Moans, Palpable Groin, and Entrapment of Bone: A Case of Malignant Peripheral Nerve Sheath Tumor in an otherwise Healthy Hispanic Male

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Background: Malignant peripheral nerve sheath tumors (MPNTs) are rare malignant soft tissue sarcomas that have an incidence of about 0.001 %. MPNTs typically occur in individuals who have neurofibromatosis or secondary to radiation therapy and rarely occur sporadically [1,2] We present a case of a previously healthy 56-year-old gentleman who was diagnosed with MPNTs.

Case: A healthy 56-year-old gentleman presented with worsening LLQ abdominal pain for 6 months. Associated symptoms included bloating, LLE swelling, early satiety for the past 2 months, and a 5-10lb unintentional weight loss. Patient denied recent cough, night sweats, dyspnea, fever, chills, melena or hematochezia. Vitals were WNL. Physical examination revealed a palpable mass on LLQ extending into the groin and edema of the left leg with diminished strength 3/5. CBC and CMP were unremarkable. CT abdomen showed a large necrotic mass in the left retroperitoneum infiltrating along the iliopsoas musculature, extending into the left hip and into the left side of L3, L4, L5 vertebral bodies and through L4 transverse process measuring up to 24.6 x 11.5 x 13 cm. Pathology revealed spindle cell sarcoma composed of moderately atypical, elongated spindle cells positive for vimentin, with loss of H3/K27me3, and negative for SMA, S-100 consistent with MPNSTs.

Conclusions: The is a rare case of a sporadic presentation of MPNTs. Treatment depends on the extent of tumor burden and metastatic disease is typically treated with chemotherapy. CT chest showed innumerable pulmonary nodules. Patient is currently being treated with Doxorubicin and Ifosfamide with minimal response.

References:

1. Perrin RG, Guha A. Malignant peripheral nerve sheath tumors. Neurosurg Clin N Am. 2004 Apr;15(2):203-16. doi: 10.1016/j.nec.2004.02.004. PMID: 15177319.

2. Kim DH, Murovic JA, Tiel RL, Moes G, Kline DG. A series of 397 peripheral neural sheath tumors: 30-year experience at Louisiana State University Health Sciences Center. J Neurosurg. 2005 Feb;102(2):246-55. doi: 10.3171/jns.2005.102.2.0246. PMID: 15739552.
3. Bhattacharyya AK, Perrin R, Guha A. Peripheral nerve tumors: management strategies and molecular insights. J Neurooncol. 2004 Aug-Sep;69(1-3):335-49. doi: 10.1023/b:neon.0000041891.39474.cb. PMID: 15527099.