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Hemangiopericytoma of the Forearm

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ABSTRACT

Hemangiopericytomas are rare soft tissue tumors of vascular origin. They occur in any organ but most frequently have been reported in the extremities. We report a case of hemangiopericytoma of the left forearm treated with optimal debulking surgery followed by postoperative adjuvant external beam radiotherapy using intensity modulated radiotherapy (IMRT). The value of recurrence-free survival after adjuvant radiotherapy is discussed. Currently, the patient is free of disease and disability at a mean follow-up period of 5 years.

INTRODUCTION

Hemangiopericytomas are rare tumors with aggressive behavior characterized by high rate of local recurrence and distant metastases[1,2]. Approximately 300 new cases of hemangiopericytomas have been reported in the literature[3,4]. Basically, hemangiopericytomas are soft tissue sarcomas of vascular origin arising from pericapillary cells or pericytes of Zimmerman[5].

The tumor was first described by Schmidt in 1937 and named by Stout and Murray in 1942[6]. Hemangiopericytomas have been reported in the extremities, pelvis, head and neck, back, retroperitoneum, abdomen, central nervous system and bones[5]. Surgery is the primary mode of treatment[7]. The value of radiation treatment in the management of hemangiopericytomas is controversial[8,9]. Local recurrence or metastasis can occur several years after the treatment[5].

The aim of this presentation is to highlight the rarity, aggressive behavior and treatment of hemangiopericytomas.

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THE CASE

A forty-two-year-old woman presented with four months history of progressive painful swelling of the anterior compartment of the left forearm. There was no history of trauma. She was a previous smoker, non-alcoholic and no history of drug abuse. Her medical history was significant for gastro-esophageal reflux disease, irritable bowel syndrome and bronchial asthma on medications. Clinical examination showed a firm, non-tender, non-pulsatile, soft tissue mass measuring 14x10 cm in the anterior aspect of the proximal left forearm. There was no neurological deficit or limitation of the joint movement and no palpable lymph nodes.

Ultrasound of the left forearm showed 7x4 cm mixed echoic mass deep to the muscles with increased vascularity. MRI of the left forearm showed well-defined lobulated lesion in the proximal and mid-forearm measuring 4.5x5x10 cm, compressing the neurovascular bundles and causing cortical thickening of the adjacent radius. The lesion started distal to the attachment of the biceps and brachialis tendons and abutting the anterior cortex of the radius. The lesion extended between the lateral margin of the radius and the adjacent muscle. There was no adjacent lymph node enlargement. Staging work-up including CT scan of chest, abdomen and pelvis and whole body bone scan showed no evidence of distant metastasis.

The FNAC and core biopsy from the lesion were non-conclusive. On 13 November 2007, the patient had wide local excision of the lesion. No significant postoperative complication was noted. On gross examination, a thinly encapsulated fleshy multi-lobulated tumor; the largest lobule measuring 9x6x3.5cm with grayish-pale whorled cut surface. Microscopically, it showed spindle fibroblast-like cells arranged as tight clusters and bundles with rare storiform pattern with empty blood vessels in-between and few lymphocytic sprinkling, see figure 1. Cellular pleomorphism was focal and minimal without increased mitosis (0-1 mitosis per 20 HPF) or necrosis, see figure 2. No foam cells or giant cells were noted. The surrounding skeletal muscle fibers were not involved by the neoplasm. There was no evidence of extension of the tumor outside the capsule. Immunohistochemistry showed that Vimentin CD34 was diffusely positive, whereas MyoD1, SMA, Desmin, S-100, and factor VIII related antigen were negative. Based on light microscopic features and supported by immunohistochemistry findings, a diagnosis of hemangiopericytoma of soft tissues (forearm) was made with predicted benign behavior.

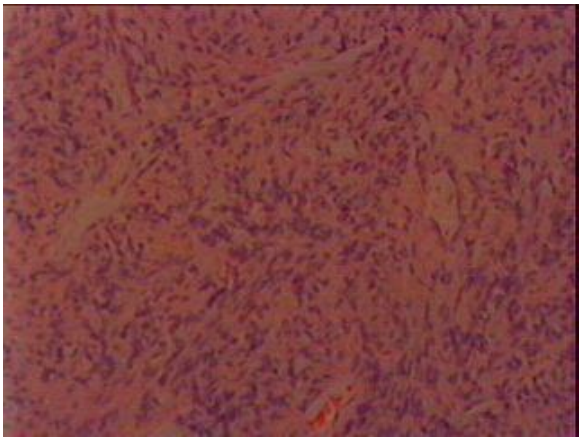

Figure 1: Spindle Fibroblast-like Cells Arranged around Thin Capillaries with Vague Storiform Pattern (H&E stain x100)

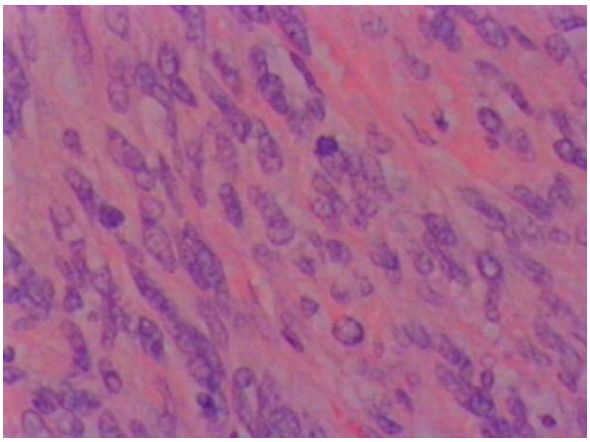

Figure 2: Spindle Cells Clearly with Minimal Pleomorphism with a Rare Mitosis (H&E stain x 400)

The patient received postoperative external beam radiotherapy using IMRT technique to the preoperative tumor volume with the margin. A total dose of 50.4 Gy in 28 fractions was given between 9 January 2008 to 21 February 2008, see figure 3.

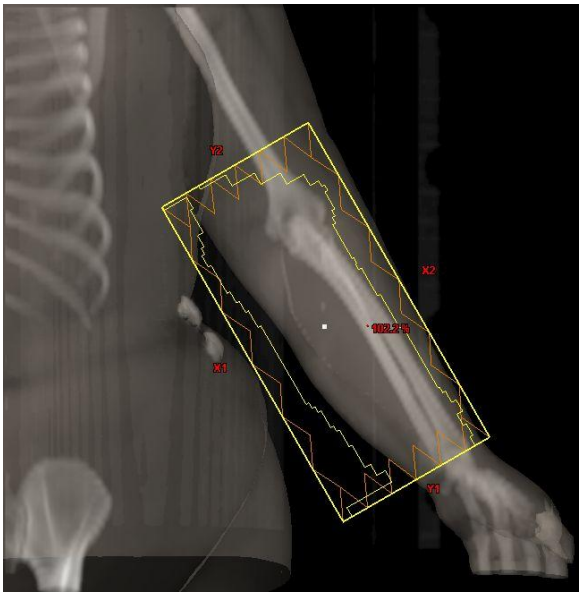

Figure 3: Digitally Reconstructed Radiograph (DRR) Showing the Site of Irradiation

The patient has been on regular follow-up and remained asymptomatic with normal function of the left upper limb for five years. Regular follow-up scans have shown no evidence of any local spread or distal metastasis.

DISCUSSION

Hemangiopericytomas are locally aggressive tumors; hence, local recurrence is common after complete surgical excision[9,10]. About two-third of cases develop local recurrence prior to metastases[10]. Lung and bones are the common sites of metastases[10]. In view of unpredictable behavior of this tumor, adjuvant radiotherapy is usually advised[5]. Hemangiopericytomas appear to be relatively radiosensitive and radiocurable tumors[9]. Since most of these tumors are large at diagnosis, there is significant risk of residual microscopic disease after wide local excision even in patients with negative margins; therefore, Staples et al advised postoperative radiation therapy[8,9]. A total tumor dose of 50-55 Gy in 5-5.5 weeks have been recommended in the literature[2,9]. Adjuvant external beam radiotherapy after initial surgery was found to extend the disease-free interval from 154 months to 254 months, although, it did not prevent the development of metastasis[1]. The mean recurrence-free interval was found to be 126.3 months long in those patients with external beam radiotherapy and complete resection[1].

CONCLUSION

Hemangiopericytomas are rare tumors with unpredictable behavior. Adjuvant postoperative radiotherapy provides prolonged recurrent-free survival. However, long-term follow up is needed as this tumor tends to recur after a prolonged period post treatment.

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