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Case report

A rare case of metastatic extramammary Paget disease of the spine and review of the literature

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ABSTRACT

Extramammary Paget disease is an intraepithelial neoplasm affecting cells rich in apocrine glands-often located in the vulvar, scrotal, or perianal region. It typically affects older patients, between the ages of 50 and 80 years old, and is most often limited to the epidermis. A 47-year-old Asian male first presented with enlargement of the right inguinal lymph node. A subsequent biopsy revealed extrammamary Paget disease of the scrotum. The patient eventually developed significant worsening back pain with bilateral lower extremity numbness and weakness three months later. Imaging demonstrated a pathologic compression fracture of the L4 vertebral body with metastatic epidural spinal cord compression. The patient underwent surgical decompression of the spine with bilateral L4 laminectomy, resection of epidural tumor, and pedicle screw fixation from L2 to S1. Surgical pathology demonstrated metastatic adenocarcinoma consistent with extramammary Paget disease. Although two other case reports have described spinal metastases from extramammary Paget disease, to the author's knowledge, this represents the first report of surgical decompression and fusion for extramammary Paget disease of the spine.

1. Introduction

Extramammary Paget disease is an intraepithelial neoplasm affecting cells rich in apocrine glands—often located in the vulvar, scrotal, or perianal region. It typically affects older patients, between the ages of 50 and 80 years old, and is most often limited to the epidermis [1]. As a rare, slow-growing malignancy with low metastatic potential, the incidence of metastases is estimated to be 0.11 per 100,000 person-years [2]. Herein, we present a rare case of a patient with metastatic extramammary Paget disease of the spinal column.

2. Case report

A 47-year-old Asian male with a previous history of E-antigen-negative hepatitis B was treated for fungal infections of the scrotum for several years. However, he noticed enlargement of the right inguinal lymph node. A subsequent biopsy revealed extrammamary Paget disease of the scrotum. Histologic sections showed an infiltration of large histiocytic cells with prominent nucleoli, amphophilic cytoplasm, and occasional intracytoplasmic vacuoles (Fig. 1A-B). By immunohistochemistry, these cells were: strongly and diffusely positive for cytokeratin 7 (Fig. 1D), epithelial membrane antigen (EMA, Fig. 1F), and gross cystic disease fluid protein 15 (GCDFP-15, Fig. 1G); focally positive for monoclonal carcinoembryonic antigen (CEA, Fig. 1C) and NKX3.1 (Fig. 1H); and negative for cytokeratin 20 (Fig. 1E), PAX-8 (Fig. 1I), and HER2/neu (not shown). A special stain for mucicarmine highlighted the presence of rare intracytoplasmic mucin vacuoles.

The patient eventually developed significant, and progressive, worsening back pain, bilateral lower extremity numbness, and weakness three months later. Imaging demonstrated a pathologic compression fracture of the L4 vertebral body with metastatic epidural spinal cord compression (Fig. 2A-C).

The patient underwent surgical decompression of the spine with bilateral L4 laminectomy, resection of epidural tumor, and pedicle screw fixation from L2 to S1 (Fig. 2D-F). Post-operative imaging demonstrated spinal cord decompression with instrumented arthrodesis.

Surgical pathology demonstrated metastatic adenocarcinoma consistent with extramammary Paget disease (Fig. 3). Histologic sections showed sheets and nests of epithelioid cells with large

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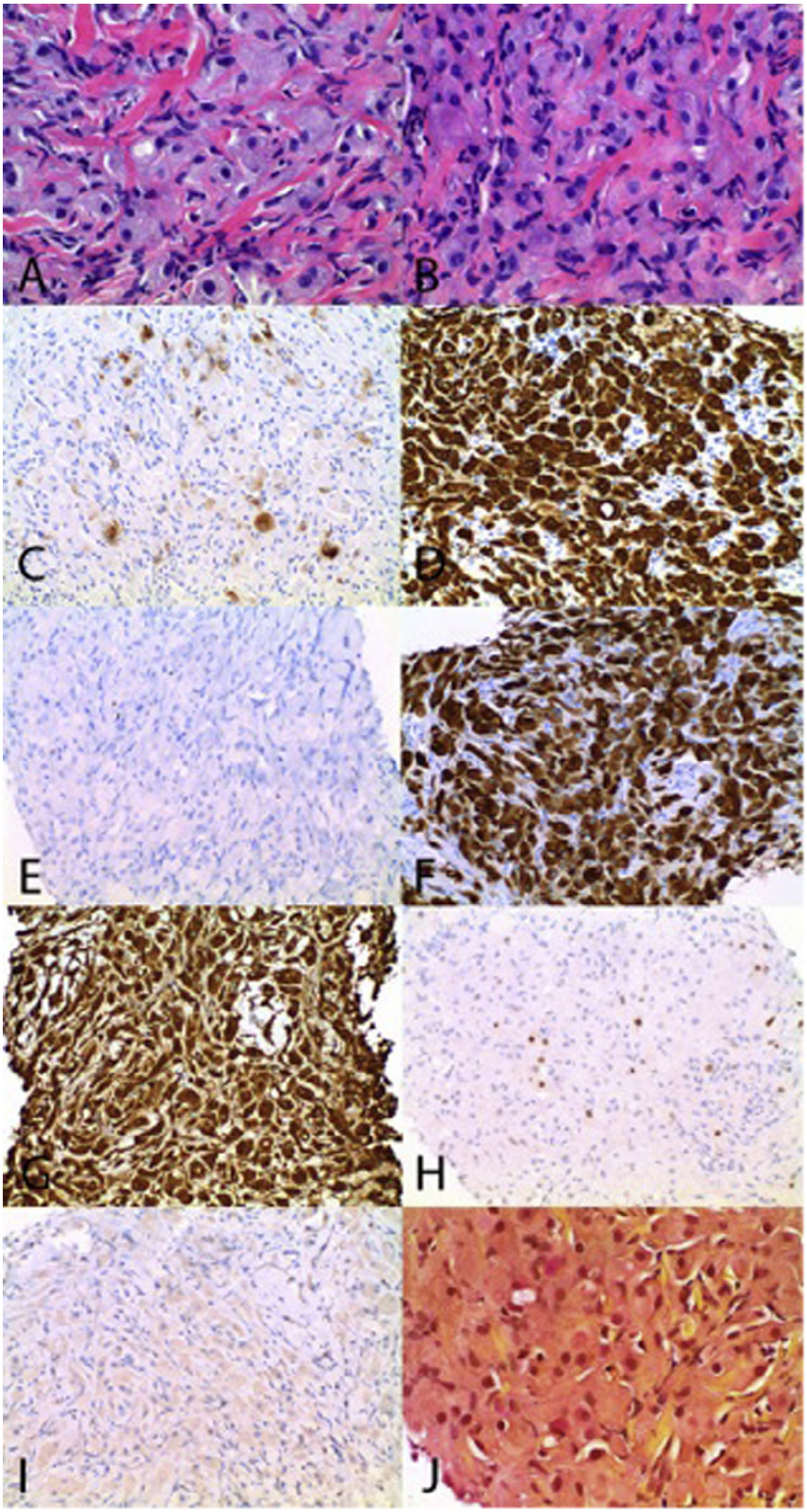

Fig. 1. Immunohistochemistry of right inguinal lymph node metastasis. A-B. Histologic sections showed an infiltration of large histiocytic cells with prominent nucleoli, amphophilic cytoplasm, and occasional intracytoplasmic vacuoles. By immunohistochemistry, these cells were: strongly and diffusely positive for cytokeratin 7 (D), epithelial membrane antigen (EMA, F), and gross cystic disease fluid protein 15 (GCDFP-15, G); focally positive for monoclonal carcinoembryonic antigen (CEA, C) and NKX3.1 (H); and negative for cytokeratin 20 (E), PAX-8 (I), and HER2/neu (not shown). A special stain for mucicarmine highlighted the presence of rare intracytoplasmic mucin vacuoles.

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