THE DIAGNOSIS AND TREATMENT OF PIGMENTED VILLONODULAR SYNOVITIS

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The clinical and pathological distinction between pigmented villonodular synovitis and malignant synovioma has in the past often posed difficult problems: there is no doubt that many amputations have been performed for what is now considered to be an entirely benign condition, and mistakes may still be made by those who are unfamiliar with the two conditions.

The purpose of the present report is to review the literature on pigmented villonodular synovitis and to present information with regard to a series of eighty cases studied personally, in which histological material and detailed clinical information were available.

HISTORICAL REVIEW

The first account of what we now know as pigmented villonodular synovitis was that of Chassaingac (1852) who described lesions of the nodular form arising in relation to the flexor tendon sheaths of the middle and index fingers. Simon (1865) described a large pedunculated nodule in the knee. Both considered the lesions to be sarcomata. Moser (1909) reported the first example of the diffuse type of lesion: the ankle was affected and the patient was free of recurrence seven years after synovectomy. Dowd (1912), who described a diffuse synovial lesion of the knee, was the first to question the malignant nature of this type of lesion.

In the early literature the nomenclature is often confusing, the terms xanthoma or giant-cell tumour (Targott 1897), villous arthritis (Dowd 1912), benign synovioma (Stewart 1948) and myeloxanthoma (Dor 1898) being used. These names suggested a neoplastic origin, although Dor had first suggested that the nodular lesion was inflammatory. More recently Wright (1951) has maintained that the lesions in question are benign synoviomas.

In 1941 Jaffe, Lichtenstein and Sutro introduced the term pigmented villonodular synovitis. Their paper, based on clinical and pathological experience with twenty cases involving joints, tendon sheaths and bursae, is still the definitive account of the condition. They observed that the nodular and diffuse lesions were histologically similar, and suggested that they were part of the same disease process. The benign course, as well as the histological appearance of the lesions, led them to conclude that the condition was not a tumour, but an inflammatory response to an unknown agent.

In 1954 Young and Hudacek produced changes which they regarded as similar to those of pigmented villonodular synovitis by repeated injection of blood into the knees of dogs. It has been suggested, however, that these changes are not really comparable with those of pigmented villonodular synovitis, but more closely resemble those seen in haemophilia (Hoagland 1967).

Fisk (1952) attributed the changes of pigmented villonodular synovitis to repeated minor trauma to synovial fringes, with consequent hydrarthrosis. This was thought to set up a
self-perpetuating process which led ultimately to pigmented villonodular synovitis. Geschickter and Copeland (1949) suggested that the lesions originate from osteoclasts in sesamoid bones, but the anatomical sites of the lesions (fingers, hips, knees, ankles) make this view untenable. A disorder of lipoid metabolism has been postulated by several authors, including de Santo and Wilson (1939) and Galloway, Broders and Ghormley (1940), who found raised blood cholesterol levels in some of their patients. More recent investigators do not appear to have confirmed this observation. The true nature of the disease process is therefore still in dispute. Most writers now favour an inflammatory process, but the etiological agent is still unknown.

CLINICAL FEATURES

Pigmented villonodular synovitis usually affects young adults (Atmore, Dahlin and Ghormley 1956), and published series of cases include approximately equal numbers of men and women. In the nodular form, lesions of the fingers are the most numerous and involve the anterior aspect more frequently than the dorsum (Sherry and Anderson 1955; Phalen, McCormack and Gazale 1959). Nodular lesions also occur in the larger joints, notably the knee, where a pedunculated nodule may give rise to instability and locking (Jaffe 1958, Granowitz and Mankin 1967). The diffuse form usually affects the knee, but lesions of the hip, ankle, subtalar joint, elbow, wrist and shoulder have all been recorded. The disease is characteristically monarticular: in the diffuse form only one well documented example of multiple joint involvement, where both knees were affected, has been described (Greenfield and Wallace 1950), but several cases of multiple nodular lesions are on record (Chassaingac 1852, Galloway et al. 1940).

In both the diffuse and the nodular form the disease process is usually slowly progressive, and two or three years often elapse before the patient seeks medical advice (Jaffe 1958, Wright 1951). Although pain is not usually severe, it can occasionally be a definite symptom (de Santo and Wilson 1939, Atmore et al. 1956). Swelling is nearly always present. Joint stiffness is common in cases of long duration. In the diffuse form there is diffuse synovial thickening, and a bloodstained effusion is often present. The joint is usually not tender, but its temperature may be slightly raised (Jaffe 1958, Larmon 1965).

Lewis (1947), Breimer and Freiberger (1958) and Smith and Pugh (1962) have drawn attention to radiological changes in adjacent bone. These are most often seen in the fingers and hip: typical lesions show cystic erosions on either side of the joint without calcification or sclerosis, and with no loss of joint space or demineralisation of surrounding bone, and there may also be lobular swelling of the soft tissues.

TREATMENT

All authorities agree that the nodular lesions should be treated by local excision, despite a high rate of recurrence, which ranges from 16 per cent (Galloway et al. 1940) to 48 per cent (Wright 1951). In the diffuse form extensive synovectomy is advocated by most writers (Shafer and Larmon 1951, Atmore et al. 1956). In the hip, Chung and Janes (1965) have used arthroplasty with encouraging results.

The high recurrence rate after synovectomy has led to the use of radiotherapy, either in combination with synovectomy (Shafer and Larmon 1951, Friedman and Schwartz 1957) or alone (Greenfield and Wallace 1950). There is no doubt that both synovial and intraosseous lesions will regress after radiotherapy, but McMaster (1960) and Atmore et al. (1956) feared that joint stiffness might be precipitated and were not convinced that this form of treatment is effective or justified. Furthermore, Chung and Janes (1965) showed that satisfactory long-term results can be obtained even when the disease has not been completely eradicated.
PRESENT SERIES

The records of the Royal National Orthopaedic Hospital and the Middlesex Hospital over a period of approximately thirty years were searched, and more than 200 examples of miscellaneous joint disease were found. Cases in which the following diagnosis had been made were reviewed: pigmented villonodular synovitis, benign synovioma, malignant synovioma, giant-cell tumour of tendon sheath, xanthoma, non-specific synovitis, traumatic synovitis and haemarthrosis.

The histological sections were examined by four of us who are pathologists, first independently and in the absence of clinical information, and then as a group with the clinical details. An agreed diagnosis of pigmented villonodular synovitis was reached in 126 cases. Each of us approached the diagnosis of pigmented villonodular synovitis independently, on the basis of the information in the literature and his own experience. Lack of agreement was expected but was, in fact, rarely encountered. When there was disagreement, opinion was between pigmented villonodular synovitis and non-specific synovitis in eleven cases, and between pigmented villonodular synovitis and a malignant tumour in eight cases. The eleven cases were regarded as uncertain and were not included in the final group of cases: of the eight cases in which malignancy was initially suspected, three were agreed on discussion to be malignant and five to be pigmented villonodular synovitis. Follow-up information substantiated this view.

<table>
<thead>
<tr>
<th>Site</th>
<th>Number of cases</th>
<th>Diffuse</th>
<th>Nodular</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knee</td>
<td></td>
<td>24</td>
<td>13</td>
<td>37</td>
</tr>
<tr>
<td>Fingers</td>
<td></td>
<td></td>
<td>26</td>
<td>26</td>
</tr>
<tr>
<td>Foot</td>
<td></td>
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<tr>
<td>Ankle</td>
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<tr>
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<td>2</td>
</tr>
<tr>
<td>Wrist</td>
<td></td>
<td>2</td>
<td></td>
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<tr>
<td>Shoulder</td>
<td></td>
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</tbody>
</table>

In eighty of the 126 histologically diagnosed cases detailed clinical and follow-up information was available, and the present report is based on this group. Seventy patients were examined personally by one of us, and in the remaining ten the necessary information was obtained by correspondence. The follow-up period varied from three to thirty-five years from the time of treatment.

DISTRIBUTION OF LESIONS

All cases in the present series showed either joint or tendon sheath involvement: no bursal lesions were encountered. The distribution of lesions is shown in Table I. Lesions of the knees and fingers occurred most commonly (46 per cent and 33 per cent of all cases), while feet, ankles, hips, wrists and shoulders were affected with decreasing frequency. In only one patient, with involvement of both ankles, was more than one joint affected. In the fingers, lesions were mostly on the palmar aspect of the distal or intermediate phalanx: they did not always have origin from the tendon sheath and sometimes appeared related to the interphalangeal joint.
AGE AND SEX

The condition is chiefly one of young persons or those in early middle life (Fig. 1). The most frequent age at presentation was in the fourth decade, with an even distribution above and below that age. The youngest patient was eleven and the oldest sixty-eight. Thirty-three patients were men and forty-seven women.

SYMPTOMS

Swelling, which was by far the most frequent presenting symptom, was seen in seventy patients. Pain was present in thirty-three, but was usually mild: its severity was not necessarily related to the size of the swelling. Joint stiffness (eleven patients) was usually associated with the larger swellings. Instability and locking occurred only in knee lesions, usually of the nodular form. In one patient symptoms resulted from pressure on the ulnar nerve at the wrist.

The interval between the onset of symptoms and clinical presentation ranged from one month to fifteen years, with an average of two and a half years. In forty-seven patients the interval exceeded one year. A history of trauma was obtained in eighteen patients.

PHYSICAL SIGNS

In the diffuse form the joint was swollen, partly from synovial thickening and partly from synovial effusion. Aspiration of bloodstained fluid from a chronically swollen joint is suggestive of pigmented villonodular synovitis and was a helpful diagnostic point. The involved joints were often slightly warm, but not tender. Limitation of movement was seen in the long-standing cases with marked swelling. Knee lesions occasionally presented as localised swellings in the popliteal fossa, initially regarded as semimembranosus bursitis.

FIG. 1
Age at onset of pigmented villonodular synovitis.

FIG. 2
Radiograph showing invasion of femur and pelvis in case of pigmented villonodular synovitis of hip.
In nodular lesions of the knee there was sometimes an effusion, and in two-thirds of the cases a mass was palpable. In many cases a diagnosis of loose body or meniscus injury had been made before operation.

In several knee lesions the appearances suggested a progression from the nodular to the diffuse form.

RADIOLOGICAL FINDINGS

In most cases there was no radiological abnormality apart from soft-tissue swelling. In 15 per cent, however, unmistakable evidence of bone involvement was shown: this, together with uncertain histological interpretation, sometimes led to unnecessarily radical treatment. Figure 2 shows the very invasive appearance in a case of subsequently proven pigmented villonodular synovitis, in a man of forty-seven with one year's history of pain and stiffness of the hip. Biopsy appeared to confirm the radiological diagnosis of malignant synovioma and hindquarter amputation was performed. At operation pigmented synovial tissue was found to invade the femur and the pelvis (Fig. 3). Subsequent re-examination of the histological sections established the correct diagnosis; the patient is alive and well nine years later.

Erosions of phalanges of the hand and foot are shown in Figures 4 and 5. In each case local excision was performed and the diagnosis confirmed histologically. Follow-up for periods of four and seven years has revealed no sign of recurrence.

Figure 4—Antero-posterior and lateral radiographs showing invasion of the proximal phalanx of a finger in the nodular type of lesion. Figure 5—Radiograph showing invasion of the proximal phalanx of toe in nodular type of lesion.
FIG. 6
Diffuse lesion of the knee joint showing darkly pigmented tissue.

FIG. 7
Figure 7—Another diffuse lesion of knee joint showing villous and nodular structures.

FIG. 8
Figure 8—A nodular lesion from the tendon sheath of a finger.

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HAEMATOLOGICAL INVESTIGATIONS
Estimation of haemoglobin, sedimentation rate, white cell count and the Rose Waaler test for rheumatoid arthritis were carried out in all patients. Normal results were almost always obtained.

PATHOLOGICAL FINDINGS
The common pathological features of the group of cases studied consisted of villous or nodular proliferation of synovial tissues, and pigmentation due to the presence of haemosiderin and lipid. The gross appearance is variable (Figs. 6 to 8). A given lesion may show a predominantly villous or nodular appearance, or both may be seen. Extensive areas of synovial membrane may be covered with fine or coarse villi which are usually a brownish red colour. The synovial tissues may be diffusely thickened and sessile or pedunculated nodules, up to several centimetres long, may be present. The degree of pigmentation ranges from a barely detectable yellow colour to dark brown. In the vast majority of cases the amount of pigment is enough to be recognised by the naked eye, but in a few cases it can be found only when histological sections are examined. The tissue can be soft and fleshy, solid, or even hard. In occasional cases gross examination shows extension of pigmented tissue into adjacent bones, and this may sometimes be extensive (Fig. 3). Despite the alarm that such a finding produces, it must be emphasised that it is not, with this type of lesion, an indication of malignancy.
On microscopic examination the abnormal tissue in pigmented villonodular synovitis consists of a stroma of reticulin and collagen fibres in which a variety of cells, including lymphocytes, plasma cells, histiocytes, fibroblasts and multinucleated giant cells, are found. The villous lesions contain relatively few reticulin or collagen fibres, and this is also true of the fleshy tissue of the softer nodules (Figs. 9 and 10). In such lesions the reticulin fibres are coarse and form an open network (Fig. 9): they usually surround small groups of two or three cells. Firmer lesions, usually nodular, have a more collagenous stroma, usually present in the form of strands of varying thickness which separate smaller or larger groups of cells (Fig. 11). As the stromal tissue increases in amount, the strands coalesce to form large sheets.
of collagen dotted with small spaces. Some lesions are highly collagenous: the fibrous tissue sometimes shows hyaline change and some areas may be mistaken for osteoid or cartilage (Fig. 12). The tissue may be highly vascular (Figs. 14 and 15), but the vessels have a normal structure and their appearance does not suggest that vasoformative activity is part of the basic disease process.

![Image of tissue sample]

**Fig. 11**
More prominent stromal tissue, with groups of cells, spaces and multinucleated giant cells. (Haematoxylin and eosin, $\times 205$.)

![Image of tissue sample]

**Fig. 12**
Highly collagenous material resembling osteoid tissue or cartilage. Scattered multinucleated giant cells are present. (Haematoxylin and eosin, $\times 130$.)

The great majority of cells are mononuclear and are round or oval in shape. Some have the features of plasma cells, while others resemble small lymphocytes. The majority are larger, and their exact nature is less easily specified. Some, containing haemosiderin pigment or lipid, appear to be histiocytes (Figs. 16 and 17). Others appear to be related to spaces in the tissue: it is tempting to regard them as synovial in nature, but this point needs to be
examined in more detail. A variety of tissue spaces are, in fact, present in pigmented villonodular synovitis. Some are simply artefacts; others are vascular, while others appear to be ramifications of the main synovial cavity (Figs. 14 and 15) produced by the coalescence of synovial villi. The spaces, however, which show the closest relationship to the cells of the tissue are of another type (Fig. 11). They appear to originate in the tissue: some are lined by mononuclear cells resembling those of the more solid areas; others contain the same mononuclear cells, or multinucleated giant cells, lying free in the central cavity. The lining cells rarely form a continuous layer, and do not produce mucin: their appearance thus contrasts strikingly with that of the "pseudo-epithelial" structures of a malignant synovioma. This type of space was found in a high proportion of the present series and is consequently to be regarded as a rather characteristic histological feature of pigmented villonodular synovitis. It is possible that the structures in question are formed by the proliferation of entrapped surface synovial cells, or that they represent a process of synovial differentiation. Alternatively, it is possible that they may be a consequence of the progressive condensation of the fibrous stromal component to which reference has already been made. The question of their synovial

![Image](https://image-url.com)

**FIG. 13**

Highly cellular tissue with multinucleated giant cells and strands of collagenous material. (Haematoxylin and eosin. × 205.)

nature must consequently be left undecided at present: in this context it must be emphasised that synovial cells, like other types of mesothelial cell, are notoriously difficult to define and identify.

Multinucleated giant cells are present in all lesions and are a conspicuous histological feature in about half of them (Figs. 11, 12, 13 and 16). In most the nuclei are distributed throughout the cell, although occasionally they are located peripherally. In the cellular lesions the giant cells often appear as clusters of nuclei with only an indistinct cytoplasmic border separating them from the adjacent stromal cells (Fig. 13). When the stroma is more collagenous the giant cells are more characteristically surrounded by spaces (Fig. 11). The size of the cells is variable, and the number of nuclei may be as many as fifty or more. Giant cells may occur in areas devoid of haemosiderin pigment or lipid.

Spindle-celled fibroblastic tissue was not conspicuous in lesions of the present series, although the formation of the collagenous stroma presumably results from the activity of this type of cell.
The diagnosis and treatment of pigmented villonodular synovitis

Although the degree of cellularity of some areas of tissue in pigmented villonodular synovitis may be disturbingly high, and occasional mitotic cells may be present, the experience gained during the examination of the present series of cases was that the nuclear pleomorphism, hyperchromatism and abnormal mitosis characteristic of a malignant tumour were never seen. The appearance of the cells was, in fact, always that of a benign condition.

![Fig. 14](image1)
Villous synovial lesion showing strands of vascular cellular tissue. The dark patches are areas of haemosiderin pigmentation. (Haematoxylin and eosin, ×55.)

![Fig. 15](image2)
A more solid villous lesion, showing spaces which appear to be ramifications of the joint space. An area of palely staining lipid-containing macrophages is present. (Haematoxylin and eosin, ×55.)

The substances responsible for the pigmentation of the tissue are haemosiderin (Figs. 14 and 16) and lipid (Figs. 15 and 17); the quantity of these varies greatly, although all the lesions studied contained at least microscopic amounts of each of them. They may be scattered quite diffusely throughout the tissue, but it is more usual for them to be concentrated focally. The lipid material is always within macrophages; the haemosiderin is
nearly always in mononuclear cells, although a few granules may be found extracellularly or in multinucleated cells. In one of the cases studied, large amounts of haemosiderin pigment were present in regional lymph nodes, the gross appearance wrongly suggesting metastasis of the pigmented synovial lesion.

**Fig. 16**
Collections of iron-containing pigment (haemosiderin) are present in many cells. (Perls' reaction for haemosiderin, ×220.)

**Fig. 17**
Collections of palely staining lipid-containing macrophages. (Haematoxylin and eosin, ×205.)

The histological structure of the lesions of pigmented villonodular synovitis which had extended to bone was not different from that of other cases, the same combination of round cells, multinucleated giant cells and fibrous stroma being found (Fig. 18).

**DIFFERENTIAL HISTOLOGICAL DIAGNOSIS**

Conditions to be considered include other types of synovitis and synovial reaction, and true tumours occurring in the neighbourhood of joints and tendon sheaths.
The synovitis group may be a cause of confusion, particularly in traumatic cases when iron-containing pigment may be present. But other features of pigmented villonodular synovitis such as conspicuous giant cells and lipid accumulations are rarely present, and the lesions do not have a villous or nodular appearance. This type of reaction is commonly seen in the knee joint after meniscus injuries (Soeur 1949); it has also been described in the shoulder in association with rotator cuff tears (Cotton and Rideout 1964); essentially similar findings have been reported in other types of haemarthrosis.

Rheumatoid synovitis is frequently villous in appearance, but the synovial tissues fail to show any appreciable degree of pigmentation. The characteristic change is a prominent accumulation of lymphocytes and plasma cells; the more varied cell types of pigmented villonodular synovitis are not seen.

The question of distinguishing between pigmented villonodular synovitis and malignant synovioma was the starting point for the present investigation. That confusion had occurred in past years was clear; four patients of the present series had, in fact, been treated by major amputation on the supposition that the lesions were malignant. On review of the material, however, it became clear that, with experience, effective histological distinction between the two groups could be made. The lesions of pigmented villonodular synovitis never showed the histological characteristics of a malignant tumour; they never metastasised, although they sometimes showed local extension to adjacent tissues, including bone. In contrast, lesions referred to as malignant synovioma have a predominantly spindle-celled structure which is unquestionably neoplastic; they are distinguished from other soft-tissue tumours by the presence of characteristic "pseudo-epithelial" structures which are quite different from the spaces of pigmented villonodular synovitis. They lack the pigmentation and the characteristic villous and nodular appearance of the latter type of lesion. It is of interest that they rarely, if ever, involve the internal aspect of a joint, or take origin from preformed synovial tissue.

The term benign synovioma has, in the past, been used to describe what is now referred to as the nodular type of pigmented villonodular synovitis (Stewart 1948, Wright 1951). Jaffe and his colleagues (Jaffe et al. 1941, Jaffe 1958) have argued at length against the neoplastic
nature of this type of lesion, and the findings in the present group of cases appear to us to be in keeping with the "inflammatory" or "reactive" nature of the condition. Furthermore, the possibility of confusion between benign and malignant synoviomas, and the absence of any intermediate forms, or of examples of malignant change in the benign condition, are additional reasons for adopting the term pigmented villonodular synovitis.

The fundamental requirement in making a diagnosis of pigmented villonodular synovitis is to be aware of the condition and to have experience of it; reaching the correct diagnosis then presents little problem.

**TREATMENT**

**Finger lesions**—Except in one case, lesions of the fingers were initially treated by local excision. Although digital nerves were sometimes damaged, patients were relieved of their symptoms. There was, however, a recurrence rate of 27 per cent. This is attributable to an attempt, in some cases, to excise the nodule in the out-patient department under local anaesthesia or without a tourniquet. In two patients, fingers were amputated for repeated recurrences; in one case primary amputation of a finger was carried out for a mistaken diagnosis of synovial sarcoma.

**TABLE II**

**RESULTS OF OPERATION FOR PIGMENTED VILLONODULAR SYNOVITIS OF THE KNEE**

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Complete relief of symptoms</th>
<th>Improved but some symptoms persist</th>
<th>No change</th>
<th>Worse since operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Synovectomy</td>
<td>2</td>
<td>6</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Excision of tumour</td>
<td>6</td>
<td>3</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Biopsy only</td>
<td>1</td>
<td>3</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Arthrodesis</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

**Knee lesions**—This group of lesions fell into two categories—diffuse and nodular—and these showed a marked difference with regard to the results of treatment. In the nodular form, excision gave satisfactory results (Table II). Six of the thirteen patients were completely relieved of their symptoms and only four were not improved, possibly because of further lesions undetected at operation. There were only two definite recurrences.

The diffuse lesions did not respond so well to surgery. In most an extensive synovectomy was performed and the results were poor. Only two patients were cured and in three the knees actually became stiffer and more painful. The rest had residual symptoms or reported no appreciable change after treatment. Two patients required a quadricepsplasty after synovectomy in order to regain the range of movement before operation. Biopsy was the only operation in five patients; in no case did deterioration follow this procedure although the residual deformity sometimes remained. In one of the patients biopsy was followed by a course of radiotherapy; later examination showed a satisfactory result.

The recurrence rate after synovectomy was 46 per cent and is probably attributable to the difficulty of performing a complete synovectomy in this joint. Arthrodesis was performed in three patients, in two as a primary procedure and in one for recurrence following synovectomy and radiotherapy. Two above-knee amputations were performed in cases where an erroneous histological diagnosis of malignant tumour (synovial sarcoma and fibrosarcoma) had been made. Both patients were free of recurrence at subsequent examination.

**Other joints**—Two below-knee amputations were performed for foot lesions in cases where the original biopsy specimens had been reported as showing malignant synovioma. In a third
case a histological diagnosis of malignant synovioma was made following excision biopsy. The patient, a girl of nineteen, refused amputation. At follow-up none of these patients showed any evidence of recurrence. The other nine lesions of the foot and ankle responded well to local excision; the recurrence rate was of the same order as in other parts of the body, but no patient was made worse by surgery.

One patient with a lesion of the hip has already been discussed. A second patient with a hip lesion is also of special interest. Pain was a marked feature and was again accompanied by bone involvement. Synovectomy and curettage of the bone lesions were performed and followed by radiotherapy. Subsequently he became impotent, and later developed a femoral neck fracture which united. Radiologically, the lesions have regressed, but the patient has been left with a stiff, painful hip.

Radiotherapy—Only eight patients were given a full course of irradiation, some as long as thirty years ago. The numbers are too small to allow any firm conclusions, although there is no doubt that radiotherapy causes both bone and soft-tissue lesions to regress. On the other hand, one suspects that the impotence and fracture in the patient with the hip lesion may have been caused by it. Two patients with knee lesions treated by radiotherapy after synovectomy developed increasing joint stiffness; one eventually required an arthrodesis. The results of radiotherapy were thus satisfactory in only two cases.

DISCUSSION

Pigmented villonodular synovitis is a relatively rare condition, only 126 cases being found in the records of the two hospitals during a period of approximately thirty years. Some of these had been mistaken for malignant tumours, and it is therefore important to stress the benign nature of this condition. No death from pigmented villonodular synovitis has been recorded, nor are there any authenticated reports of lymphatic or haematogenous spread.

It is possible in almost all cases to make the diagnosis on the basis of the macroscopic appearances at operation, and to confirm this by histological examination. Our findings are in all essentials similar to those of most other writers in that it is a monarticular, relatively painless, chronic condition affecting young adults. In 80 per cent of the series either the knee joint or the finger was affected.

The typical nodule on the finger usually presents little diagnostic difficulty, although radiological evidence of bone invasion may mislead the unwaried into diagnosing a malignant lesion. Recurrence may further alarm both the surgeon and the pathologist, but, provided the natural history of the condition is borne in mind, it should not be difficult to reach the correct diagnosis. Treatment of the finger lesions present's little difficulty, although it must be stressed that excision should always be performed with great care and under proper conditions, using general anaesthesia and a tourniquet. Although we have no proof we feel that the high recurrence rate is simply due to incomplete excision.

Nodular lesions in the knee are usually readily diagnosed and removed; adequate exposure should be used to ensure that no additional lesions remain undetected. Sometimes a pedunculated nodule gives the clinical picture of a loose body or torn cartilage, and its true nature only becomes apparent at arthrotomy. It is the diffuse form which causes most difficulty, the differential diagnosis resting between pigmented villonodular synovitis, tuberculosis, rheumatoid arthritis and malignant synovioma. The characteristic monarticular involvement and normal sedimentation rate excludes rheumatoid arthritis, as does the aspiration of a bloodstained effusion. This latter finding is also unusual in tuberculosis, as is the normal erythrocyte sedimentation rate. Malignant synovioma may be suspected in the presence of bone involvement, but the intra-articular origin of the lesion and the typical reddish-brown pigmented appearance should make a diagnosis of pigmented villonodular synovitis possible even before histological material has been examined.

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In all cases in which amputation is considered, a biopsy must be performed and its interpretation discussed with the pathologist; errors are less likely to occur when both the surgeon and pathologist are aware of the possibility of pigmented villonodular synovitis and have discussed the problem together. A pathologist whose experience of this type of lesion is limited should not hesitate to seek a second opinion.

Treatment of diffuse lesions of the knee by extensive synovectomy has proved to be unpredictable and in some cases unsatisfactory. There is a risk of aggravating the patient's symptoms and the chances of a complete cure are only about 17 per cent. The fact that biopsy alone was often followed by remission suggests that the condition may be self-limiting. Further evidence in favour of this view is that synovectomy, although necessarily incomplete, leads to recurrence in less than 50 per cent of cases. We therefore suggest that in this form of pigmented villonodular synovitis, once the diagnosis has been established by a biopsy or by a limited synovial resection, no further surgery be carried out as an initial procedure. Only if the patient's symptoms become serious or disabling—and this rarely occurs—should extensive synovectomy be considered; it may then have to be carried out as a calculated risk, but the possibility of subsequent arthrodesis or hinge arthroplasty must be borne in mind should it fail. Radiotherapy may also be used, particularly in older patients; we are, however, reluctant to recommend its use for a benign condition in a young patient, not only because of its possible carcinogenic effect, but also because of the considerable risk of producing disabling stiffness of the joint.

At other sites, such as the wrist or the ankle, synovectomy, if performed with care, offers a good chance of success. The hip presents a special problem, as treatment may be required for disabling symptoms at a relatively early stage in the development of the lesion. In these cases, particularly when bone involvement has occurred, the best functional results are probably achieved by more radical surgery such as arthrodesis or total replacement arthroplasty.

SUMMARY
1. The literature on pigmented villonodular synovitis has been reviewed and a series of eighty additional cases is reported.
2. The condition usually presents either as a nodule in a finger or knee, or as a diffuse lesion in a knee. The lesions, although benign, sometimes erode or invade the tissue of adjacent bones.
3. Distinction from malignant synovioma can be made on the basis of the macroscopic appearance of the lesion at operation (relationship to joints or tendon sheaths: villonodular appearance: pigmentation), and by histological examination.
4. Treatment of the nodular form by excision is satisfactory but extensive synovectomy for diffuse lesions of the knee gives poor results.
5. The etiology of pigmented villonodular synovitis is unknown, but it appears to be a self-limiting process, possibly inflammatory in nature.

REFERENCES
THE DIAGNOSIS AND TREATMENT OF PIGMENTED VILLONODULAR SYNOVITIS

Dor, L. (1898): Relations des tumeurs à myélopoxes et des xanthomes. Congrès de Chirurgie, 12, 553.


