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PIGMENTED VILLONODULAR SYNOVITIS

By

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Pain and swelling in the knee are commonplace symptoms. It is not always so simple, however, to diagnose the causal relation of these symptoms. If faced by a patient with such symptoms, which have not disappeared within a reasonable time—and when no other cause has been disclosed—pigmented villonodular synovitis should be contemplated.

Pigmented villonodular synovitis is no new disease. It was not classified more closely, however, until in 1941 *Jaffe, Lichtenstein & Sutro* considered—on the basis of experimental research and their own clinical material, as well as the perusal of previously published papers —that they were able to establish that it was a lesion of inflammatory nature.

Due to the polymorphous and greatly variable histological picture the disease has been given different names in previous publications. Those in most common use have been: synovial giant cell tumour, xanthoma, xanthogranuloma, pigmented giant cell xanthosarcoma, benign synovioma, polymorphous cellular tumour, hemorrhagic villous synovitis, sclerosing hemagioma, giant cell fibrohemangioma, and pigmented villonodular synovitis.

As indicated by these different designations, the joint demonstrates besides profuse brownish-yellow to clear exudate, villous processes and nodules which may become rather large. They have a fungous appearance, of characteristic yellowish-brown colour with more strongly pigmented spots. The histological picture is characteristic. Processes are found, covered by several layers of epithelial cells, amply vascularized stroma, lymphocytes, plasma cells, lipoid macrophages with cholesterol deposits, areas with "foam cells", extracellular and intracellular hemosiderin, multinucleated giant cells. At a more advanced stage of the disease the vascularization deteriorates to hyalinization, fibrosis, sclerosis. The picture varies somewhat with the phase of the disease, and

furthermore, some cell forms may predominate. The latter circumstance is reflected in the varying terms used for this disease.

Even if the above-mentioned authors intended to record the lesion as an inflammation, they were unable to demonstrate any releasing factor. The exudate has always proved to be sterile, no infection being revealed, therefore. Due to the deposits of cholesterol in the cells the possibility has been entertained, that the lesion might be caused by an inflammatory reaction to a local disturbance of the cholesterol metabolism. Injections of cholesterol, cholesterol-ferrum chloride, or cholesterol esters caused no disease, however. Neither has it been demonstrable that persistent hemorrhage, due to minor traumas, has been a causative agent. The lesion does not occur in hemophilia patients nor have injections of autoblood in rabbits' joints produced pigmented villonodular synovitis. The hemosiderosis may be secondary—a sequel to the crushing of villous processes by ordinary articular function.

Efskind (1947) has shown by his studies on the anatomy of the knee joint capsule, that subject to some reservations, the subsynovial tissue may be classified in the reticulo-endothelial system, and on this basis pigmented villonodular synovitis may possibly be recorded as reticulo-endotheliosis.

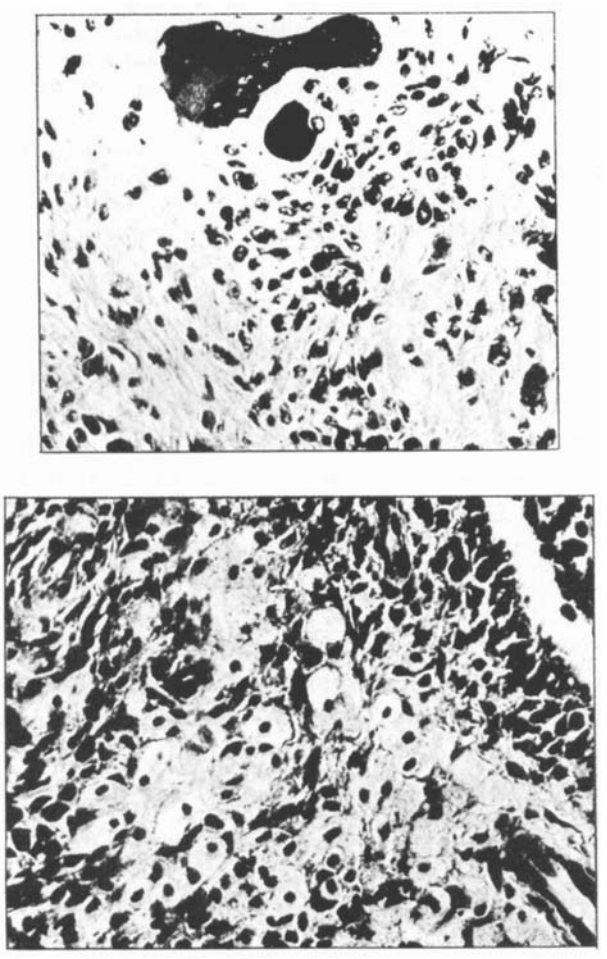
The disease is primarily localized to the knee joint, though it has been found also in other joints, tendon sheaths and bursae. At the latter places it is associated with minor complaints, and the hyalinized-fibrous form has been observed most frequently (Harbitz 1927).

Apart from one case (*Wallace-Greenfield*, 1950) with simultaneous localization to two joints, the lesion is monarticular. It is found in a diffuse form and in a localized form with one nodule up to three-four villous nodules.

The patients, who are in the age group 18–60 years, complain of the ordinary symptoms: pain and swelling in the knee joint. Initially these are intermittent, frequently lasting for several years. Less common symptoms are locking and lameness. There is no restriction of movement, beyond the moderate, usually no increase in temperature. Objectively exudate is demonstrated, and nodules may be palpated at times. Puncture of the joint produces a yellowish-brown or clear exudate. This is rapidly reproduced. X-ray examination as a rule is negative. A film of the soft parts may show nodular shadows, however. If the villonodular character of the lesion is recognized, the differential diagnosis of synovialoma (synovial sarcoma) may be considered. The latter is extracapsular, however, contains amorphous calcium, and invades the bone (*Lewis*, 1947).

The treatment is combined surgical/radiological. As much as poss-

ible of the affected tissue is removed with the greatest care, and afterwards the operative field is treated with roentgen doses between 1500–3000 r (*Shafer & Larmon*, 1951).



Microscopical picture is polymorphous and greatly variable. See description in text.

On March 23rd 1953 a 53 year old female was admitted to the Surgical Department A. of Rikshospitalet, complaining of pain and swelling in the right knee for fifteen years. The condition would improve somewhat when she kept quiet, but was never quite satisfactory, and during the past 2–3 years the complaints had increased. Physical examination showed normal findings. The right knee joint region was somewhat swollen, particularly downwards medially to the patella, where a distinct roll was palpable. This was somewhat tender. A moderate exudate was demonstrable in the knee joint. Severe pain was elicited by attempt at flexion beyond 90 degrees. There was no abnormal lateral movement, no demonstrable

atrophy of the muscles. X-ray examination with arthrography showed nothing special. Cholesterol content of blood was 170 mg. per 100 ml.

On March 27th 1953 arthrotomy was performed. A reddish-brown exudate was evacuated. Large villous nodules were demonstrated, and edematous, brownish, fungous masses, most of which were removed. Histological examination showed the characteristic picture of pigmented villonodular synovitis (see fig.). The patient received roentgen irradiation, therefore, with a total dosage of 2700 r. The postoperative course was complicated by thrombosis-emboli; the symptoms disappeared rapidly, however, after treatment with heparin and antiprothrombin.

The patient has subsequently been re-examined several times, the last time on January 9th 1954. She is subjectively free from symptoms and is very satisfied with the result. No demonstrable swelling or exudate in the knee joint. She is able to flex the knee to 135 degrees.

SUMMARY

A survey of pigmented villonodular synovitis in the knee joint has been given in connection with a case of complete recovery after combined treatment with surgery and deep roentgen irradiation.

The histology indicates a specific reaction in the reticulo-endothelial tissue of the joint capsule.

RESUME

Compte rendu de l'observation d'une synovite villonodulaire pigmentée dans le genou en connexion avec un cas de guérison complète après traitement opératoire combiné à une profonde irradiation aux rayons X.

L'histologie indique une réaction spécifique dans le tissu réticulo-endothélial de la capsule articulaire.

ZUSAMMENFASSUNG

Eine Übersicht der pigmentierten villonodulären Synovitis im Kniegelenk wurde im Zusammenhang mit dem Bericht eines Falles von vollständiger Heilung nach chirurgischem Eingriff, kombiniert mit tiefer Röntgenbestrahlung, vorgelegt.

Das histologische Bild spricht für eine spezifische Reaktion im retikulo-endothelialen Gewebe der Gelenkskapsel.

LITERATURE

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