COLLAGENOUS PSEUDOTUMOURS OF THE HANDS

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Soft-tissue tumours on the dorsum of the hand in an area between the level of the radiocarpal and metacarpo-phalangeal joints frequently present a diagnostic problem.

These "collagenous pseudotumours" seem to represent a separate entity. Cases with less swelling have been regarded as instances of rheumatoid tenosynovitis, but in fact these lesions differ pathologically from those of tenosynovitis. In the past they have not been adequately treated, and the only effective treatment that can be suggested at present is drastic surgical eradication.

Material

Six cases have been selected from a series of ten recently investigated. Particulars of the six patients are given in the Table.

Case 1, coloured female, aged 25, first seen July 7, 1945, had noted painful "masses" about the size of an egg on both hands for the past 2 years. These "masses" were extremely painful, especially on movement of the fingers. The clinical diagnosis was multiple tumefaction of both hands, possibly pigmented villonodular tenosynovitis.

TABLE

SUMMARY OF HISTORIES OF THE SIX PATIENTS WHOSE CASES ARE REPORTED

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Race</th>
<th>Age</th>
<th>Family History</th>
<th>Previous History</th>
<th>Present Illness</th>
<th>Operation</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>C</td>
<td>25</td>
<td>Mother living, has tuberculosis and cancer</td>
<td>—</td>
<td>1945 Both hands</td>
<td>Left hand</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>W</td>
<td>29</td>
<td>None</td>
<td>1938 Arthritis of left middle finger; 1942 Subdeltoid tumour (removed at operation) 1944 Arthritis of left knee (fusion operation)</td>
<td>1947 Both hands</td>
<td>Both hands (with 3-week interval)</td>
<td>Yes</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>C</td>
<td>20</td>
<td>—</td>
<td>1945 &quot;Rheumatism&quot; 1947 Contractures of finger joints</td>
<td>1948 Left wrist, hand, and fingers Left hand</td>
<td>(1) Left hand and wrist (2) Left hand</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>C</td>
<td>63</td>
<td>—</td>
<td>1945 Severe rheumatoid arthritis successfully treated</td>
<td>1947 Left hand</td>
<td>Left hand</td>
<td>Yes</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>W</td>
<td>29</td>
<td>—</td>
<td>—</td>
<td>1948 Right hand Two on right hand and one on right ankle</td>
<td>(1) Right hand (2) Right hand</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>C</td>
<td>30</td>
<td>None</td>
<td>1939 Miscarriage 1942 Hysterectomy</td>
<td>1950 Right wrist</td>
<td>Right wrist</td>
<td>Yes</td>
</tr>
</tbody>
</table>
COLLAGENOUS PSEUDOTUMOURS OF THE HANDS

Laboratory Findings.—Urine normal. Complete blood count normal. Blood sedimentation rate 35 mm./1 hr (Westergren). Tuberculin test positive. No sputum could be examined.

Operation.—Three tumours—one just over the styloid process of the ulna, one over the midportion of the fifth metacarpal, and one just between the two—were seen on the left hand. A 3-in. longitudinal incision was made over the dorsolateral aspect. Sharp and blunt dissection was carried out and much oozing was encountered. A soft, greyish, friable tumour mass surrounded the tendons of the common extensors of the fingers; the tendons were strangulated and atrophied, and the mass was so intensely adherent that it was impossible to remove all of it. After careful haemostasis and closure, an anterior moulded plaster-of-Paris splint was applied. The post-operative course was uneventful.

Pathologist's Report (Dr. H. L. Jaffe).—A number of fragments of thickened and congested tendon sheaths, were found on section to be hypertrophied, oedematous, and inflamed, and heavily infiltrated by lymphocytes, plasma cells, and macrophages, and in places by nests of polymuclear leucocytes. The inflamed tenosynovium was covered in places by fibrin exudate. This was described as "chronic but apparently still active tenosynovitis, possibly of rheumatoid origin".

Comment.—No tourniquet was used and this was one reason why the tumour could not be removed completely. It recurred in less than 5 months; was painful, and impaired tendon function. X-ray therapy (100r weekly) was given, but no improvement was noted for the next 6 months. The patient would not submit to another operation and was lost sight of.

Case 2, white female, aged 29, had been seen in 1942 when a large cystic mass containing more than 100 rice bodies, but with no evidence of tuberculosis, was removed from the right subdeltoideal area. She was reported to have had arthritis of the right middle finger in 1938, with spontaneous recovery.

In 1944 she complained of increasing pain and limitation of motion in the left knee. She connected this with an automobile accident in 1941. At this time x rays showed changes characteristic of rheumatoid arthritis. Blood sedimentation rate 37 mm./1 hr (Westergren). Formogel reaction positive. At operation the left knee joint showed extensive destruction of all its cartilaginous constituents, and appearances characteristic of rheumatoid arthritis. A fusion operation was performed and recovery was uneventful.

In 1947 a soft mass had formed upon the dorsum of each hand, 1½ in. distally to the anatomic snuffbox on the right, and more towards the centre of the dorsum of the wrist on the left. A fusiform swelling of the middle joint of both long fingers was noted. Active and passive motion of the metacarpal and proximal interphalangeal joints of the index and middle fingers was limited.

Laboratory Findings.—Sedimentation rate 39 mm./1 hr (Westergren). Blood count normal. Wassermann reaction negative.

Operation.

Right Hand: A greyish-red, almost rubbery, growth was tunnelled by and adherent to the tendons of the extensor pollicis longus and extensor carpi radialis brevis and longus in the right hand.

Left Hand: (3 weeks later) the tendons of the common extensor, indicis proprius, and digitus quintus proprius were found to be involved, some being half their usual size, though their power was not restricted.

In each case a pneumatic tourniquet was used and all the collagenous tissue removed by careful dissection.

Comment.—So far there has been no recurrence.

Case 3, coloured female, aged 20, on March 3, 1948, had painful swellings the size of an egg on the left wrist, hand, and fingers. These swellings had first been noted 3 years previously, and had remained unchanged, except for pain during the last year, especially after scrubbing.

Three years previously the patient had been in bed with "rheumatism" for 6 months, and swelling of the right wrist had been treated by "injections".

For the past year she had noted the onset of flexion contractures of the little and middle fingers at the proximal interphalangeal joints of both hands. The involved joints had never been painful or swollen.

Radiological Examination.

March 8, 1948: irregularity of outline of distal ulna and radius, and a curious beak-like projection distal to the semi-lunar in the lateral.

Laboratory Findings.—Urine and blood count normal. Sedimentation rate 38 mm./45 min. Kahn test negative. Tuberculin test negative.

A diagnosis was made of possible fibroma or lipoma of the tendon sheath.

First Operation.—A curved incision 4 in. long was made, originating over the dorsum of the second metacarpal and extending to about 1 in. above the ulnar styloid. The subcutaneous tissues were divided and the tumour mass dissected out. It was found to be constricted by the dorsal carpal ligament, located in the tendon sheath, completely encircling all the extensor tendons. It was impossible to remove it completely because of the extensive bleeding, since no tourniquet had been used. There were areas of cystic degeneration, the cysts being filled with greenish viscid fluid. The tendons incorporated in the mass showed villous surface growth and were dissected free as far as possible. The dorsal retinaculum was approximated and the hand placed in a moulded plaster-of-Paris splint.

Fluid recovered from the cystic areas showed a moderate amount of white cells, but no bacteria. The culture remained sterile for 48 hours.

Pathologist's Report (Dr. Jaffe).—Tendon sheath tissue shows delicate villous hypertrophy. Microscopic sections show sub-acute and chronic tenosynovitis suggesting rheumatoid tenosynovitis.
Later Developments.—The post-operative course was uneventful, but there was a painless recurrence of the mass on the dorsum of the left hand. It was about 4-5 × 9 cm., and apparently involved the sheath of the extensor communis digitorum as well as that of the abductor longus and extensor brevis. The skin over the swelling was warm and appeared doughy and cystic. The volar surface of the hand was not involved (Fig. 1).

Radiological Examination.

Wrists: slight irregularity in cortices of radii bilaterally at level of styloid process.

Carpal bones: normal.

Knees: large effusion in suprapatellar pouches.

Left tibia: subcondylar resorption in external condyle.

Right tibia: subcondylar resorption on outer aspect of external condyle.

Laboratory Findings.—Sedimentation rate (December 14, 1949), 72 mm./45 min. Non-protein nitrogen, 43 mg. Total protein, 8.4 per cent. Cholesterol, 169; free cholesterin, 27 per cent. Tuberculin test negative. No anaemia. No sickle cells. Fluid from cystic areas negative for tubercle bacilli, sterile after 48 hours. Guinea-pig negative for tuberculosis. Rosen test negative.

Second Operation.—This was performed with a pneumatic tourniquet. Through a 6-in. incision, the previous scar was excised. A cystic, irregular, greyish, boggy mass was found extending from the middle of the dorsum of the left hand to about 2 in. above the extensor retinaculum, superficial to the deep fascia. It was undermined and separated from the deep fascia from the proximal area down to the extensor retinaculum, where it was found to adhere intermittently to the extensor retinaculum, and to dip through it. When the muscles were exposed, this greyish mucoid granulation tissue was found to encompass the distal ends of the muscles, and the tendons of all the extensor muscles, including the pollicis longus, were also involved. The mass was dissected out from the tendons and cleaned down to the mid dorsum, removing with it the extensor retinaculum (Fig. 2, opposite). The tendons were thin and attenuated. A fascia lata graft was applied in place of the extensor retinaculum. Layer-by-layer closure was done and a cockup splint applied.

The fascial graft was extruded on the seventh post-operative day. The rest of the incision healed by primary intention.

Pathologist’s Report (Dr. Jaffe).—A frayed cyst-like structure roughly the size of a goose egg. The cyst had been opened in many places. The wall was fibrous and in many areas villous hypertrophy of the synovial lining was noted. Sections of the synovium showed villous hypertrophy and cellular hyperplasia, with many focal areas of lymphocytic infiltration as well as acute exudate. The muscle showed focal lymphocytic infiltration.

Comment.—In the light of further experience we believe that had the first operation been performed with the use of a pneumatic tourniquet, the chances of recurrence would have been lessened or even eliminated.

The patient was last seen on February 26, 1951, when there was excellent function of all the extensor tendons and no sign of recurrence.

Case 4, coloured female, aged 63, first seen in 1945 with multiple joint lesions due to rheumatoid arthritis of 25 years’ duration, had been unable to walk for the past 2 years because of pain and stiffness about the right knee, and had been bedridden for 2 years. The right knee was swollen, and the skin temperature raised; the joint was held in flexion at 110°, and no active or passive extension beyond this was possible, except with great pain. The para-articular structures were enlarged, though only a moderate amount of fluid was noticed in the joint. With the exception of the temporomandibular joints, all were affected to some degree.

Laboratory Findings.—Sedimentation rate 72 mm./1 hr (Westergren), marked secondary anaemia, Wassermann negative, serum protein normal, blood calcium 7, phosphorus and phosphatase normal, blood uric acid 4/1 mg. per cent.

Therapy.—Gold salts were administered and the patient’s anaemia corrected; the contracture and acute inflammatory lesion of the knee was treated partly by
traction and partly by manipulation, and a plaster-of-Paris encasement was applied with the knee in the position of 175°. The patient started to walk with the aid of crutches in one week after the application of the plaster, and finally made a satisfactory recovery.

Later Developments.—Two years later she reported a swelling on the dorsum of the left hand. This had increased gradually and become very painful during the past 2 months; it has been diagnosed as a ganglion and excision recommended, but we thought it was a collagenous soft tissue mass of rheumatic type, and the involvement of the tendons of the common extensor was quite obvious clinically.

Operation (February, 1947).—The gross and microscopic findings were the same as in previous cases, but cystic degeneration with accumulation of fluid was perhaps more prevalent. A pneumatic tourniquet was used and complete removal obtained by sharp dissection.

Comment.—The patient was seen last in January, 1951; the other lesions had progressed considerably, but there was no sign of recurrence of the hand lesion.

Case 5, white male, aged 29, who had been working 2 years before as a lumberjack, was first admitted on June 18, 1949, with painful swelling of the right hand; this had begun 12 months before but had only been painful for 6 months. A mass the size of an egg was seen over the dorsum of the right hand at the level of the radial styloid; whence it extended 2 in. proximally and 3 in. distally in a fusiform fashion. It seemed to be firmly attached to the common extensor tendons. Wrist motions were free, except that palmar flexion was limited to about 80 per cent. of that on left side. The mass was painful upon pressure and active motion. The grasping power of the left hand was diminished, but finger motions were not impaired. A pre-operative diagnosis of ganglion of the right wrist was made.

Laboratory Findings.—Blood sedimentation rate 8 mm./45 min., red blood count 4,960,000, white blood count 5,500, differential count normal.

First Operation (June, 1948).—No tourniquet used. Recovery uneventful.

Pathologist’s Report (Dr. Jaffe).—Two pieces of tendon sheath (3 × 1½ × 1 cm. and 4 × 1½ × 1 cm.). The inner surface was wrinkled and in places villously transformed. Parts of the inner surface were brown. A few tiny rice bodies adhered to the surface. Sections showed typical changes of rheumatoid inflammation, including...
fibrinoid necrosis, rice-body formation, and rheumatoid nodule formation.

Later Developments.—He was re-admitted on November 7, 1949, 17 months later. Slightly tender fluctuant swellings were seen on the dorsum of the right hand and the anteromedial aspect of the right wrist, and a swelling over the dorsal aspect of the right ankle about half way between the lateral and medial malleolus corresponding with the tendons of the common extensors of the toes and peroneus tertius. No limitation of motion in the joints and no increase of skin temperature.

Laboratory Findings.—Sedimentation rate normal—6 mm./45 min. Wasserman test negative. No anaemia.

Second Operation.—Masses excised similar to those removed at previous operation. Tourniquet used.

Case 6, coloured female, aged 30, had noticed a mass on the dorsum of the right wrist for 3 years, since striking her hand against a sink. The mass had gradually increased in size for the past year with a mild and constant pain, and inability to extend the fingers fully. She had had a miscarriage in 1939, and a hysterectomy in 1942. There was no history of night sweats, weight loss, or known tuberculosis. On the dorsum of the wrist was a mass measuring 1½ × 1½ × 1 in. It was a circumscribed, multilocular, cystic soft structure. The patient was unable to dorsiflex the wrist beyond the neutral position, or to extend the fingers except with the wrist flexed.

Radiological Examination.—X ray of the wrist taken immediately on admission revealed some asymmetry and spotty demineralization with relative increased densities at the styloid process of the radius.

Laboratory Findings.—Urine normal. Sedimentation rate 59 mm./45 min. Haemoglobin 9-8 g.

Operation (October, 1950).—A pneumatic tourniquet was used. The tumour, which involved the dorsal extensor tendons, was a reddish-yellow mass, and felt soft and meaty. There were numerous rice bodies, yellow, pebble-shaped masses with a very small amount of fluid, in a conglomerate gelatinous mass, through which the markedly frayed and atrophied tendons, quite adherent to each other, were shining as if they were made from wax.

It was not possible to differentiate macroscopically between collagenous disease and tuberculosis, but the latter now seemed most likely.

Pathologist's Report (Dr. Jaffe).—Microscopic examination revealed caseous and non-caseous tuberculous tenosynovitis.

Further Laboratory Findings.—Tuberculin test positive. Sputum was collected although the patient denied its existence and a few acid-fast bacilli reported on one occasion and negative findings on three others. Streptococcus agglutination positive 1 : 1,280. Wassermann positive.

Cerebrospinal fluid protein 21 mg. per cent., Kahn negative, colloidal gold normal, cell count normal.

Later Developments.—There was no primary healing of the operative incision. A sinus developed, but healed later. Diffuse swelling persisted in the operated area, but there was some improvement in the range of active extension of the fingers.

The patient next developed an acute exudative tuberculosis of the lung, and was transferred to another hospital for treatment.

Comment.—This case represents the only erroneous diagnosis in this series. It shows the difficulty of differentiating the collagenous tumours from tuberculous synovitis. Even at the time of operation it was not possible to make the correct diagnosis, notwithstanding that tuberculosis is always kept prominently in mind in dealing with the type of lesion. Had tuberculosis been diagnosed our surgical approach might have been modified by the simultaneous use of streptomycin.

Pathology

In exploring these lesions one is surprised by the extensive invasion of the surrounding tissues by the "tumour". It is particularly the tendon structure itself that is inseparably invaded, and it is hard to tell whether the growth originates in the epitelen or paratenon. The "tumour" tissue does not invade the subcutaneous or cutaneous structures, but does invade tendon and muscle tissue proper as is seen in the microscopic sections. The name tenosynovitis seems inadequate to describe these extensive lesions which far exceed the inflammatory character of tenosynovitis proper. This condition as found in the cases described above is invasive, expansive locally, and extremely cellular, and shows the characteristics of collagenous tissue. It is also destructive in that it destroys and replaces muscle tissue and diminishes the size of the tendons which become much thinner and seem to atrophy.

The gross surgical appearances differ depending whether a tourniquet is applied or not. When the tourniquet is used a greyish, almost rubbery, definitely soft, light-coloured mass bulges out, and is found upon dissection to follow the structure of the tendon sheath like a sleeve. If this tissue is incised right down into the tendon itself, it will be found that the tendon runs through it as through a tunnel and is closely adherent to it. Thus only meticulous sharp dissection will free the tissue from the tendon proper, and even so small fringes of the tissue will remain in places. These can be removed by rubbing with a dry gauze pad. In nearly every case the tendon itself was found to be almost half of its normal size. Fibrin (rice) bodies were present in half of the cases. Between the lobules of the "tumour" mass, there almost always appeared cyst-like structures filled with yellowish, turbulent, not very viscous fluid. When this fluid escaped during dissection, the mass, to some extent, collapsed. Two of these cysts were almost as large as
goose eggs (see Pathologist’s Report on Case 3). The cysts had fibrous walls with villous hypertrophy of the synovial lining.

Microscopic sections show varying histological forms. At the synovial lining, villous hypertrophy with lymphocytes, plasma cells, and fibrinoid necrosis is prevalent. In the bulk of the tumour an immense number of collagenous fibres is the usual picture, and occasionally there are a great many fibroblasts and fibrocytes. Dark and light nuclei vary and giant cells with two to four nuclei are infrequently seen (see Figs 3, 4, and 5). In no instance (apart from Case 6) were these tissues tuberculous. Greatly thickened arterial and venous walls are commonly seen with para-adventitial lymphocytic infiltration.

Discussion

Diagnosis.—“Collagenous pseudotumour of the tendon sheaths”, or “tumorous rheumatoid tenosynovitis of the hand” has its importance as
a diagnostic feature, and correct diagnosis will lead to correct treatment. During cortisone or ACTH administration these lesions regress temporarily but do not disappear. This is due partly to the absorption of the fluid in the cyst-like pouches in the lobulated mass, and partly to the elimination of the inflammatory changes.

Such tumours are by no means restricted to the hand, but this is their commonest location.

That such nodular tumours may be regarded as forerunners of generalized rheumatoid arthritis, was observed in two patients of the present series, and one other patient had such a lesion as the only rheumatoid manifestation in the tendon sheath of the tibialis posticus of the left foot.

X-ray Findings.—In all but two cases there were changes in the distal portion of the radius in the form of some irregularity, rarefaction, or atrophy. Circumscribed, "punched out" areas without the characteristics of gouty lesions were seen in the distal radius, the carpal, or the metacarpal bones and phalanges.

Nomenclature.—We have described these lesions as "tumours" because of their clinical and pathological features. There is little reason to describe them as examples of rheumatoid granulation tissue, and no evidence of pannus or necrosis.

Summary

(1) Six cases of tumorous tenosynovitis, rheumatoid in appearance, are described in detail.
(2) It is suggested that this clinical entity should be called "collagenous pseudotumour".
(3) Reasons are given for drastic surgical treatment.
(4) It is reported that these lesions were not found to be improved by cortisone or ACTH, except temporarily and to a moderate degree.

My thanks are due to Dr. Isadore Zadek for his interest and co-operation.

BIBLIOGRAPHY


Pseudo-tumeurs collagénès des mains

RÉSUMÉ

(1) On relate en détail six cas de ténosynovite tumécente d'apparence rhumatismale.
(2) On suggère que cette entité clinique devrait porter le nom de "pseudo-tumeur collagène".
(3) On présente des raisons pour un traitement chirurgical drastique.
(4) On signale que ces lésions ne sont pas améliorées par la cortisone ni par l'ACTH.

Seudotumores colagenos de las manos

SUMARIO

(1) Se describe detalladamente seis casos de tenosinovitis tumoral, aparentemente reumatoide.
(2) Se sugiere que esta entidad clínica debería llamarse "seudotumor colageno".
(3) Se aduce razones para un tratamiento quirúrgico drástico.
(4) Se señala que estas lesiones no se vieron mejoradas con el tratamiento con la cortisona o la ACTH.
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