Synchronous Tricompartmental Benign CNS Tumors with Tonsillar Herniation, Cervicodorsal Syringomyelia and Hydrocephalus

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Abstract In this study, three primary central nervous system tumors of different histological cell types occurring together without neurofibromatosis are reported. These included a sellar pituitary adenoma with apoplexy, a large torculo-tentorial meningioma in the posterior fossa, and a cervical spinal intramedullary schwannoma. Displacement of primitive multipotent cells in different central nervous system compartments or the oncogenic effects of activated signaling of growth factor receptors are the proposed pathophysiological mechanisms for the simultaneous genesis of different types of tumors. There was associated tonsillar herniation, cervicodorsal syringomyelia, and hydrocephalus. The radiological features and treatment strategy of these rare synchronous tumors are highlighted.

Keywords Synchronous tumors · Central nervous system tumors · Tentorial meningioma · Simultaneously occurring tumors · Intramedullary schwannoma · Suboccipital craniectomy

Introduction

Multiple primary central nervous system (CNS) tumors of different histological cell types occurring simultaneously are rare. A 55-year-old male patient presented with ataxia and lower cranial nerve impairment for 3 years and symptoms of raised intracranial pressure for 6 months. His contrast-

enhanced magnetic resonance imaging revealed synchronous benign tumors in three separate CNS compartments: a predominantly posterior fossa-based peritortorial tentorial meningioma (TM) invading into the supratentorial compartment and the straight sinus reaching the posterior third ventricular region confirming to Yasargil's primary type IV (infra/supratentorial posteromedial) TM [1]; a pituitary chromophobe adenoma with apoplexy (with subacute hemorrhage); and a cervical intramedullary cystic schwannoma [Fig. 1]. The large posterior fossa meningioma with raised ICP caused tonsillar herniation (up to C2 lamina), alteration of cerebrospinal fluid dynamics at foramen magnum, leading to cervicodorsal syringomyelia (extending from C2-T2 levels) and hydrocephalus. The possibility that the tonsillar descent represented a previously existing congenital Chiari I malformation also existed. Displacement of primitive multipotent cells in the different CNS compartments [2] or the oncogenic effects of activated signaling of growth factor receptors [3] are the proposed pathophysiological mechanisms for the simultaneous genesis of different types of tumors. An appropriately staged surgical strategy was as follows: a) CSF diversion; b) transsphenoidal pituitary adenoma excision; and c) a suboccipital craniectomy, C1 posterior arch-C2 lamina removal with simultaneous torculo-TM excision (sparing a rim of tumor tissue invading the straight sinus and surrounding the major veins of the posterior third ventricular region subsequently addressed by stereotactic radiosurgery), cervical posterior midline myelotomy with schwannoma excision, and a duroplasty to address the tonsillar herniation. Despite having a synchronous CNS meningioma and schwannoma, this patient does not strictly confirm to the National Institutes of Health criteria of neurofibromatosis-2 (NF-2) due to the absence of a first-degree relative with NF-2 as well as the lack of other stigmata associated with the phacomatoses [4].

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Fig. 1 **a** and **b** Contrast enhanced axial, **c** sagittal, **d** coronal T1-MRI showing a posterior fossa peritortulo-tentorial meningioma invading into supratentorial compartment, straight sinus and posterior third ventricular region with hydrocephalus, and a pituitary adenoma with

subacute hemorrhage, **e** T1, and **f** T2-MRI showing a cervical intra-medullary cystic schwannoma, tonsillar herniation, and cervicodorsal syringomyelia

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