Intramuscular spindle cell hemangioendothelioma

Abstract  Spindle cell hemangioendothelioma occurring in skeletal muscle is extremely rare. No reported studies have performed an imaging evaluation of intramuscular spindle cell hemangioendothelioma. We report on such a tumor arising in an unusual site, the right extensor digiti minimi, in a 46-year-old woman. An en bloc resection was performed and the patient has been disease free for 8 years. Radiologic imaging in the present case showed similar findings to those described in intramuscular hemangioma.

Key words  Spindle cell hemangioendothelioma, skeletal muscle, MRI, forearm

Introduction

Spindle cell hemangioendothelioma (SCH) is a peculiar vascular neoplasm first described by Weiss and Enzinger in 1986 [1]. They reported 26 patients with this neoplasm, which they described as a low-grade vascular tumor that shares histologic features with Kaposi sarcoma and cavernous hemangioma. As the tumor behaves in an indolent manner and is subject to local recurrence, they described these lesions as spindle cell hemangioendothelioma, representing a vascular tumor of intermediate malignancy. Most of the reported SCHs involve the dermis and the subcutaneous tissue, with occurrence in skeletal muscle being extremely rare. This report describes the imaging features of intramuscular SCH.

Case report

A 46-year-old woman had a tumor on the extensor aspect of her right forearm for 1 year, with intermittent discomfort and mild tenderness localized to that area. The mass was becoming increasingly tender. Her symptoms did not limit her activity and her medical history was otherwise normal. Physical examination revealed a 6-cm palpable mass in the right extensor muscles (Fig. 1A). There were no overlying skin changes, with a full range of motion and strength in her elbow and wrist and no neurovascular compromise.

A CT scan revealed a soft tissue mass, which minimally enhanced with contrast (Fig. 1B). MR images showed a heterogeneous 6×2×2 cm mass in the right extensor digiti minimi (EDM) (Fig. 1C,D). The lesion demonstrated markedly increased signal intensity on the T2-weighted images (Fig. 1D). No other lesions were seen. Bone scintigraphy and a CT scan of the chest revealed no metastasis. Histology done on a biopsy...
of the neoplasm suggested SCH. An en bloc resection including the entire muscle belly of the EDM and a portion of the extensor digitorum communis (EDC) was performed. The distal cut end of the EDM tendon was woven through the EDC tendon and sutured in the tendon.

The surgical specimen measured 6×2×1 cm and appeared poorly circumscribed. Centrally, the lesion had a soft, hemorrhagic cut surface. Microscopic examination revealed many cavernous blood-filled spaces lined by endothelial cells (Fig. 2A). These vascular spaces were separated by interlacing arrays of elongated, fibroblast-like cells with spindle to slightly plump nuclei with a fine chromatin pattern (Fig. 2B). No mitoses were observed. Admixed with this pattern were foci of somewhat rounded epithelioid endothelial cells. Some cells possessed intracytoplasmic lumina or vacuoles. A small amount of collagen and a few chronic inflammatory cells were present in the stroma.

The right forearm and hand were splinted with the wrist in dorsiflexion for 4 weeks. At 2 months postoperatively, the patient gained full extension of the right wrist and fingers with no functional limitations. She has been disease free for 8 years.

Discussion

The neoplasm in this report was characterized by a mixture of cavernous blood vessels and intervening areas of spindle cells, which resembled Kaposi sarcoma. These histologic features are consistent with the recently recognized vascular tumor termed “spindle cell hemangioendothelioma (SCH).” SCHs are usually localized to the dermis and subcutaneous tissues. They may be confused clinically with hemangioendothelioma (SCH), dermatofibroma, or Kaposi sarcoma. The present patient had a solitary lesion in muscle. To date, 71 cases of SCH have been reported in English language journals [1–11]. Among these cases, only four patients (5.6%) were found to have intramuscular lesions [2–7]. Ono et al. reported a case of SCH occurring in the left gluteus maximus muscle [7]. Ding et al. reported two additional intramuscular tumors, which were