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CASE REPORT

Malignant Triton Tumor of the Cervical Spine: Report of One Case and Review of the Literature

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Malignant triton tumor (MTT) is a highly aggressive malignant neoplasm, classified as a variant of malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation. MTT is rarely reported in children, and its true prevalence may be underestimated. We herein report such a case in an 8-year-old boy who presented with a mass over the trapezius muscle. He was previously diagnosed with neurofibromatosis in the same area. Four years later, a follow-up magnetic resonance imaging revealed an intradural tumor recurrence at the level of C1—C7. An immunohistochemical test result was positive for S-100 protein and desmin, which confirmed the diagnosis. The patient outcome was fatal despite multimodal therapy. The possibility of this rare but devastating tumor must always be considered when patients present with new compressive spinal symptoms.

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1. Introduction

Malignant peripheral nerve sheath tumor (MPNST) accounts for approximately 5—10% of all soft-tissue sarcomas. The subgroup of tumors in which malignant Schwann cells coexist with malignant rhabdomyoblasts is termed as malignant triton tumor (MTT).[1] MTT is generally seen in the head, neck, extremities, and trunk.[2] Although an interesting tumor, only limited reports on MTT are available in the neuro-oncology literature. It can occur in sporadic form in the setting of neurofibromatosis type 1 (NF-1).

2. Case report

An 8-year-old boy presented with a progressively enlarging mass in his right trapezius muscle for 6 months. Results of magnetic resonance imaging (MRI) revealed an intramedullary tumor from levels C1—C7 along the continuity of the right upper brachial plexus. Four years before, he was operated for a plexiform neuroma entangled in the right brachial plexus, which was dexterously performed by the cooperation of a neurosurgeon and a plastic surgeon. Upon further questioning, he elicited a past history of NF; the

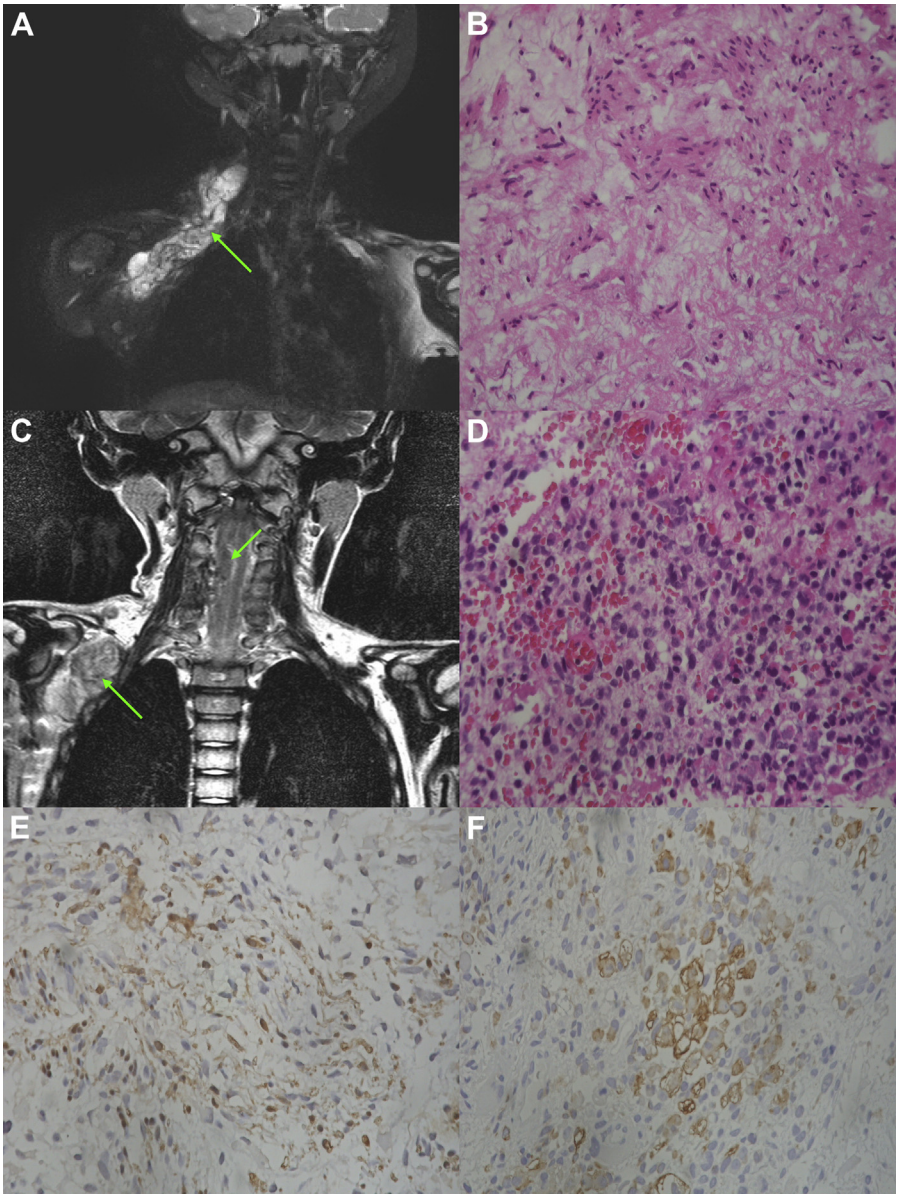

Figure 1 (A) Coronal T2-weighted image with fat saturation demonstrates a characteristic "bag of worms" appearance (arrow) of plexiform neurofibromatosis. (B) Pictomicrograph showing spindle cells with wavy nuclei and cell bodies arranged in a loose myxoid matrix [hematoxylin and eosin (H&E stain; 400×)]. (C) Coronal turbo spin-echo T2-weighted image shows an intradural tumor at the C1—C7 levels, with cord edema extending to the T2 level (arrows). (D) Pictomicrograph showing hypercellular areas characterized by hyperchromatic pleomorphic nuclei and prominent nucleoli (H&E stain; 400×). (E) Pictomicrograph of the tumor showing immunohistochemical positivity for S-100 protein in both elongated and rounded rhabdomyoblastic cells (400×). (F) Immunohistochemical staining revealing marked cytoplasmic positivity for desmin in the areas of rhabdomyoblastic differentiation (400×).