknown : inflammatory, hemorrhagic and neoplastic processes have been considered as pathogenic factors in the literature (4, 7).

Now the question is posed whether in view of the heterogeneity of the associated diseases, a connection between Sweet's syndrome and pigmented villonodular synovitis also exists. Because of the pathogenic mechanism of Sweet's syndrome, the role of pigmented villonodular synovitis as a possible antigen is surely worth consideration. Until now this relationship has not been discussed.

*In summary* it can be stated that because of the heterogeneity of the diseases which are associated with Sweet's syndrome and the underlying pathogenic mechanism, we cannot exclude that pigmented villonodular synovitis was responsible for the appearance of Sweet's syndrome in our patient.

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## SAMENVATTING

*G. GOSHEGER, A. HILLMANN, T. OZAKI, H. BUERGER, W. WINKELMANN. Syndroom van Sweet geassocieerd met gepigmenteerde villonodulaire synovitis.*

Het syndroom van Sweet, eerst beschreven in 1964, bestaat in een koortstoestand gepaard gaande met pijnlijke huidrupties onder vorm van papels en vlekken, arthralgiën en leucocytose. Het wordt vooral gezien bij neoplastische, infectieuze en immunologische aandoeningen. Vermoedelijk is het te wijten aan een reactie op een antigeen, waarbij immunologische complexen zich opstapelen en inflammatoire stoffen vrijkomen. De schrijvers observeerden een geval, waarbij het syndroom van Sweet samenging met een gepigmenteerde villonodulaire synovitis. Vermoedt wordt dat PVS de antigene rol heeft gespeeld in dit geval.

## RÉSUMÉ

*G. GOSHEGER, A. HILLMANN, T. OZAKI, H. BUERGER, W. WINKELMANN. Syndrome de Sweet associé à une synovite villo-nodulaire pigmentée.*

Le syndrome de Sweet (dermatose aiguë, fébrile, avec neutrophilie) a été décrit en 1964. Ses symptômes associent typiquement une hyperthermie, une leucocytose périphérique, des éruptions cutanées douloureuses (papules, plaques) et des arthralgies. On a décrit en particulier l'association d'un syndrome de Sweet avec des affections néoplasiques, infectieuses et immunologiques. La pathogénie du syndrome peut s'expliquer par une réaction à une structure antigénique, avec accumulation de complexes immunologiques et libération de médiateurs de l'inflammation. Les auteurs rapportent un cas princeps de syndrome de Sweet associé avec une synovite villonodulaire pigmentée, qui a pu jouer le rôle antigénique dans la genèse du syndrome de Sweet.