Meningioangiomatosis of the brain stem

Case report

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The case is reported of meningioangiomatosis of the brain stem in a 31-year-old girl who suffered from vomiting, left facial weakness, difficulty in swallowing, and ataxia. This is believed to be the first reported case of meningioangiomatosis in the brain stem. Computerized tomography showed an intensely enhancing hyperdense mass in the left restiform body. Magnetic resonance imaging revealed that the lesion was isointense to gray matter on the T1-weighted image and hypointense on the T2-weighted image, with a surrounding zone of high T2 signal and intense enhancement. Angiography was normal. Surgical exploration demonstrated an intramedullary firm mass that was partially resected. Histologically, the mass consisted of a low-grade lesion of meningeal origin with spindle cells in a whorling pattern that were occasionally focused around small vessels. On 2-year follow-up imaging, the lesion remains unchanged in size. Certain particularities of this lesion are discussed in the context of the literature.

KEY WORDS • meningioangiomatosis • brain stem • histopathology

Meningioangiomatosis is a rare but benign process, probably of hamartomatous origin, characterized histologically by proliferation of meningeal cells along proliferating parenchymatous blood vessels.15 It was initially described as a forme fruste of neurofibromatosis,19 but recently increasing numbers of isolated cases have been described.1,4,5,7,10,11,14,16-18 Classically, the process is localized in the cerebral cortex and also involves the overlying leptomeninges.5,7,13,15 Brain stem involvement has not previously been described. We report a case of meningioangiomatosis located in the posterior medulla and describe the histological characteristics of the lesion as well as the clinical and therapeutic implications of its unusual location.

Case Report

This 31-year-old girl, previously healthy with an uneventful prenatal course, presented with a 2-week history of vomiting and left facial weakness.

Examination. Physical examination revealed an alert, irritable child with marked left-sided facial droop and prominent fasciculations of her tongue, which she protruded with difficulty. During hospitalization her symptoms progressed, including difficulty in swallowing and occasional ataxia. Laboratory evaluation of blood and cerebrospinal fluid revealed no abnormalities. She had no stigmata or family history of neurofibromatosis.

Pre- and postcontrast computerized tomography (CT) of the head disclosed a well-defined, slightly hyperdense mass in the left restiform body, which enhanced intensely (Fig. 1). Magnetic resonance (MR) imaging before and after gadolinium administration showed a well-circumscribed round mass 1.5 cm in diameter. Although predominantly intra-axial, the possibility of an extra-axial mass invading the medulla could not be excluded. The mass was isointense to gray matter on T1-weighted MR images and slightly hypointense on T2-weighted images, surrounded by a zone of hyperintense T2 signal. The lesion enhanced intensely on MR imaging after contrast administration (Fig. 2). Cerebral angiography was normal.

After a trial of antibiotics and steroids, the patient had transient improvement of symptoms despite a persistent left facial palsy and intermittent ataxia. A repeat MR image 3 weeks later showed no change in the lesion's size or signal characteristics.

Operations. A posterior fossa craniotomy was performed. The inferior vermis was divided, exploring the floor of the fourth ventricle. Intraoperative ultrasound studies demonstrated the intra-axial location of the lesion. On biopsy, the lesion was very firm. A preliminary
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**FIG. 1.** Preoperative computerized tomography scans. **Left:** Axial scan showing a slightly hyperdense mass in the left restiform body (arrows). **Right:** Contrast-enhanced axial scan showing intense enhancement of the mass (arrow). The mass is surrounded by an area of low attenuation likely representing edema.

Histology report was consistent with a fibroblastic process, probably reactive rather than neoplastic.

The patient was discharged with improvement and was maintained on a course of steroids. Two months later she was readmitted with difficulty in swallowing and tremors of the head and trunk. Repeat MR imaging showed the lesion had enlarged slightly but there was no change in signal characteristics. Repeat biopsy and surgical resection were performed. An area devoid of vascular markings was present in the region of the left restiform body. After incision, a firm lesion was identified appearing almost fibrocytic and like a meningioma. At least 50% of the lesion was resected. Because dissection at the lesion's margins resulted in significant bleeding, no further dissection was performed.

**Pathological Examination.** Histologically, the lesion consisted of interlacing and whorled fascicles of spindle cells with fibrillary cell processes and abundant collagen matrix (Fig. 3). Mitosis and nuclear pleomorphism were absent, and psammoma bodies were not seen. Focal capillary vascularity was prominent. Electron microscopy showed cells containing abundant cytoplasmic intermediate filaments, prominent rough endoplasmic reticulum, minimal external lamina, and a few primitive cell junctions. Immunohistochemical studies revealed that most cells were reactive with antibodies to cytokeratin (CAM-5.2 and MAK-6) and, in an irregular pattern, to antibodies against vimentin, Leu-7, and neuron-specific enolase. Few cells were immunoreactive for S-100 protein. Tests for glial fibrillary acidic protein, neurofilament, and epithelial membrane antigen were negative. Despite this lesion's histological similarities with meningioma (especially of the meningothelial type) its benign histological features, absence of pleomorphism, lack of leptomeningeal tumor, and intra-axial location argue against this possibility. The lesion was classified within the spectrum of meningoangiomatosis, implying a hamartomatous rather than a neoplastic origin. The term “meningiomatosis” was also suggested because the meningeal reaction predominated to vascular proliferation.

**Postoperative Course.** Two years after the second operation, the patient uses a walker, and has minimal facial asymmetry and ataxia. Follow-up MR examinations showed no changes in the postoperative appearance of the lesion.

**Discussion**

Meningioangiomatosis is an uncommon benign lesion of the central nervous system, presumably of malformative origin, that predominantly involves the cerebral cortex and often the overlying leptomeninges. The condition was first described in 1915 by Bassoe and Nuzum as an incidental autopsy finding in a 15-year-old boy with type 1 neurofibromatosis, and was named in 1937 by Worster-Drought, *et al.* who considered it a forme fruste of neurofibromatosis. Since then only a few cases have been reported in the literature.

Meningioangiomatosis is composed of two components: 1) an intraparenchymal proliferation of small...
vessels accompanied by concentric perivascular proliferation of spindle-shaped meningeal cells similar to those found in a meningioma, and 2) an overlying leptomeningeal component of proliferating meningotheial cells that exhibit marked degenerative reactions such as calcification, fibrocortilage, and bone formation. Grossly, the lesions are usually firm, solitary, and sharply demarcated, with the intraparenchymal component limited to the cerebral cortex. Unique features of meningioangiomatosis include: lack of leptomeningeal involvement, lack of leptomeningeal calcification, association with Alzheimer-type neurofibrillary tangles within neurons in and around the lesion, leptomeningeal arteriovenous angioma without perivascular proliferation of meningocytes, intracortical clusters of fibroblast-like cells that do not form vessels, dense gliosis in the adjacent cortex, multiple lesions, and involvement of locations other than the cerebral cortex.

Similar to the pathology of meningioangiomatosis, our case had meningotheial, spindle cell proliferation forming an irregular whorling pattern associated with small blood vessels, and immunohistochemical and electron microscopic evidence of meningeal origin but without the classic appearance of a meningioma. Because of these unique histological features and the indolent growth rate of the lesion, we categorized this process as meningioangiomatosis. The peculiarity of this case was the unique anatomical location of the lesion in the brain stem, which to our knowledge has not been reported previously.

Several theories have been advanced to explain the histopathogenesis of meningioangiomatosis. Russell and Rubinstein reported it as a true neoplasm, considering that the leptomeningeal component represents a meningioma “en plaque” that infiltrates the underlying brain via penetrating vessels, which are also proliferating. However, lack of the leptomeningeal component in several lesions and of the continuation between the leptomeningeal and intraparenchymal lesion make this explanation unlikely. Kasantikul and Brown proposed that meningioangiomatosis is a hamartoma or developmental abnormality that undergoes degenerative changes as seen by the presence of collagen and hyalinization of the vascular component. This contention is further supported by the association of meningioangiomatosis with neurofibromatosis.

The favored hypothesis, proposed by Kasantikul and Brown, claims that initial development of angiomatous tissue within the brain would initiate proliferation of meningocytes around the blood vessels. Chronic stimulation of the leptomeninges by the underlying cerebral lesion would eventually lead to the changes seen in the overlying leptomeninges. This theory is based on the observation that leptomeningeal cells ensheathing the perforating blood vessels penetrate deep into the brain tissue to the capillary level, and seems to apply to our case because no extra-axial component was seen by MR imaging or at surgery although leptomeningeal involvement may not be apparent without microscopic examination. In our case, the anatomical location in the brain stem led to early symptomatology and diagnosis, which precluded chronic stimulation of the overlying tissues to induce the leptomeningeal changes usually seen in lesions located in the cerebral cortex.

Clinical presentation is related to the anatomical location of the lesion and presence or absence of neurofibromatosis. In many cases, the lesion is asymptomatic and recognized as an incidental finding, usually in patients with other manifestations of neurofibromatosis. Symptomatic patients usually present with focal or generalized seizures attributable to the superficial location of the lesion in the cerebral cortex. In our case, the medullary location of the lesion was responsible for the unique clinical manifestation of cranial nerve involvement and ataxia.
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The diversity of imaging characteristics of meningioangiomatosis reported previously reflects the complex histological features of the lesion.1,5,7,10,12,16,17 Angiography is usually normal or shows signs of an avascular mass; only two cases have shown evidence of abnormal vascularity.2 On CT, the lesion is usually iso- to slightly hyperintense with the presence of calcification (from faint psammomatous to dense osteoid) and variable enhancement. Low-density masses as well as normal CT have also been reported.12 The MR imaging appearance has been reported for only seven cases, revealing well-circumscribed lesions, iso- to slightly hypointense on T1-weighted images, and iso- to markedly hypointense on T2-weighted images surrounded by a hyperintense rim, which corresponded to edema or gliosis in the adjacent brain. Postcontrast MR images, obtained in only two previously reported cases, showed marked enhancement in one and lack of enhancement in the other.12,17 In our patient, the lesion was slightly hyperdense on CT and iso- to slightly hypointense on MR imaging with intense enhancement after contrast administration. These signal characteristics, similar to those of a meningioma, were expected considering the histological predominance of the meningial component.

The rarity of the lesion and the nonspecific imaging characteristics make the diagnosis difficult, especially in cases not associated with neurofibromatosis. Differential diagnosis, depending on the location, includes vascular abnormalities,3 neoplastic conditions such as meningioma or glioma (especially oligodendroglioma,17 and non-neoplastic conditions such as granulomatous processes (sarcoidosis tuberculo- sis or syphilitic gumma).1,5 Definite diagnosis is only provided by the pathology.

The few reported cases do not allow any definite conclusions regarding the natural history of meningioangiomatosis. Although a progression of clinical symptomatology was noted in several cases, only one case had follow-up radiology for 6 years and this showed no perceptible change in the mass size.12 Histological evidence suggests progression, but this process seems to be slow.2,11,17 Its benign nature is indicated by the duration and stability of clinical symptomatology and remarkable recovery after surgical removal.2 The relatively rapid progression of clinical symptomatology in our case is attributed to the “malignant” location in the brain stem rather than to the histological characteristics of the lesion.

Surgical excision is the treatment of choice in patients with meningioangiomatosis and appears to be curative. Excellent results have been reported after complete or partial removal of lesions located peripherally in the cerebral cortex.1,5,7,9,10,14,16 Intimate association with the motor cortex and middle cerebral vessels of a more extensive lesion may preclude complete surgical excision.11 Partial removal was successfully performed on our patient; complete excision was not feasible given the unusual location in the medulla, the lack of a plane of dissection, and the hemorrhage encountered when a more radical excision was attempted.

Acknowledgments

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