

Case 3

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Clinical Abstract:

This 7 year old boy was in excellent health until 10 months prior to demise. He first developed intermittent nausea, vomiting, and progressive headache followed by double vision and ataxia. CT scan revealed a contrast enhancing mid-line lesion of the posterior fossa. A craniotomy was performed with gross total resection of tumor. Pathological diagnosis was medulloblastoma. Post-operatively the patient had residual cerebellar deficits as well as signs of "aseptic meningitis" with nuchal rigidity. Two weeks after the operation he was referred to the Hospital for further treatment. Evaluation at that time included a myelogram which revealed extramedullary obstruction from T3 to T5 and multiple filling defects from T5 to the sacral sac. The CSF cytology was positive for malignancy. CT scan of the brain was unremarkable with no evidence of recurrent tumor in the cerebellum. He received extensive chemotherapy with cyclophosphamide, intrathecal Ara-C, hydroxyurea, and nitrogen mustard. Subsequently a full course of radiotherapy was given which included 3,600 rads to the neuraxis, with 1200-1500 rad boosts to T1 to T6, the posterior fossa and the cribriform plate. Repeat myelogram revealed resolution of the previously observed filling defects. Three months following the completion of radiotherapy, (four months prior to his demise) cerebellar signs increased and a CT scan revealed contrast enhancement in the meningeal compartment of the cerebellum. A month later a repeat myelogram demonstrated recurrent tumor in the mid-thoracic and cauda equina region. He received further chemotherapy including cis-platinum and PCNU with no improvement. He eventually expired with cardiorespiratory arrest.

On gross examination of the brain no tumor was seen in the region of the previous operation. The major bulk of tumor was in the subarachnoid space of the brain and the spinal cord. Patches and streaks of grey-white firm tissue covered the superior surface of the cerebellum and the base of the cerebrum. Tumor infiltrated the underlying parenchyma in these areas. The brain stem and the spinal cord were extensively encased and superficially infiltrated by similar tumor tissue. The cranial nerves, especially II through VIII, and spinal roots of the mid-thoracic and lumbo-sacral cord were matted by the tumor.

Material Submitted: 1 H & E stained section from autopsy and one unstained.

Points for discussion: 1. Diagnosis
2. Pathogenesis