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A case of aggressive pigmented villonodular synovitis

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An unusual case of pigmented villonodular synovitis affecting a knee joint is described. Shortly after synovectomy, extensive bone and ligament destructions occurred and made conservative surgery impossible.

Pigmented villonodular synovitis usually progresses slowly, with a diagnostic delay of one to several years (Jaffe 1958, Myers 1980). We report a case of rapid destruction of the knee in spite of early synovectomy.

Case report

After operation for a breast carcinoma 4 years ago, a 78-year-old woman gradually developed swelling and pain with impaired motion of her left knee. She had no previous injury of the knee. Arthrosis was suspected and fenestration of the left tibia was performed 3 years ago with no effect on the symptoms.

On admission the left knee was fixed in 30° of flexion with passive flexion to 80°. A 15×15 cm soft tumor was palpated proximal to the patella. The knee was stable. A few milliliters of turbid fluid was aspirated. Radiographs showed large lytic destructions of the distal femur and proximal tibia. On reexamination of the radiographs taken 3 years earlier, minor destruction in the distal femur could be seen (Figure 1). Because metastasis from the earlier breast carcinoma was suspected, an open biopsy was planned. At arthrotomy the synovium was found to be thickened, and it filled the entire joint. Invasion of both the femoral and the tibial epiphyses was seen. Total synovectomy with curettage of the osseous lesions was performed. Histologic examination showed a solid tumor consisting of uniform cells without marked polymorphism. The cytoplasm was clear, with vacuoles, and in certain areas abundant hemosiderin. Only a few mitoses were seen. The number of multinucleated giant cells was moderate. No sign of metastasis from the breast carcinoma was found (Figure 2). Immunohistochemical analyses showed a negative reaction to desmine, keratine, and vemintine, but a positive reaction to alpha-1-antichymotrypsine, alpha-1-antitrypsine, and a slight positive reaction to muramidase, indicating histocytic genesis. The histologic diagnosis was aggressive proliferating pigmented villonodular synovitis.

The postoperative course was prolonged due to severe pain, but gradually the patient was mobilized with crutches and discharged from the hospital after 1 month with knee motion of 20–100° without instability.

Three months later, the patient was readmitted because of increasing pain, swelling, and instability of the left knee with subluxation and severe bone destructions (Figure 1). Further examination revealed absence of the patellar and cruciate ligaments. The patient refused amputation. She was confined to a wheelchair and discharged to her local hospital with the left knee immobilized in an orthosis. Three months later, she died of a pulmonary embolus. Autopsy was not performed.

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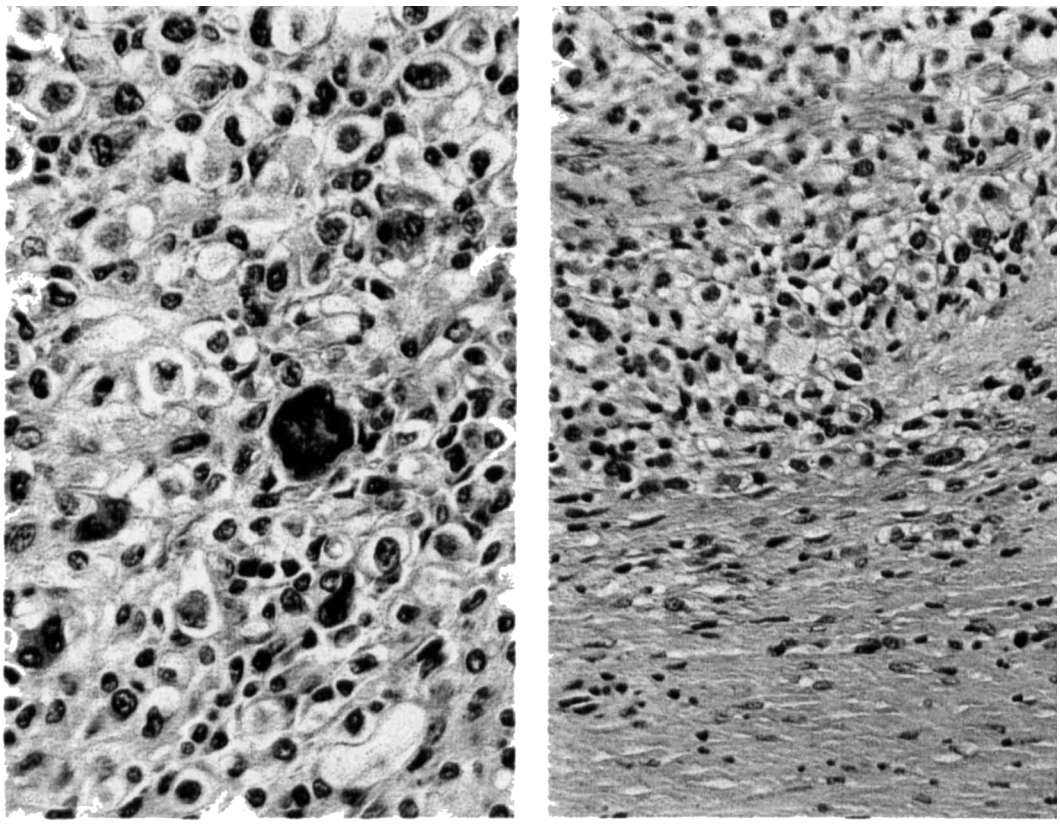
Figure 1. A 78-year-old woman with pain and swelling in the left knee for 4 years.
A. Three years ago. Destructions in the distal femur.
B. At admission. Note clusters of cysts in the distal femur and proximal tibia.
C. Three months after synovectomy. Note posterior dislocation of the tibia, destruction of the tibial tuberosity and the anterior of the tibial joint surface. The patellar articulation has been destroyed by an extensive cyst in the femoral condyle.



A B

Figure 2.
A. The center of the lesion with a few multinucleated giant cells and many histiocytes with a smaller rounded nucleus and clear cytoplasm. HE×300.
B. Periphery of the lesion with foamy histiocytes invading the connective tissue. HE×300.

Discussion

In a summary of reports on pigmented villonodular synovitis affecting the knee, 21 of 81 joints had either bone cysts or minor cortical erosions (Dorwart 1984). The radiographic appearance is typically a single cyst or clusters of cysts involving the juxtaarticular bone on both sides of the joint, but with the joint space preserved (Scott 1968). Extensive local destruction of bone and invasion of soft tissue, as in our case, are very rare (Atmore 1956, Jergesen 1978, Kindblom 1978). The rapid course of the disease after synovectomy and the extensive destruction of bone with involvement of soft tissues clearly distinguish our case from the commonly benign behavior of this condition.

The recurrence rate is high, ranging from 17 to 46% (Myers 1980, Rao 1984). We have chosen the designation "aggressive" to indicate that the lesion in our case represented a borderline malignant variant. A true malignant counterpart to these lesions in which both the tumor and the metastases contain identical morphologic features has only been reported once (Enzinger 1983).

Histologically, the main differential diagnosis is synovial sarcoma. Multinucleated giant cells, which are rare in synovial sarcoma, foam cells, hemosiderin-containing histiocytes, foci of inflammatory cells, and the absence of cellular anaplasia distinguish pigmented villonodular synovitis from synovial sarcoma (Nilsonne and Moberger 1969). In clear cell sarcoma the multinucleated giant cells are less predominant, with the nuclei located peripherally. The inflammatory

or xanthomatous forms of malignant fibrous histocytoma have a predominantly acute inflammatory background as compared with the modest number of chronic inflammatory cells in pigmented villonodular synovitis (Enzinger 1983). The histologic picture in our case showed fewer multinucleated giant cells; the xanthomatous cells had a more abundant cytoplasm; and the collagenous stroma between cells was more delicate when compared with the usual histology of the benign pigmented villonodular synovitis.

Total synovectomy combined with curettage of the bone cysts is considered by most authors the best treatment (Byers 1968, Scott 1968, Granowits 1976). The use of radiotherapy in the treatment of recurrent pigmented villonodular synovitis is controversial (Atmore 1956, Wiss 1982). If destruction of the joint cartilage and minor bone destructions are present, arthrodesis or arthroplasty is indicated. In our patient, none of these alternatives were considered feasible, and amputation was therefore suggested.

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