Image of the Month

Tracheal obstruction caused by cervical malignant peripheral nerve sheath tumor

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Figure 1.

Figure 2.

Figure 3.

Figure 4.
A 73-year-old man presented with dyspnea and needed tracheal intubation. He had no past medical or family history of neurofibromatosis. Physical examination revealed no evidence of palpable mass, lymphadenopathy or pigment macule. Hematological investigation revealed normal levels of tumor markers. A contrast-enhanced computed tomography demonstrated a 55-mm, low-density mass in the posterior mediastinum without enhancement, and no evidence of metastases (Fig. 1). A contrast-enhanced magnetic resonance imaging demonstrated a posterior mediastinal tumor, appearing heterogeneously high on T2-weighted imaging, low on T1-weighted imaging and markedly high on diffusion-weighted imaging (Fig. 2). The tumor showed peripheral enhancement with central necrosis. Echo-guided biopsy of the lesion was performed. Light microscopy revealed nests of spindle cells with abnormal nuclei, positive staining for bcl-2 and β-catenin, but negative staining for other markers. The MIB-1 index was >20%. Sarcoma was confirmed, but the subtype was not determined. Resection was not considered possible due to suspected invasion to the cervical spine and trachea apparent on imaging. Tumor growth was rapid and caused tracheal obstruction. The patient died 31 days later due to respiratory insufficiency. On autopsy, the tumor was found to be 82 × 65 × 45 mm in size, white and solid, with necrosis (Fig. 3). The tumor invaded the esophagus and trachea, and was coalescent with the spine. Light microscopy revealed nests of spindle cells with abnormal nuclei. The cells were focally positive for S-100 (Fig. 4), but other stains yielded negative results. Electron microscopy revealed spindle cells with non-tapered branching cytoplasmic processes (Fig. 4). Malignant peripheral nerve sheath tumor (MPNST) was confirmed based on the immunohistochemistry and electron microscopy findings. MPNSTs are defined as any malignant tumor arising from a peripheral nerve or showing nerve sheath differentiation, with the exception of tumors originating from the epineurium or peripheral nerve vasculature. MPNST should be considered among the differential diagnoses of rapidly growing tumors.