Meningeal fibroma: a rare meningioma mimic

Report of 2 cases

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Meningeal fibromas are rare intracranial tumors that mimic meningiomas radiologically as well as histologically. The authors report 2 cases of meningeal fibroma with detailed clinical, radiological, histopathological, and immunohistochemical features, and discuss the differential diagnosis of this entity. Knowledge of this rare tumor is essential for pathologists to be able to distinguish it from more common meningeal tumors, especially in younger patients. This knowledge is also essential for neurosurgeons, as incomplete resection may lead to tumor recurrence, and such patients require close follow-up.

Case Reports

Case 1. A 15-year-old boy complained of intermittent headache and vomiting for 3 months. His parents had noted changes in his personality and behavior over this period. On examination, there was no evidence of any sensorimotor deficits or any stigmata of neurofibromatosis. Magnetic resonance imaging revealed a large, heterogeneously enhancing mass lesion in the frontotemporo-parietal region, suggestive of a meningioma (Fig. 1). The patient underwent a right frontoparietal craniotomy and gross-total excision of the tumor. Intraoperatively, a grayish-white, hard, extraaxial mass with minimal vascularity was noted, attached to the falx. The tumor was excised completely, along with its falcine attachment (Simpson Grade I excision) and was submitted for histopathological evaluation. The patient remains recurrence-free 3 years after the surgery.

Case 2. An 8-year-old boy presented with a history of intermittent headache and abnormal behavior, includ-
Ing hyperactivity and nonsensical speech, for 1 year. He had undergone operations twice at another hospital, after which he had remained symptomatic postoperatively. On both the previous occasions, a histopathological diagnosis of WHO Grade I fibroblastic meningioma had been made. Preoperative MRI of his head showed a well-defined falx region mass that was hypointense on T2-weighted images and showed peripheral dense enhancement on T1-weighted images with contrast administration. There was no evidence of calcification. Repeat imaging revealed a residual tumor (Fig. 2). The tumor was approached through a bifrontal craniotomy. Intraoperatively, a well-demarcated, firm, lobulated, poorly vascularized tumor was noted attached to the middle third of the falx cerebri, which was reaching up to the base of the falx, more on the right side. Simpson Grade II tumor excision was achieved, and the operative specimen was submitted for histopathological examination. Sections from the primary tumor stained with H & E were also reviewed. The patient was followed up for a period of 1 year, during which time no further recurrence or metastasis occurred.

Pathological Examination. Tumor tissue was fixed in 10% neutral-buffered formalin, routinely processed, and paraffin embedded. Five-micron-thick sections were cut for routine H & E staining and immunohistochemical analysis. A labeled streptavidin biotin kit (Universal, Dako) was used as a detection system. Antigen retrieval was performed in a microwave oven using citrate buffer at pH 6.0 for all antibodies. Monoclonal antibodies against vimentin (Diagnostic BioSystems, 1:100), epithelial membrane antigen (EMA; Dako, 1:100), pan cytokeratin (Neomarkers, 1:200), claudin-1 (Neomarkers, 1:50), smooth muscle actin (SMA; Dako, 1:50), progesterone receptors (Neomarkers, 1:50), estrogen receptors (Neomarkers, 1:50), CD34 (Dako, 1:100), bcl-2 (Neomarkers, 1:100), CD99 (Dako, 1:100), desmin (Dako, 1:50), glial fibrillary acidic protein (GFAP; Dako, 1:1500), beta catenin (BD Transduction, 1:200), p53 protein (Santa Cruz Biotechnology, 1:1000), and proliferation marker MIB-1 (Dako, 1:200) were used. For assessing the MIB-1 labeling index, at least 500 cells were counted using a 1-square-mm eyepiece pinhole. For each batch, appropriate positive and negative controls were taken. Electron microscopic examination was performed on paraffin-embedded tumor tissue, after deparaffinization, rehydration, and postfixation in osmium tetroxide.

Microscopic Examination. Sections stained with H & E from tumors from both patients showed similar features (Fig. 3). The tumors were sparsely cellular, composed of elongated spindled-shaped cells with a scant to moderate amount of pale eosinophilic cytoplasm, and normochromatic oval to elongated nuclei with delicate chromatin and inconspicuous nucleoli. The tumor cells were arranged in long parallel fascicles that were embedded in a densely collagenized matrix. Areas of hyalinization were present and were more prominent around blood vessels. Sections from both cases showed focal myxoid change in the stroma. The tumor cells did not demonstrate nuclear atypia, pleomorphism, mitoses, or areas of necrosis. There was no evidence of meningothelial differentiation in the form of whorl formation or sheeting of the tumor cells. Mast cells were observed interspersed between the tumor cells. Review of sections prepared from tumors excised at the first and second surgeries in Case 2 showed similar histomorphological features.

The tumor cells were immunopositive for vimentin;
however, they were immunonegative for EMA, S100 protein, claudin-1, estrogen receptors, progesterone receptors, CD34, CD99, bcl-2, beta catenin, desmin, SMA, pan cytokeratin, and GFAP (Fig. 4). The MIB-1 labeling index was less than 1% in both tumors, and no immunopositivity for p53 protein was noted. Ultrastructural examination (Fig. 5) of paraffin-embedded tumor tissue from both cases showed the presence of artifacts; however, the features were sufficient to recognize the tumor cells as fibroblasts. Spindle-shaped tumor cells with elongated nuclei (F), perivascular hyalinization (G), and interspersed mast cells (H).

Fig. 3. Photomicrographs showing tumor cells in long intersecting fascicles (A and B) with areas of hyalinization (C), focal myxoid change (D), and thick bundles of collagen in the stroma (E). High magnification shows bland spindle-shaped cells with elongated nuclei (F), perivascular hyalinization (G), and interspersed mast cells (H). H&E, original magnification ×40 (A), ×100 (B and C), ×200 (D and E), and ×400 (F–H).
Long nuclei, some of which appeared indented, and abundant dilated rough endoplasmic reticulum were identified. Some of the cells showed the presence of elongated processes. Bundles of collagen fibers were present in the extracellular matrix. Intercellular junctions and interdigitations were not observed, excluding the possibility of a meningo-epithelial origin.

Discussion

Intracranial fibromas are rare, benign, fibrous lesions that arise from the meninges or from within the brain parenchyma. Less than 20 such cases have been described to date, and their incidence remains unknown. At our institute, they accounted for 0.12% (2/1648) of all meningeal tumors excised over a 10-year period. Intracranial fibroma was first described in the English literature by Koos et al. in 1971 in an 11-month-old boy. These fibromas are usually noted in the first 2 decades of life, and show a male preponderance. These tumors are usually bulky, well-circumscribed masses that, on histological examination, are found to be composed of elongated spindle cells in a dense, eosinophilic, hyalinized collagenous matrix. Due to the rarity of these tumors, no definite management protocol is defined for meningeal fibromas. Because these tumors are benign lesions, complete resection is curative. However, incomplete resection can lead to recurrence or regrowth of the tumor, as noted in one of our cases.

The differential diagnoses of meningeal fibroma include meningioma, SFT, fibromatosis, cranial fascitis, meningeal myxoma, and meningeal glioma (Table 1). While psammomatous calcification and osseous and chondroid metaplasia may be encountered in these tumors, the absence of whorls and meningo-epithelial features helps to distinguish them from fibroblastic meningiomas. Immunoreactivity with EMA, claudin-1, S100 protein, and progesterone receptors in meningiomas further aids in this distinction. Another close differential diagnosis is SFT. These tumors also arise from the meninges, but are usually observed in adults. Solitary fibrous tumors are more cellular than fibromas, and are composed of interfacing fascicles of spindle cells rather than long parallel fascicles. They are immunopositive for CD34, CD99 and bc1-2, while fibromas are negative for these markers. Meningeal gliomas are extremely rare; the absence of collagen accompanied by immunopositivity for GFAP helps to differentiate between the two tumors. Fibromatosis is a histologically benign but locally invasive condition involving the dura, but it has infiltrative margins, whereas fibromas are well circumscribed. Fibromatosis may occur de novo or at the site of previous surgery or trauma. Intracranial myxomas are rare lesions that arise from the skull bones and may show attachment to the dura. Radiologically, they are usually limited to the inner and outer tables of the skull, while histologically, they are typified by a loose myxoid matrix containing spindled-to-stellate-shaped cells with minimal pleomorphism. Cranial fascitis is a reactive proliferation of fibroblastic and myofibroblastic cells in a variably hyalinized and myxoid stroma, which involves the soft tissue of the scalp and adjacent cranium, and may extend to involve the dura. Its predilection for the first 6 years of life, presence of a lytic skull lesion, prominent myxoid matrix, and inflammatory cell infiltrate on histology help to distinguish it from meningeal fibroma.

Nonmeningeal tumors of the meninges, including fibromas, are much less common than meningiomas. Knowledge of these entities is essential for pathologists to include them in the differential diagnosis of meningiomas, especially in younger patients; this knowledge is also essential for neurosurgeons because incomplete resection may lead to tumor recurrence, and these patients need to be closely followed up.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.
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References


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