

Intrapontine malignant nerve sheath tumor : MRI and MRS features

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Abstract

The primary source of malignant intracerebral nerve sheath tumors is still unclear.

We report the imaging and MR spectroscopic findings in a 39-year-old man with a very rare brain stem tumor. MR examination revealed the presence of intraaxial brain stem tumor with a partial exophytic growth. On pathological examination, the neoplasm appeared to be an intrapontine nerve sheath tumor, originating most likely from the intrapontine segment of one of the cranial nerve fibres. The tumor showed exophytic growth, with consequent spread to adjacent subarachnoid space. MR spectroscopy revealed the presence of very high concentration of choline, associated with no creatine and N-acetyl aspartate resonance, suggesting non-glial origin of the intraaxial neoplastic infiltration. MR spectroscopy seems to be a helpful diagnostic modality not only in the estimation of the grade of astrocytomas that has been already confirmed in literature, but also in the differentiation between glial and non-glial origin of primary intracranial neoplasms when MR imaging is not conclusive.

Key words : Intracerebral malignant nerve sheath tumor ; MR imaging ; MR spectroscopy.

Introduction

Benign intracerebral nerve sheath tumors are extremely rare lesions, while malignant intracerebral nerve sheath tumors are even less common neoplasms (1-6). It is still debated if malignant intracerebral nerve sheath tumors arise from Schwann cells. No previous reports on MR imaging and MR spectroscopy of intrapontine malignant intracerebral nerve sheath tumor (MINST) are available in literature, most likely due to the rare character of this neurooncologic entity.

Report

Neurologic examination revealed the presence of horizontal nystagmus, mild paresis of the right upper extremity and severe and moderate paresis of the right and left lower extremity, respectively, in a 39-year-old man with one-year long history of the

left facial paresthesia, ataxia and progressive dysarthria. The presence of Romberg sign was noted. Central facial nerve palsy on the right was evident. No diplopia was present.

Brain MR examination revealed the presence of an ill-defined expansile mass lesion infiltrating the left lateral aspect of pons, invading the left middle and inferior cerebellar peduncles, and extending into the ipsilateral subarachnoid space with consequent involvement entry/exit root zones of the trigeminal, facial and vestibulocochlear nerves. The mass was slightly hyperintense on T2-weighted images (Fig. 1) and showed inhomogenous, faint contrast enhancement on T1-weighted images (Fig. 2). The size of the tumor was 30 mm in diagonal antero-posterior and 32 mm in diagonal cranio-caudal dimension. Proton MR spectroscopy (TR = 1500, TE = 135 ms) detected the presence of a prominent peak of choline (Cho) at 3.22 ppm. There was a "shoulder" like appearance at the bottom of Cho peak, which could represent the creatine (Cr) resonance. No presence of N-acetyl aspartate (NAA), lactate and lipid peaks was identified (Fig. 3). Surgical exploration with biopsy was performed and a histologic evaluation showed the features of malignant schwannoma (Fig. 4). The labeling index of proliferation marker Ki-67 was above 20%. Extensive immunoreactivity to S-100 protein was present. No overexpression of p53 marker was noted. Development of the peripheral left facial nerve palsy was evident in the early post-operative course.

Discussion

Various theories regarding the histogenesis of intraaxial schwannomas are considered. According to the majority of authors, MINST is a preferable term to malignant intracerebral schwannoma, since schwannomas rarely undergo malignant alteration (3). While the presence of Schwann cells accompanying the perivascular nerve plexus in the spinal cord is considered as a probable source of the intramedullary schwannoma (7, 8), the occurrence of Schwann cells in the brain remains controversial.

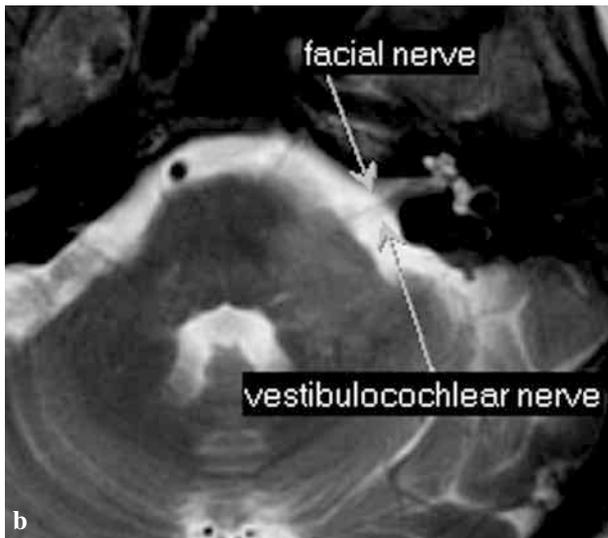
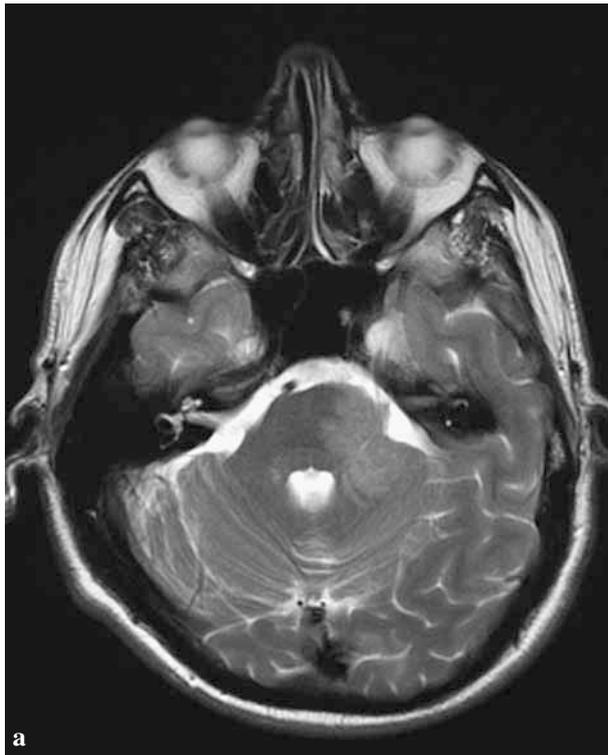


FIG. 1. — Axial T2-weighted image through the brain stem showing a large expansile, slightly hyperintense mass lesion occupying mainly the left side of the pons, extending into the left middle cerebellar peduncle and affecting the root entry zone of the left trigeminal nerve (a) and entry/exit root zone of the left facial and vestibulocochlear nerve (b).

Feigin and Ogata suggested the possibility of Schwann cells arising from the differentiation of the multipotential mesenchymal elements within the brain tissue (9). Nelson and Remels reported that Schwann cells could be found in different pathologic conditions, like multiple sclerosis and old infarcts (10). Disordered embryogenesis was also identified as a theoretical possibility, since younger age groups are predominantly affected (11).

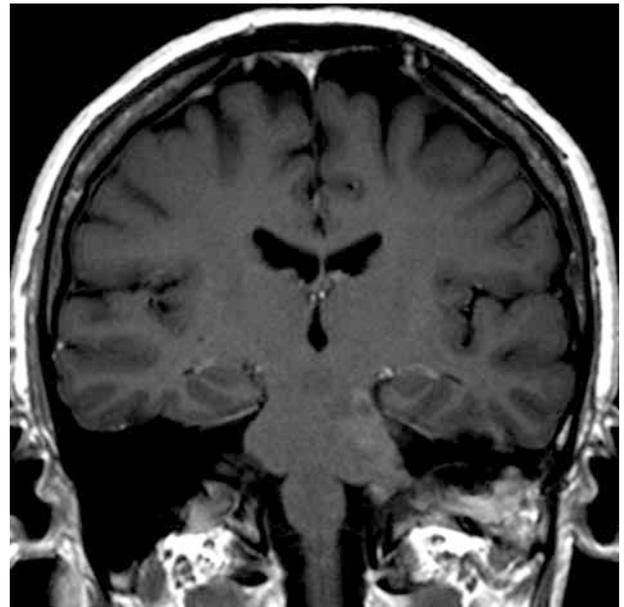


FIG. 2. — Coronal T1-weighted postcontrast image showing inhomogeneous, slight to moderate contrast enhancement of the MINST, affecting the left lateral aspect of pons and showing exophytic growth to the subarachnoid space.

In our patient, the tumor had diffusely infiltrating appearance, invading the pons and showed the involvement of the trigeminal nerve entry root zone and entry/exit root zone of the facial and vestibulocochlear nerve.

In the recent report of Kozic *et al.*, malignant oculomotor nerve sheath tumor in 9-year-old boy involved the cerebral peduncle (area of the parenchymal-cisternal segment of the third cranial nerve) with advanced perineural extension. The patient exhibited pure oculomotor nerve palsy in the initial course of the disease (12). Ueda *et al.* reported the presence of huge, inhomogeneously enhancing anterior cranial fossa mass, histologically compatible with MINST. This tumor most likely originated from the meningeal branch of the trigeminal nerve (1).

The differentiation between benign and malignant extraparenchymal cranial nerve schwannomas on MR imaging is rather difficult except on the basis of rapid growth and aggressive infiltrative characteristics. It has been reported that the T1 and T2 signal intensity characteristics of benign and malignant cranial nerve schwannomas may be indistinguishable (13). With respect to contrast-enhancement, it has been noted that complete absence or minimal, variable postcontrast enhancement might be associated with the facial nerve schwannoma (14). In our patient with MINST, the T2 signal was discretely hyperintense and there was minimal and moderate enhancement on early and delayed postcontrast scans, respectively.

The main spectroscopy findings were the presence of a very prominent Cho peak and the absence of NAA peak. Maheshwari *et al.* reported

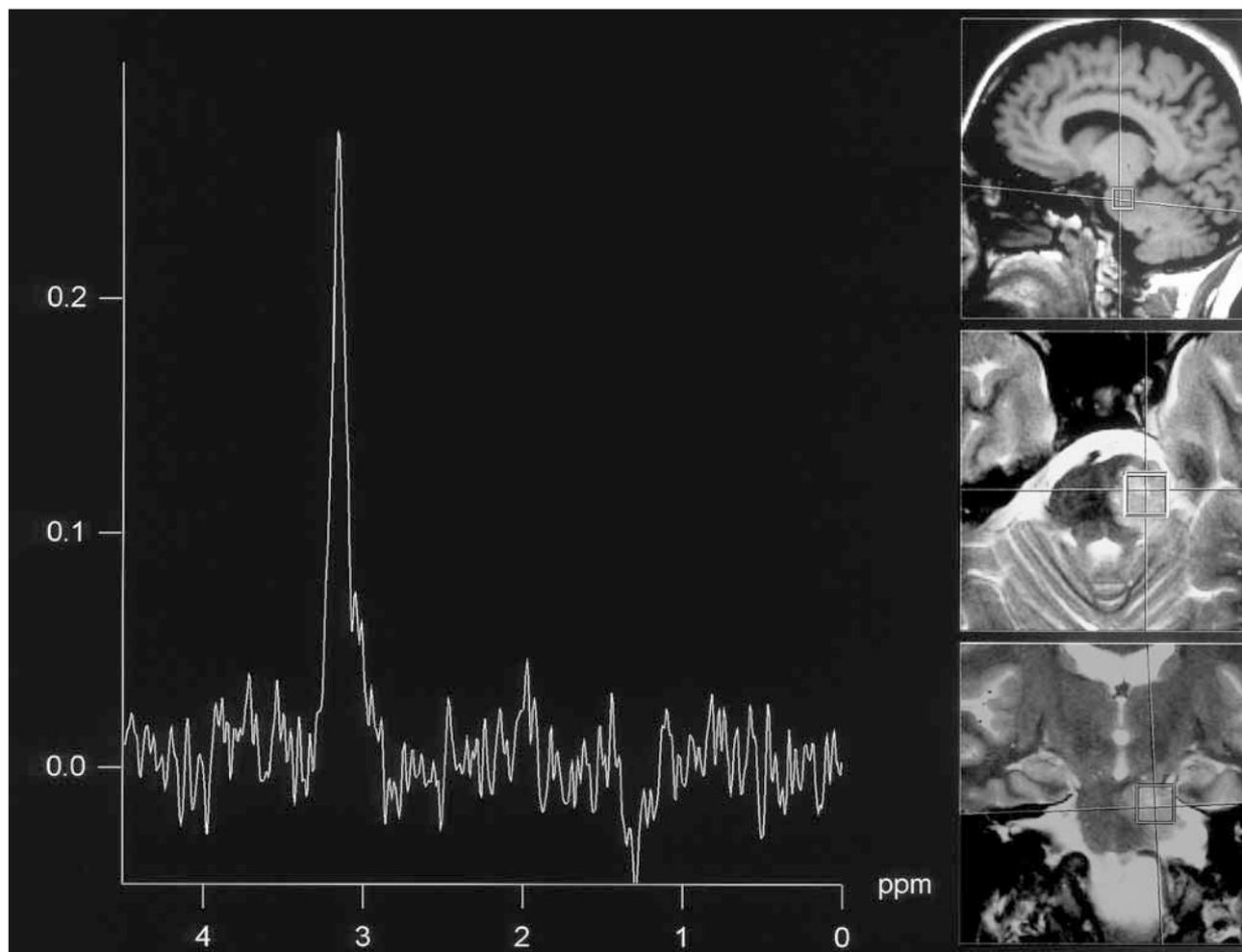


FIG. 3. — Prominent Cho peak with no NAA concentration and with the presence of a “shoulder” resonance at the bottom of Cho peak (most compatible with Cr resonance), suggesting non-gliar origin of the primary brain tumor.

the presence of elevated Cho peak in the vagal schwannoma, associated with an impression of a small “shoulder” peak, most compatible with a minimal creatine resonance (15). A similar impression was noted in our patient, too. Elevated Cho peak may be explained by cellular proliferation and cell density (16). Proliferative capacity of schwannomas is most compatible with their ability to produce abundant amounts of Schwann cells and collagen (12, 15).

The brainstem glioma with exophytic growth was also included in differential diagnosis. We found no presence of lactate and lipid peaks compatible with anaerobic metabolism and cellular necrosis, respectively, usually identified in high-grade gliomas. The alanine peak, typical for meningiomas was also absent. It has been reported that NAA level was reduced to 40-70% of its normal value in astrocytomas, with a trend of more prominent NAA level decrease with the advanced tumor grade (17-20). However Bulakbasi *et al* could not find any statistically significant relation between the decrease in NAA level and the tumor grade (21). Oya *et al.* evaluated the utility of proton

MRS in bone and soft tissue tumors, in order to detect whether or not NAA signal could be recognized at 2.0-2.1 ppm. The signal was not found in neurofibroma (9/9), schwannoma (6/6), pheochromocytoma (2/2) and other mesenchymal tumors of non-neuroectodermal origin (22). Neuronal marker NAA tends to decrease in malignant intraaxial tumors due to neuronal destruction, however complete absence of NAA seen in our patient, in association with other imaging and spectroscopy data, suggested the non-gliar origin of the neoplasm. Also, even in high grade gliomas, both Cho and Cr peaks are usually well defined and separated while in extraaxial tumors the Cr peak is either completely not detected or presents with a “shoulder” like appearance in the bottom of the Cho peak (15).

Information obtained from MR spectroscopy, consistent with a rather typical “extraaxial neoplasm pattern” was quite reliable indicator that helped in the differentiation between glioma and non-gliar nature of the unusual malignant cranial tumor. In the investigation of Moller-Hartman *et al.* the additive information from MR spectroscopy led to a 15.4% higher number of correct diagnoses and

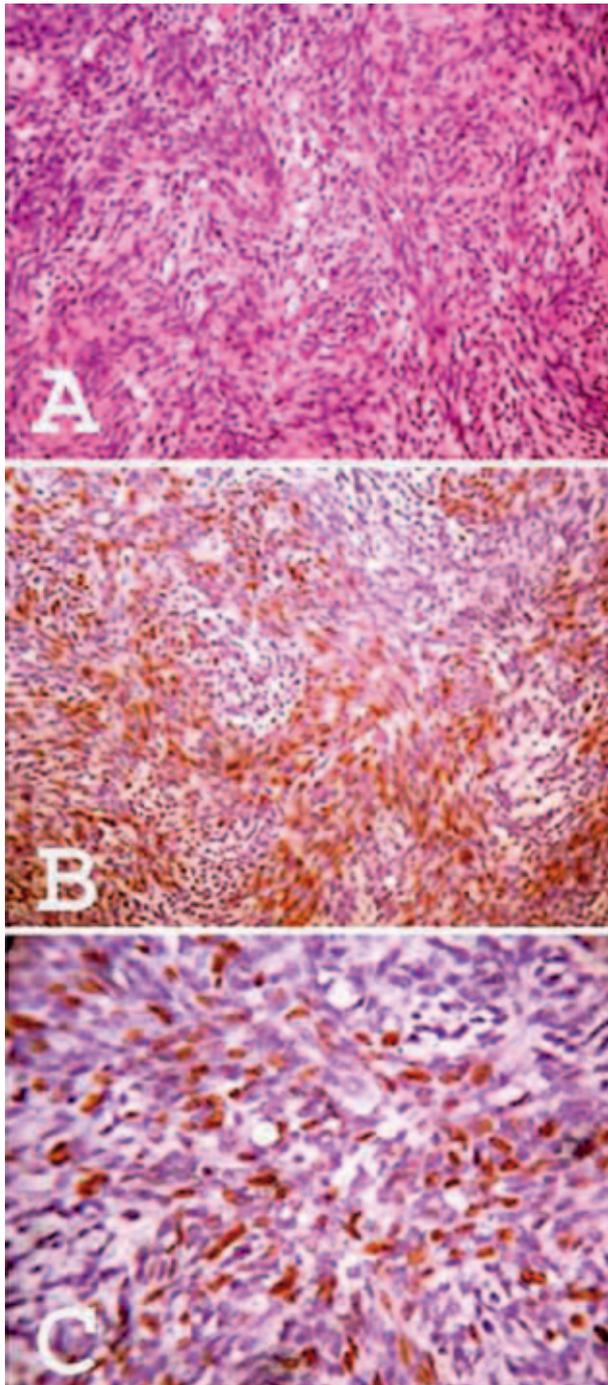


FIG. 4. — Malignant nerve sheath tumor. (A) The majority of tumor cells is tightly packed and arranged in fascicles (B) S-100 immunopositivity present in tumor cells. (C) High Ki-67 labeling index corresponding to infiltrative potential of the tumor.

to a 6.2% lower number of incorrect compared to MR imaging alone, while 16% fewer equivocal diagnoses were also observed (19). In the study that included 164 patients, neither MRI nor MRS alone provided a degree of accuracy sufficient for reliable preoperative differential diagnosis of brain tumors. However, by combining these two modalities, a high level of diagnostic accuracy was achieved (23).

In conclusion, we would like to emphasize that MR spectroscopy seems to be helpful diagnostic modality not only in the estimation of the grade of astrocytomas (increased Cho/Cr ratio and decreased NAA) that has been already confirmed in literature, but also in the differentiation between glial and non-glial origin of primary intracranial neoplasms, like in our patient in whom the aspect of malignant nerve sheath tumor was in complete contradiction with the usual findings of an extraaxial cerebello-pontine angle schwannoma and resembled brain stem glioma. This even more stresses the importance of MR spectroscopy.

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