CASE REPOSITORY

Intramuscular Epithelioid Sarcoma Presenting as Extrinsic Flexor Tightness in the Forearm

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Epithelioid sarcoma is an uncommon soft tissue sarcoma involving predominantly the distal extremities of adolescents and young adults. Its rarity makes it difficult to diagnose accurately and treat properly in the early stages. We discuss the delayed diagnosis of a 37-year-old man who presented with extrinsic flexor tightness of the wrist and fingers. We initially thought that the lesion resulted from inflamed soft tissue of the flexor muscles causing contracture. However, histological examination of a biopsy specimen revealed nodular proliferation of epithelioid and spindle cells, which were immunoreactive to epithelial and nonepithelial markers, respectively, leading to the final diagnosis of epithelioid sarcoma. *(J Hand Surg Am. 2018;■(■):1.e1-e5. Copyright © 2018 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Epithelioid sarcoma, extrinsic flexor tightness, flexion contracture, forearm, soft tissue sarcoma.

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EPITHELIOID SARCOMA IS AN uncommon soft tissue sarcoma first described by Enzinger[1] in 1970, predominantly affecting the distal extremities of adolescents and young adults.[1–4] Although epithelioid sarcoma is one of the most common soft tissue sarcomas of the upper extremity,[1–4] the condition is frequently misdiagnosed, which can result in inadequate surgical resection.[1] In this case report, we present the delayed diagnosis of an epithelioid sarcoma in a 37-year-old man who presented with extrinsic flexor tightness of the wrist and fingers.

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CASE REPORT

The patient was a 37-year-old man with a 1-year history of finger numbness and painless swelling of the left upper limb without a history of trauma. No abnormalities of the affected forearm were detected on radiographs, which were taken at an outside hospital. Despite more than 1 year of nonsurgical treatment, symptoms worsened, which prompted a visit to our institution. During the initial visit, the patient reported a tingling sensation in the distribution of the median nerve. Full extension of the wrist and fingers was not possible because of extrinsic flexor tightness in the forearm (Fig. 1). Although no abnormalities were detected on plain radiographs, magnetic resonance imaging showed a high-intensity area on T1- and T2-weighted images in the flexor compartment of the left proximal forearm (Fig. 2).

Because diagnostic imaging did not provide a definitive cause for the flexion contracture of the patient's fingers, surgery was considered to be indicated and was performed under general anesthesia. Intraoperative findings showed that the flexor

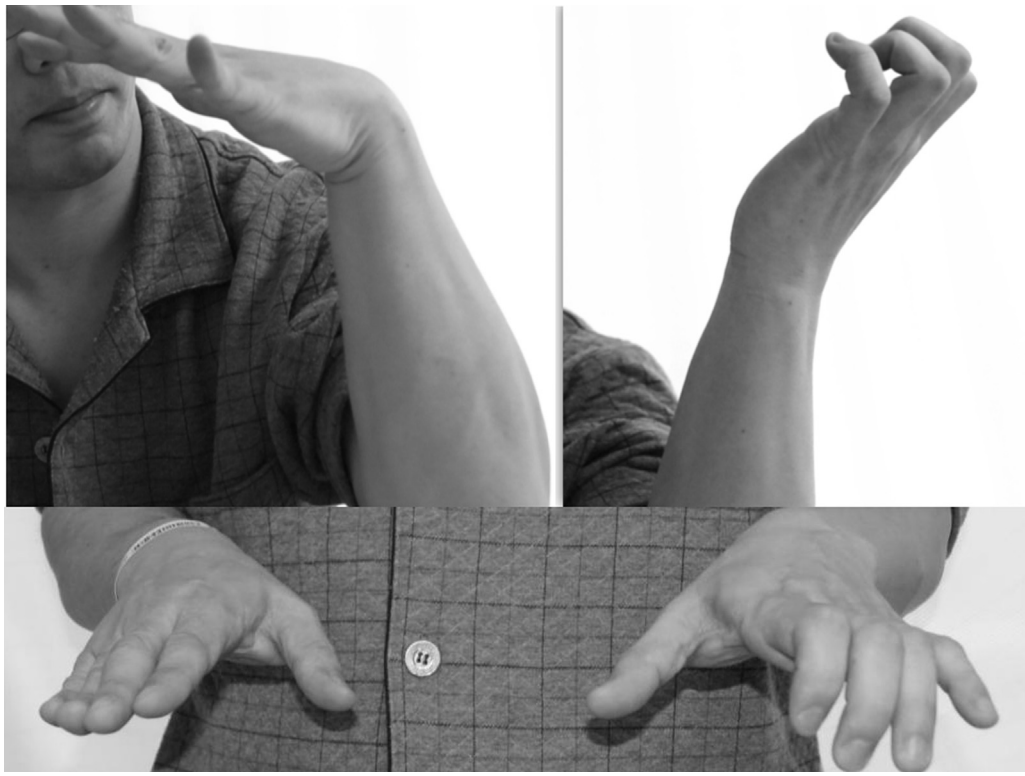

FIGURE 1: Preoperative photographs showing flexion contracture.

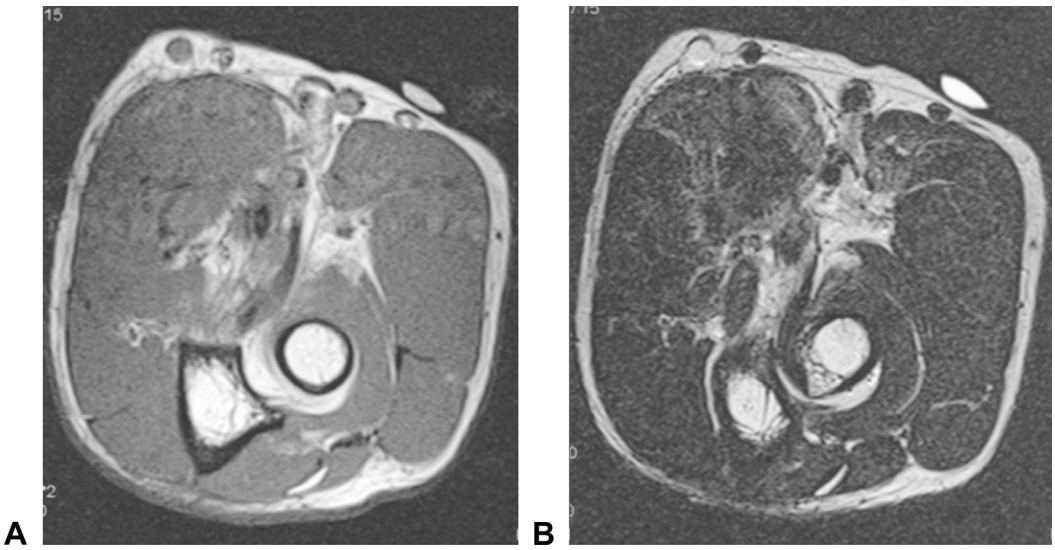

FIGURE 2: Preoperative magnetic resonance images of the left forearm, showing a partial high-intensity area on (A) T1- and (B) T2-weighted images in the flexor muscle compartment.

muscles contained visible scar-like tissue with no obvious lesions or masses (Fig. 3). The scar-like tissue was resected and sent for pathological analysis. In the immediate postoperative period, finger extension improved after this excision.

Histological examination revealed sheets of plump, oval to spindle-shaped atypical cells with prominent nucleoli accompanied by focal necrosis, suggestive of a malignant tumor. The tumor exhibited predominantly a nodular growth pattern and an epithelial appearance (Fig. 4A) with wide infiltration into the flexor muscles of the forearm; lymphatic and vascular invasion was suspected. High-power views of the histology specimen revealed tumor cell