Introduction

Hemangioendothelioma (HE) is a vascular neoplasm composed of endothelial cells and is considered to be of an intermediary pathology between hemangioma and frankly malignant angiosarcoma [1]. These tumors can occur in almost all locations but rarely involve the neuraxis. Three histologic subgroups including epithelioid, spindle cell and malignant endovascular HE have been defined [2]. The etiology is unknown, but it may be associated with vascular malformations involving other organs. It can also be associated with hemolytic anemia, thrombocytopenia and coagulopathy [3].

We describe a rare case in the literature of an intraspinal (intradural with intramedullary extension) HE occurring in a young man with a complete resolution of residual tumor seen on MRI after postoperative radiotherapy.

Case Report

In March of 1999, a 41-year-old gentleman presented with a 3 months’ history of low back pain that radiated to his right leg. His pain developed acutely and was predominantly right-sided. The pain progressed despite analgesics and physiotherapy. One month later, he developed numbness in the right inner thigh, scrotum and perianal region. He also developed urinary retention with dribbling, difficulty with defecation and abdominal distention. Clinical examination revealed a decreased sensation to light touch, pinprick and temperature in the right gluteal and perianal region. Deep tendon reflexes in the knee and ankle were absent. An urgent CT-myelogram showed a complete blockage at the level of T12–L1. An irregular, lobular, markedly contrast enhancing mass opposite to T12 vertebral body was seen on MRI (fig. 1).
The patient then underwent a laminectomy from T12–L1. After removal of the overlying bone, the dura on the right-hand side was found to have a slightly bluish discoloration. The dura was tense with a firm mass palpable beneath the surface. Upon opening of the dura, a blue-black tumor was identified with many vessels present on its surface. The tumor was attached to the filum terminale. A biopsy was taken and the tumor was found to be quite vascular and firm. After severing the filum and dissecting the surrounding roots from the tumor, dissection proceeded superiorly, separating the tumor from the pial surface of the spinal cord. Debulking of the tumor was accomplished by using an ultrasonic aspirator. Near the superior portion, the tumor was found to extend into the spinal cord. This intramedullary portion was carefully dissected from the spinal cord. The tumor was separable from the spinal cord in most places but there was no well-developed capsule. Using bipolar coagulation and ultrasonic aspirator, gross removal of all visible tumor was completed. Augmentation duroplasty was then performed with a dura graft and the wound was closed in layers.

The pathology specimen consisted of multiple fragments of soft hemorrhagic tissue measuring in aggregate approximately 2.0 × 3.0 cm. This was fixed in 10% buffered formalin. Histologic sections were stained with hematoxylin and eosin (HE) in the standard fashion, as well as with an extensive immunohistochemical panel. Immunohistochemical stains included endothelial cell markers, factor VIII, CD34, CD31, vimentin, glial fibrillary acidic protein,

**Fig. 1.** Preoperative sagittal T1-weighted postcontrast MRI scans showing an irregular, lobular, markedly enhancing mass (arrowed) opposite the T12 vertebral body.

**Fig. 2.** a Histology of the lesion showing an irregular anastomosing network of vascular channels with plump lining endothelial cells and a similar intervening cell population in more solid regions. HE. Original magnification ×100. b Immunohistochemistry of the lesion, showing positivity for endothelial cell marker CD34, outlining the vascular network and many of the intervening cells. Immunohistochemistry, antibody to CD34. Original magnification ×250.
keratins CAM5.2, AE1/AE3, neuron-specific enolase (NSE), S100, epithelial membrane antigen, and synaptophysin.

The lesion consisted of a vascular tumor composed largely of an irregular aggregate of vascular spaces with an endothelial cell lining and an intervening population of similar endothelial cells (fig. 2a). Vascular channels varied in caliber from very small capillary type vessels to larger more ectatic back-to-back vascular arrangements. Varying degrees of intervening collagenous stromal fibrous tissue and focal area of myxoid stromal change were present. There was evidence of previous intratumoral hemorrhage with hemosiderin deposits. The endothelial cells ranged from bland to focally more plump, large and atypical forms that were polygonal to spindled. In areas, individual cells showed intracellular cytoplasmic vacuoles. Mitotic activity was not present. The immunohistochemical profile confirmed the endothelial and vascular nature of the lesion with both the intervening as well as vascular space lining cells showing positivity for the endothelial cell markers factor VIII, CD34, CD31 and vimentin (fig. 2b). The cells were negative for keratin, NSE, S100, EMA, GFAP and synaptophysin. A diagnosis of hemangioendothelioma was made.

Postoperatively, the patient’s urinary and bowel symptoms resolved but he continued to have mild perianal numbness and left hip flexor weakness for which he used a cane. A postoperative MRI demonstrated a small enhancing nodular area at the right posterior conus at the T12 level, measuring less than 1 cm in length (fig. 3). This was adjudged to represent residual disease on radiology review and radiotherapy was offered to improve local control. He received a total of 50.4 Gy in 28 fractions over 5.5 weeks (1.8 Gy per fraction). He was followed on a regular basis with clinical examination and MRI scans. The patient is doing well with no radiation toxicity and no residual disease on the MRI at 48 months’ follow-up (fig. 4).

**Discussion**

HE is a neoplasm composed of endothelial cells, and is considered to represent a neoplasm of intermediate or borderline malignant potential [1]. More recently, histologic subtypes, including epithelioid, spindle cell and polymorphous forms, have been described [2]. The present case conforms best to the polymorphous subtype of HE demonstrating some variability in morphologic pattern with spindled cell areas, more polygonal epithelioid areas, as well as variable vascular density within the lesion.

The treatment of choice for spinal HE is complete excision of the tumor. Although radiation has been used as an adjuvant treatment, the role of irradiation has not been clearly defined. In 1980, Pearl et al. [4] treated a benign epidural HE at T8 spine with surgery and postoperative radiotherapy (radiation = 30 Gy/10 fraction) with good results. Mahdavi et al. [5] reported an intradural-extramedullary lesion that recurred twice and was irradiated after the third resection. The first local recurrence occurred 8 months after the first operation. The second local recurrence happened at about 4 months from the second surgery. However, when radiotherapy was delivered postoperatively after the third resection, no disease was seen on MRI for 28 months. Unfortunately, it is unclear whether partial or gross total resection was performed. Ronca-
Hemangioendothelioma of the Spinal Cord

References