CASE REPORT

Intracerebral fibroma: a case report and review of the literature

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Abstract

Intracerebral fibromas are among the most rare neoplasms found in the central nervous system. Ten previously reported cases have been documented in the literature including only two reported cases since 1985. As a result, little is known about these uncommon intracerebral fibrous tumors. We report a case of an intracerebral fibroma without dural or leptomeningeal attachment, discuss the pertinent diagnostic findings and briefly review all prior reports of this entity.

INTRODUCTION

We report a case of intracerebral fibroma, an exceedingly rare neoplasm found in the central nervous system. Since 1985, only two cases have been reported.

CASE REPORT

A 41-year-old woman with no known history of seizures presented to the emergency department after a witnessed fall from standing. The patient had transient loss of consciousness, suffered a seizure and experienced post-ictal amnesia. The patient reported a history of falls, occurring approximately once per month over the past 2–3 years. She denied headache, visual disturbance, gait imbalance, nausea or vomiting. The complete neurological examination was within normal limits. She was in generally good health with no major medical problems. She had no pertinent surgical history.

Diagnostic computed tomography (CT) and magnetic resonance imaging (MRI) studies of the brain revealed a right-sided temporoparietal 2.5 × 2.0 cm high-attenuation lesion with associated vasogenic edema and heterogeneous enhancement after gadolinium administration (Fig. 1). CT body imaging also indicated a 5-mm pulmonary nodule within the left upper lobe and a pelvic soft tissue irregularity with fluid in the region of the uterus. Subsequent gynecologic workup revealed this to be a simple cyst; however, given the body imaging profile, our differential diagnosis included meningioma or metastatic neoplasm.

We performed a right-sided craniotomy for resection of the solitary intracranial mass. The surgery was performed with the aid of the BrainLAB frameless stereotactic neuronavigation system. Open exploration revealed no dural attachments. The mass appeared extra-axial without invasion into the brain (Fig. 2). In addition, it was hard and mineralized. There was a pial interface between the lesion and the brain, allowing for gross total resection of the tumor and its capsule.

Frozen section pathology indicated a spindle-cell tumor. The final pathology report reflected intracerebral fibroma, which was reviewed both at our institution and confirmed at Johns Hopkins Reference Laboratories. The specimen had no meningeal cell nests attached to it. There were no whorls, psammoma bodies, meningothelial cells or other stigmata of meningioma. It did not stain for CD34 or BCL-2. Immunohistochemistry for EMA, S-100 and SMA were non-contributory (Figs 3 and 4).

The patient had an uneventful postoperative hospital course and was subsequently discharged to home on Postoperative Day 6. She was seen 2 weeks later in clinic, and the...
neurological examination revealed no deficits. Furthermore, she has remained seizure-free since the operation.

**DISCUSSION**

Intracerebral fibroma without evidence of dural attachment represents an extremely rare entity. An electronic search of the literature dating back to 1950, using the keywords ‘intracerebral fibroma’, yields only 10 previously reported cases [1–7], with only 2 cases reported since 1985 [8, 9]. Of the 10 prior cases, all but one were located supratentorially [5]. Similar to the tumor in our patient, most of these early described tumors reported a hard rubbery intracerebral mass unrelated to the meninges. In only two cases did the authors report an attachment to the falx or dura [6, 7]. Previously reported specimens indicate a tumor composed of elongated cells with the features of fibroblasts, embedded in abundant collagen bundles [7]; however, immunohistochemical staining was not described. A summary of prior reported specimens is demonstrated in Table 1.

A closely related intracranial finding is that of solitary fibrous tumor (SFT) of the meninges. SFTs account for just 0.09% of all meningeal tumors [10]. The ultrastructural findings of SFTs are similar to intracranial fibromas, in that they have spindle-cell proliferation with areas of collagenization [10]. The main difference is found in the immunohistochemical staining and site of dural attachment. Recent immunohistochemical and ultrastructural studies indicate that these tumors arise from mesenchymal fibroblast-like cells in the meningeal covering, and are classified as mesenchymal, non-meningothelial tumors [10], which is an important differential consideration. The immunohistochemical profile of SFTs includes strong reactivity for CD34 and vimentin but not EMA or S-100 [10]. CD34 is a transmembrane glycoprotein, which is expressed by hematopoietic progenitor cells, endothelium and certain populations of mesenchymal stromal cells in the dermis and, by all accounts, is the most sensitive marker for SFTs. Intracerebral fibromas do not stain for CD34 [10], which is a clear difference in differentiating intracerebral fibroma and SFT.

Currently, MRI appears to be the modality of choice for initial detection of these tumors. However, there is no clear imaging differentiation between intracerebral fibroma and other fibrous tumors found within the intracranial cavity such as SFT, meningioma or schwannoma. This increases the importance of adequate histological and immunohistochemical evaluation of these lesions in order to arrive at the appropriate tumor diagnosis.

Ultimately, the diagnosis of intracerebral fibroma was based on the tumor being unassociated with the dura, the histological evaluation of spindle-shaped cells interspersed with a dense collagen matrix and the immunohistochemical profile. Intracerebral fibromas are indeed very rare in the central nervous system. Few cases have been described, and little is known regarding the pathogenesis and diagnosis of these lesions. The diagnosis depends greatly on immunohistochemical and histologic correlation, and complete surgical resection...
remains the goal of treatment. Further reports are needed to refine the true incidence of intracerebral fibromas and to provide a more clear definition of these lesions.

**CONFLICT OF INTEREST STATEMENT**

None declared.

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| Case number | Age     | Sex   | Location                          | Pathologic specimen | Size          | CD expression |
|-------------|---------|-------|-----------------------------------|---------------------|---------------|---------------|
| 1           | 11 months | Male  | Supratentorial, right frontal lobe | Total resection     | 5 × 5 × 5 cm  | NA            |
| 2           | 9 years  | Female | Supratentorial, left temporoparietal lobe | Open biopsy, subtotal resection | 3 × 3 × 3 cm  | CD34 negative |
| 3           | 10 years | Female | Supratentorial, right frontal lobe | Observed upon autopsy | 5 × 5 × 5 cm  | NA            |
| 4           | 11 years | Female | Supratentorial, right lateral ventricle | Total resection     | 3 cm          | NA            |
| 5           | 17 years | Male   | Supratentorial, right parietal lobe | Total resection     | NA            | NA            |
| 6           | 19 years | Female | Supratentorial, right temporoparietal lobe | Total resection     | NA            | NA            |
| 7           | 19 years | Male   | Infratentorial, left cerebellar hemisphere | Total resection     | 5 × 4 × 4 cm  | NA            |
| 8           | 24 years | Female | Supratentorial, left frontotemporoparietal lobe | Observed upon autopsy | NA            | NA            |
| 9           | 42 years | NA     | Supratentorial, temporoparietal lobe | Total resection     | NA            | NA            |
| 10          | 52 years | Male   | Supratentorial, right parietal lobe | Total resection     | 5 × 2.5 × 5 cm | NA            |