Case report 682

Datla G.K. Varma, M.D., Angela Moulopoulos, M.D., Alan S. Sara, M.D., E. Edmund Kim, M.D., and Sidney Wallace, M.D.

Departments of 1 Diagnostic Radiology and 2 Pathology, The University of Texas M.D. Anderson Cancer Center, Houston, Texas, USA

Imaging studies

Fig. 1. A A sagittal, T1-weighted magnetic resonance (MR) image (SE 800/25) reveals a mass of intermediate signal intensity in the distal end of the posterior part of the thigh. The signal intensity of the mass is slightly higher than that of muscle. B An axial, T1-weighted MR image (SE 800/25) demonstrates the mass abutting on the distal posterior femoral cortex without evidence of cortical destruction. Note normal intensity of the femoral bone marrow.

Fig. 2. An axial, T2-weighted MR image (SE 2500/80) reveals a mass of high signal intensity with interspersed low intensity septations.

Clinical information

This otherwise healthy, 16-year-old, white, male patient was referred to the M.D. Anderson Cancer Center with a 4-month history of a growing mass in the right popliteal fossa, causing him discomfort during exercise. Physical examination revealed a 5 x 8 cm painless mass in the back of the knee with no associated skin changes. The range of motion of the knee joint was normal. The patient was free of systemic symptoms. Laboratory evaluation was unremarkable.

Plain radiographs of the right knee obtained at another institution revealed a mass in the soft tissues posterior to the knee with no evidence of bone abnormality (film not available). Magnetic resonance imaging (MRI) was then performed here in axial and sagittal planes. T1-weighted images demonstrated a mass of intermediate signal intensity abutting on the periosteum of the distal femoral cortex (Fig. 1). T2-weighted images revealed high signal intensity within the mass with interspersed low signal intensity septations (Fig. 2). The femoral bone marrow was normal. Fine needle aspiration of the mass was performed, subsequently followed by several courses of intraarterial chemotherapy. The mass was resected 5 months following the needle aspiration.

Address reprint requests to: Datla G.K. Varma, M.D., Department of Diagnostic Radiology, Box 57, University of Texas M.D. Anderson Cancer Center, 1515 Holcombe Blvd., Houston TX 77030, USA

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Diagnosis: Neuroepithelioma of soft tissues of the knee

Cytology smears of the fine needle aspirate disclosed aggregates of small, round to oval, malignant cells with Homer-Wright rosette formation, hyperchromatic nuclei, and scant cytoplasm (Fig. 3). Electron microscopy showed uniform cells with interdigitating, elongated cell processes, which focally contained neurosecretory granules (Fig. 4).

At surgery, an 8 x 4 x 2 cm, firm, oval mass which was adherent to the periosteum of the distal femur was resected. The cut surface was white-tan, fibrotic, and solid, with an absence of cystic degeneration. Microscopically, the chemotherapy-treated tumor was composed of large areas of hyalinized fibrosis with interspersed foci of viable tumor (Fig. 5).

Postoperatively, the patient received radiotherapy over a course of approximately 2 months. Some 24 months following resection, he developed a permeative, osteolytic metastatic lesion in the proximal portion of the left femur, accompanied by linear periosteal reaction and confirmed by percutaneous aspiration and biopsy to be metastatic neuroepithelioma (Fig. 6).

The patient is alive approximately 30 months after the initial diagnosis and is being treated for his metastatic bone disease.

Discussion

Since Stout presented the first case of malignant peripheral neuroectodermal tumor (MPNT) in the forearm in 1918, there have been relatively few publications on this exceedingly rare tumor that is believed to originate from the neuroepithelium of primitive neuroectodermal tissue [5]. In 1932, the term peripheral neuroepithelioma was introduced by Penfield to designate primitive neuroectodermal tumors arising outside the central nervous system; this appears to be the name preferred by most authors [4].

Peripheral neuroepitheliomas are more frequently encountered in

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Pathological studies

Fig. 3. A smear for cytological evaluation shows an aggregate of malignant, hyperchromatic, small, round cells with foci of Homer-Wright rosette formation (arrows) (Papanicolaou stain x 40)

Fig. 4. An electron micrograph shows malignant cells with intertwining cell processes containing occasional neurosecretory granules (arrows) (x 4000)

Fig. 5. A histological section of the resected tumor after treatment shows small malignant cells and bands of hyalinized collagen (H & E x 10)