



Epithelioid sarcoma masquerading as Dupuytren's disease

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SUMMARY. Epithelioid sarcoma is a rare and deceptive lesion, often confused both clinically and on histopathological examination with other malignant processes. The surgical course of two patients with initial diagnoses of Dupuytren's disease is described. Early biopsy of all unusual fibrotic lesions on the palm is recommended. An aggressive surgical approach to confirmed malignancy is mandatory.

Epithelioid sarcoma was first described in detail by Enzinger in 1970,¹ who analysed 62 cases of unusual sarcomas repeatedly confused with other pathological processes. Although it is the most common soft tissue sarcoma of the hand,² there have been no reports in the literature of the condition mimicking the effects of Dupuytren's contracture. In addition, we have not found any reports of malignant degeneration of the abnormal tissue of Dupuytren's disease. We report on two cases who presented with characteristics of Dupuytren's disease and who underwent initial excision of affected tissue, which was subsequently shown to be epithelioid sarcoma.

Case reports

Case 1

A 53-year-old male presented to the district orthopaedic service with progressive, painless contracture of his left little finger. A diagnosis of Dupuytren's disease was made and a palmar fasciectomy performed. After a period of several months the contracture recurred, and an examination showed evidence of fixed flexion deformity at the metacarpophalangeal joint together with skin involvement (Fig. 1).

A repeat fasciectomy was performed, followed by a ray amputation of the little finger one month later owing to persistent ulceration and deformity. The soft tissue specimens were not sent for pathological examination.

The patient was lost to immediate follow-up, but presented 8 years later to the plastic surgery service, following orthopaedic referral, with an extensive ulcer of the ulnar border of his left hand (Fig. 2A, B). He admitted that the lesion had returned soon after the amputation but that he had continued to work, with his wife dressing the lesion at home in her capacity as a nurse. Weakened grip and pain had led him to seek medical advice again, and radiological examination revealed erosion of the heads of the 4th and 5th metacarpals. An initial biopsy of the ulcer edge was reported as squamous cell carcinoma, for which the patient underwent a partial hand amputation.

The correct diagnosis was only arrived at after the tissue sample had been submitted for definitive staining, and the patient eventually underwent a mid forearm amputation. Three years following the limb ablation, he developed skin metastases of the head and neck region, for which he received radiotherapy and underwent palliative local tumour excision. He eventually died from lung and cerebral metastases 4 years later.

Case 2

A 21-year-old male joiner was referred by his GP to the orthopaedic service with a 6 month history of a painless subcutaneous nodule in the palm of his right hand. Owing to a familial history of Dupuytren's disease, this was suspected to be a Dupuytren's plaque and the patient underwent excision biopsy under local anaesthetic some 5 months later.

The surgeon at the time suspected the lesion to be of connective tissue origin, arising from the tendon sheath and surrounding tissue, and submitted the specimen for histopathological examination. The diagnosis of epithelioid sarcoma was confirmed, and the patient was referred to the plastic surgery service for urgent management (Fig. 3).

An MRI scan showed infiltration of fat by tumour superficially at the level of the 3rd and 4th metacarpals, with extension into the palm involving the intrinsic muscles (Fig. 4). A radical en bloc excision of palmar skin, 3rd and 4th lumbricals and common digital nerve to the 3rd web space was performed, with soft tissue cover provided by a distally based radial forearm fasciocutaneous flap (Figs. 5A, B). At follow-up 6 months postoperatively, he had regained full range of hand movement, with no evidence of tumour recurrence.

Discussion

Epithelioid sarcoma is a poorly understood soft tissue sarcoma affecting predominantly the distal extremities

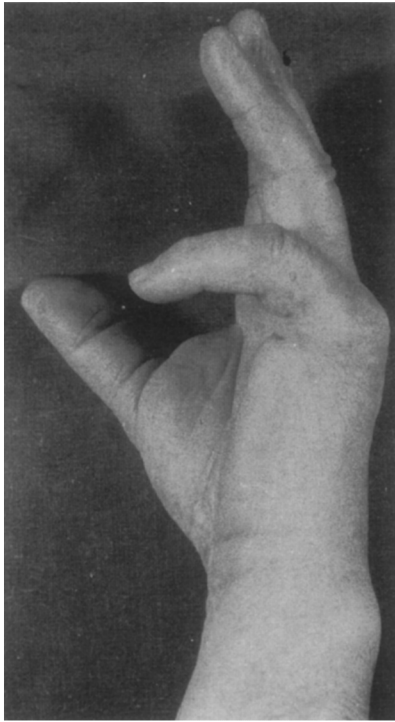


Fig. 1



Fig. 2

Figure 1—Case 1. Contracture of the left little finger due to tumour involvement of soft tissues at the MCP joint level. **Figure 2**—Case 1. (A) Palmar view and (B) ulnar view of hand showing tumour recurrence 8 years after amputation of the left little finger for suspected Dupuytren's disease.

of young adult males, and has a mortality rate of 20%.¹ It grows slowly along fascial and tendinous structures, forming multiple painless nodules which eventually necrose and ulcerate. The primary lesion may involve overlying skin and extend into the subdermal plane, resulting in subdermal lymphatic spread. It is this mode of growth that closely correlates with the natural history of Dupuytren's disease and may lead to an erroneous diagnosis. Although the initial course of the disease is slow, our two cases demonstrate the typically rapid recurrence following inadequate excision, which is variably reported as being between 63–85%.^{1,3,4}

There have been no reports in the literature on malignant degeneration of Dupuytren's disease. Tsur and Lipsker⁵ have described a patient in whom an atypical nodule at the base of a thumb was mistakenly diagnosed by the pathologist as fibromatosis. A tumour recurrence 3 years later was subsequently reported as epithelioid sarcoma and the thumb was amputated. Power *et al.*⁶ have described a spindle cell sarcoma of the hand presenting as an atypical nodule in the 2nd web space, erroneously reported as Dupuytren's disease, with subsequent recurrence after excision. In the series of Bryan *et al.*² of 85 soft tissue sarcomas of the upper limb, 13 were identified as epithelioid sarcomas after previous misdiagnoses. These included rhabdomyosarcoma, synovial sarcoma, juvenile fibroma and malignant histofibroma, but did not include fibromatoses.

A link between other fibromatoses and epithelioid sarcoma has been suggested by Moore *et al.*,⁷ who described a patient who had biopsy proven Peyronie's

disease of the penis, with subsequent extension of the soft tissue mass. Progressive focal gangrene over the ensuing 6 years eventually led to the correct diagnosis of epithelioid sarcoma being made.

A history of trauma has been suggested by a number of authors,^{3,8,9} although no direct link was identified in our series. Evans and Baer¹⁰ have identified specific prognostic features relating to distant metastases and death, which include tumour size ≥ 5 cm, presence of regional lymphadenopathy and local recurrence. Variables such as mitotic rate, perineural invasion and tumour necrosis, previously reported as significant, are now not believed to relate to early metastases.

The capricious method of spread may mimic the progressive course of Dupuytren's disease. The tendency of epithelioid sarcoma to grow proximally along tendons and fascial planes, resulting in flexion contracture, can resemble the natural history of palmar fibromatoses.³ The cells of origin are hypothesised to be either mesenchymal reserve cells or fibroblasts. An epithelial origin is doubtful, as many tumours arise in deep locations in association with tendons and fascia.

Microscopically the tumours show three particular characteristics, in the nodular arrangement of the tumour cells, their tendency to undergo central necrosis and their eosinophilic, epithelioid appearance (Figs. 6A, B). The distinctive nodular pattern of these cells varies from a well circumscribed one to an irregular fusion of the nodules. Central degeneration and necrosis is frequently noted, with occasional associated haemorrhage. The tumours may spread within fascia or aponeurosis, forming multiple bands interspersed with areas of necrosis. Lesions located within the



Fig. 3

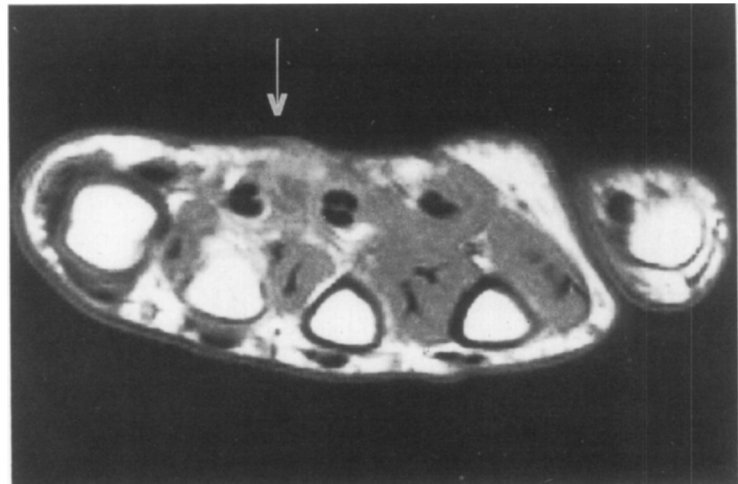


Fig. 4

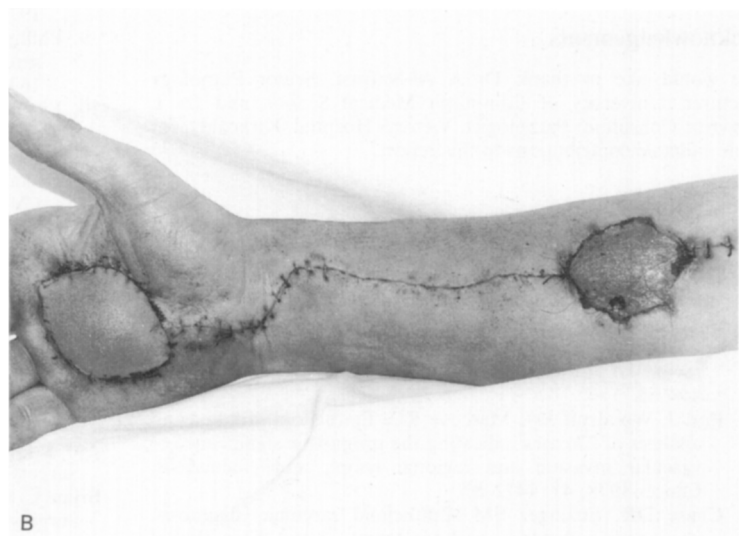


Fig. 5

Figure 3—Case 2. Scar in palm of right hand at the biopsy site for a suspected Dupuytren's nodule. **Figure 4**—Case 2. MRI scan of the right hand, showing tumour involvement of the palmar skin (arrow) and extension between the third and fourth lumbricals. (Axial gradient echo T1 weighted image.) **Figure 5**—Case 2. (A) Intraoperative photograph showing extent of radical excision of soft tissues within the palm. (B) Distally based radial forearm flap used to close the palmar defect.

dermis often ulcerate through the skin, resembling an ulcerating squamous cell carcinoma, and occur most frequently in areas with minimal subcutaneous fat, such as the fingers. Epithelioid cellular elements may vary from large polygonal cells to spindle-shaped cells resembling a fibrosarcoma or malignant fibrous histiocytoma. Multinucleated giant cells are infrequently noted and cellular pleomorphism is minimal. The distinct biphasic pattern characteristic of synovial sarcoma is not seen. Calcification and dysplastic ossification may occur in up to 20% of cases, although cartilaginous metaplasia is uncommon.¹¹

Our two cases illustrate the natural history of epithelioid sarcoma, being that of recurrence and

repeated operations, with the diagnosis often only being made after three or four excisions.

Enzinger,¹ Prat *et al.*,³ and Chase and Enzinger⁴ recommend prompt wide en bloc excision or radical mid forearm amputation if the lesion recurs. Simultaneous axillary lymph node dissection is also advised by Peimer *et al.*¹² The reported experience with chemotherapy and palliative radiation has not been encouraging, and is limited to cases with advanced metastases, with no 'cure' directly attributable to their use.⁴

We would suggest that any unusual nodular fibrous lesion on the hand, even in the presence of a familial Dupuytren's diathesis, be treated with the utmost

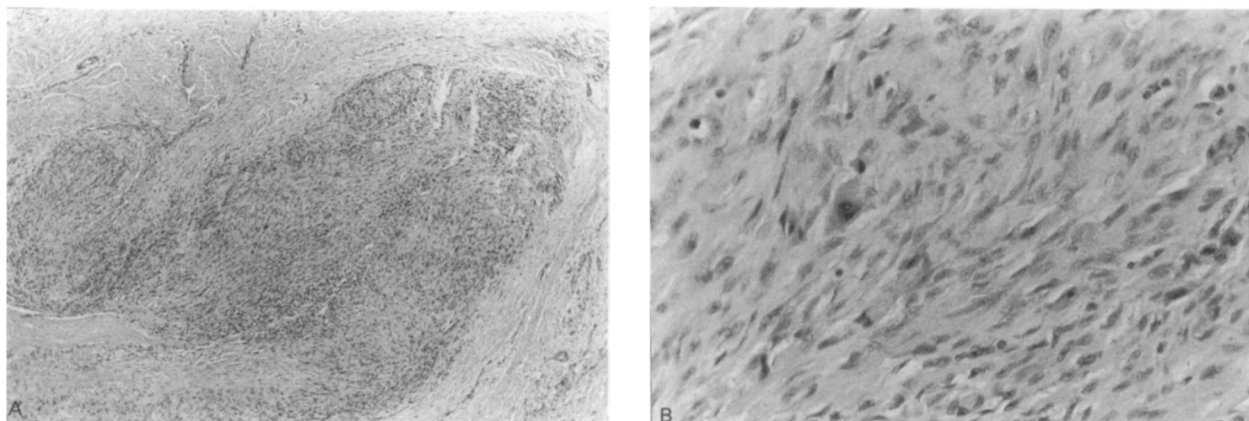


Fig. 6

Figure 6—(A) Low power photomicrograph showing nodular orientation of the tumour cells (H&E \times 100). (B) At high magnification, the dominant cells are seen to be large and epithelioid, appearing round or polygonal in shape. Numerous mitoses can be observed (H&E \times 800).

suspicion and should certainly be submitted for histopathological examination.

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