Intracranial solitary fibrous tumor/hemangiopericytoma: Report of two cases and literature review

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ABSTRACT

Intracranial solitary fibrous tumors (ISFTs) are rare mesenchymal neoplasms originating in the meninges and constitute a heterogeneous group of rare spindle cell tumors that include benign and malignant neoplasms of which hemangiopericytoma is nowadays considered a cellular phenotypic variant. ISFT usually shows benign or indolent clinical behavior. We describe two cases of ISFT managed in our institution along with a review of pertinent literature.

Keywords: Solitary fibrous tumor, hemangiopericytoma, tumor, meningioma

Introduction

Solitary fibrous tumors (SFTs) affect mainly the visceral pleura. They were first described as a primary spindle cell tumor of the pleura by Klemperer and Rabin in 1931.1 Primary SFT involving the central nervous system (CNS) was first reported in 1996 by Carneiro et al.2 Involvement of the CNS is rare and has been attributed to the paucity of true connective tissue elements. These tumors were considered to have benign histopathological features at onset although anaplastic or malignant transformation resulting from multiple recurrences has also been reported few times.3 It might involve the brain, nerve roots as well as the skull base.4

Case Report

The first patient was a 59-year-old man, referred by a dermatologist to our outpatient clinic with a 6-month history of a progressively growing mass situated medially on the head convexity: otherwise, the patient was asymptomatic. A cerebral magnetic resonance imaging (MRI) showed a parasagittal postcentral left-sided mass enhancing homogeneously, eroding the skull, extending intra- and extra-cranially, and compressing/invading the superior sagittal sinus (Figure 1a). At the latest follow-up (2 years later), neither residual nor recurrence of the tumor (Figure 1c) was found and the patient doing very well. The second patient was a 54-year-old man complaining 2-year history of progressive unsteady walk, headache, and vertigo.

An enhanced brain computed tomography and MRI scan showed a space occupying lesion involving the free edge of the tentorium, compressing the 4th ventricle, without hydrocephalus, in favor of a meningioma (Figure 1b). The

Figure 1: Magnetic resonance imaging (MRI) of intracranial solitary fibrous tumor, (a) axial pre-operative MRI of the first patient, (b) axial pre-operative MRI of the second patient, (c) axial post-operative MRI of the first patient, (d) axial post-operative MRI of the second patient
patient underwent surgery, and a gross total resection was achieved; the histopathology showed to be an ISFT. At the 4th year follow-up, a MRI scan showed neither residual nor recurrence of the tumor (Figure 1d) and the patient reporting a generalized well-being.

Discussion

There has been debate over the histogenesis of the SFT as to whether the origin is a mesothelial or mesenchymal, but recent immunohistochemical and electron microscopic studies have suggested that they originate from mesenchymal fibroblast-like cells. The main differential diagnosis of ISFT includes fibrous meningioma; it may also mimic schwannoma as a cerebellopontine angle tumor. Symptoms are mostly related to tumor location, which is similar to that of meningioma. These tumors usually occur in the fifth decade and are equally represented in men and women. Incomplete surgical excision is one of the important predictive factors of recurrence. Our cases were completely excised without evidence of recurrence at 4 and 2 years, respectively; the former is another demonstration that surgery represents the gold standard management of such a tumor.

Conclusion

For its rarity and resemblance to other more common brain tumors, ISFT is often poorly recognized and remains a diagnostic challenge. The correct diagnosis of SFT could be made only by histopathology.

References