Malignant Hemangiopericytoma: A Clinical Overview and Case Study

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Hemangiopericytoma (HPC) is considered an unusual perivascular tumor. It is believed to originate from the cells that surround endothelial tissue, which are known as pericytes of Zimmerman (Marcus, Post, & Mancuso, 1994). Therefore, HPC can occur wherever endothelial tissue exists and develops primarily in the musculoskeletal system or skin. The function of pericytes is uncertain, but they are believed to provide mechanical support to enable the capillaries to have contractile power (Perez & Chao, 1998). HPC occurs in both sexes with equal frequency and is found primarily in adults and rarely in children (Perez & Chao). Congenital or infantile HPC usually behaves as a benign tumor despite histologic evidence of mitosis and increased cellularity (Brock, Morgan, & Anderson, 1995). Cases of spontaneous regression in congenital HPC have been reported (Brock, Morgan, & Anderson; Chen, Kassel, & Medrano, 1986). However, this phenomenon is rare when present in the adult population. The etiology of this disease is uncertain, and no strong clinical data exist to indicate a convincing link to specific causative agents. Some reports indicate a relationship between HPC and occupational vinyl chloride exposure (Hozo et al., 2000), as well as herbicidal exposure (Vietnam Veterans of America, 2002).

A comprehensive literature review revealed many cases of HPC in numerous parts of the body. The most common sites were the head and neck, followed by the lower extremities and retroperitoneum (Weiss & Goldblum, 2001). Most arise from soft tissue (Daugaard, Hurltberg, Hou-Jensen, & Mouridsen, 1988), which usually causes them to be classified as sarcomas. In about three-fourths of cases, the tumors are well circumscribed or encapsulated (Rosai, 1996). They grow relatively slowly and are considered malignant when the mitotic rate exceeds four per high power-field, they have a foci of necrosis, and they have increased cellularity (Perez & Chao, 1998).

The diagnosis of HPC is somewhat controversial and often is a diagnosis of exclusion. HPC has a highly vascular pattern, usually highlighted by a reticulin stain (Saldanha & Pia, 2001). This, however, can be present in other forms of connective tissue tumors. Pathologically, HPC also must stain negatively for muscle, nerve sheath, and epithelial markers (Brooks, 1994). Malignant HPC cases represent less than 1% of all vascular tumors and about 5% of all sarcomatous tumors (Kiefer, Wertzel, Freudenberg, & Hasse, 1997). Because malignant HPC is a rare and often incurable disease, not much advancement has been made in the way of treatment and cure. Much of what has been published about the disease is from the 1970s and 1980s, with recent journal publications limited mostly to specific case studies. Many cases of HPC have an indolent behavior, although some, like the one outlined in this article, behave like high-grade sarcomas (Enzinger & Smith, 1976). The literature reports only one family in which three members were diagnosed with HPC, suggesting no evidence of an overall increased familial incidence (Plukker et al., 1988).

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