Malignant intracerebral nerve sheath tumor: A case report

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Case Report

Abstract

Intracerebral MPNST (Malignant Peripheral Nerve Sheath Tumor) is an unusual and a highly malignant tumor. Only 19 cases have been reported so far in the world literature. The prognosis of the tumor is extremely poor. The conventional histopathology and immunohistochemistry is of value to make the exact diagnosis. Of 19 cases which have been reported so far, only 3 cases are above 60 years, whereas ours patient is 65 years and is the 2nd oldest patient.

Keywords: Brain Tumor; Nerve Sheath Tumor; Intracerebral

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a highly malignant tumor that arises from the nerve sheath, either sporadically as a sarcoma or more often from dedifferentiation within a pre-existing neurofibroma in patients with neurofibromatosis 1 (NF1). MPNST that arises from brain parenchyma are termed malignant intracerebral nerve sheath tumors (MINST) and are exceptionally rare with only 19 documented cases in the literature. We hereby report a case of a malignant intracerebral nerve sheath tumor in a 65-year-old female occurring in the frontal lobe.

Case Presentation

A 65-year-old female presented to the neurosurgical out-patient-department with complaints of weakness right side of body which was progressive over a period of 2 months. On examination the patient was conscious and coherent. She had upper motor neuron 7th nerve palsy and power in the right sided limb was 3/5. Right plantar showed an extensor response. Rest of the neurological examination was normal. Her contrast enhanced CT scan head showed a large frontotemporal enhancing lesion with mass effect. Patient underwent craniotomy and excision of the tumor. Intraoperatively the tumor was rubbery yellowish in color, moderately vascular and adherent to the overlying dura. The postoperative period was uneventful. On histopathological examination the tumor comprised of pleomorphic cells arranged in a fascicular pattern with hyperchromatic nuclei and had numerous mitotic figures. Foci of extensive necrosis were also seen. Immunostains were negative for GFAP, EMA, CD117, CD34 and Myo-D. The immunostain for S-100 showed a focal positivity. Patient was subjected to chemoradiation and is doing well on one year follow up.

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Intracerebral MPNST not associated with cranial nerves is an extremely rare and a highly malignant tumor. The over-all survival is poor.

Conflict of interest

The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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