Meningioangiomatosis: Report of Three Cases and Review of the Literature

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Abstract. Meningioangiomatosis is a rare condition, probably hamartomatous, characterized by proliferation of capillary-sized vessels, meningothelial cells, and fibroblasts within the cortex of the brain. Lesions may be single or diffuse and may be associated with neurofibromatosis type II. Clinically it presents with seizures but may be asymptomatic throughout life. We report 3 cases of meningioangiomatosis, 2 localized and 1 diffuse, all with different clinical manifestations. Differential diagnoses are discussed with a review of the literature. Since this condition is rare, close clinico-pathological correlation is essential. A correct diagnosis avoids further aggressive treatment. (received 26 July 2001; accepted 3 December 2002)

Introduction

Meningioangiomatosis is a rare, non-neoplastic, intracortical proliferation of meningothelial cells, microvasculature, and fibroblasts [1] that may mimic a neoplasm clinically and radiologically. The process may be isolated or diffuse; the diffuse form may be associated with neurofibromatosis. We report 3 cases of meningioangiomatosis, 1 diffuse and 2 localized, with a review of the literature. Cytological features of Cases 2 and 3 have been described previously [2].

Case Reports

Case 1. A 15-yr-old boy with no significant medical history presented with recent onset of headache, which was treated with acetaminophen. Several weeks after the onset of symptoms, his family found him dead in bed. At autopsy, multiple, granular, firm “tumor masses” were seen in the frontal cortex, basal ganglia, optic chiasm, and mid-brain (Fig. 1). The lesions were bilateral, but more prevalent on the left side. The masses varied from a few mm to 2.4 cm in greatest dimension. Their configuration was nodular in some places and ill defined in others. In addition to nodules, there were areas of diffuse thickening of the gray matter, with loss of distinction between the gray and white matter, the texture of these lesions also being granular and firm. The cerebellum was grossly unremarkable.

On microscopic examination, the well-formed masses exhibited meningothelial cells, focally forming typical “meningioma-like” clusters (Fig. 2B). In most areas, however, the cells were arranged in a sharp palisading pattern in a background of hyalinized stroma (Fig. 2A). The adjacent intra-cortical extensions of the lesion were characterized by prominent proliferation of uniform capillary-sized blood vessels with fibroblastic and meningothelial proliferation in the Virchow-Robin spaces. Foci of calcification were seen. Apart from the focal trapping of slightly gliotic brain tissue within these lesions, there was little disruption of the cortical matter. The lesions showed scant mitoses and lack of anaplasia; their features were characteristic of extensive diffuse meningioangiomatosis involving cerebral cortex, basal ganglia, thalami, and brain stem. The cause of death was acute subarachnoid hemorrhage due to rupture of a berry aneurysm of the anterior communicating artery, unrelated to meningioangiomatosis, the latter being an incidental finding.
Fig. 1. Case 1. Gross photograph of a coronal section of the brain at autopsy showing diffuse enlargement and a granular appearance of the basal ganglia. Also visible are two firm “tumor masses” in the sub-ependymal region (arrows).

Fig. 2. A. Case 1. Section from the “tumor masses” showing palisading arrangement of meningothelial cells resembling schwannoma (H & E, x 200). B. Case 1. Meningioma-like clusters (H & E, x 200). C. Case 2. Thickened meninges with proliferation of capillary-like blood vessels in the underlying cortex (H & E, x 100). D. Case 3. Calcific deposits in the meninges (arrows). These were also present within the cortex (H & E, x 200).
**Case 2.** A 71-yr-old man presented with 8-mo history of headache, change in vision, loss of depth perception, and gait imbalance. His medical history included hypertension, cardiac disease, transient ischemic attacks and paraesthesias, depression, and short term memory failure. Magnetic resonance imaging (MRI) showed features of an infiltrative lesion in both occipital lobes, greater on the left side, extending to the parietal and temporal lobes, with a mass effect on the posterior horn of the left lateral ventricle. At operation, the surgeon noticed that the surface of the brain was boggy and non-pulsatile and had gray discoloration with small blood vessels. The gyral pattern was maintained. A frozen section was reported as benign brain tissue with gliosis. Permanent sections showed features of meningioangiomatosis (Fig. 2C). The patient had an uneventful recovery and was discharged 2 days later. He remains well 5 mo later.

**Case 3.** A 3-yr-old girl (birth weight 4.4 kg, born to a 36-yr-old woman by normal vaginal delivery) had been well until 1 day prior to admission, when she had a focal seizure on the right side accompanied by fever, perioral cyanosis, and vomiting. The child had seemed irritable for the past few weeks, especially in the morning. On admission, physical examination was normal and blood and CSF cultures were negative. MRI revealed an enhancing calcifying lesion in the left anterior inferior temporal lobe measuring 1.7 x 1.0 cm with minimal mass effect. The child underwent craniotomy and excision of the lesion. She had an uneventful recovery and was discharged 3 days later. She remains well 3 mo later.

**Discussion**

Meningioangiomatosis is a rare condition that occurs both in sporadic and syndromic forms, the latter with neurofibromatosis, type II. Most cases are asymptomatic; the symptomatic patients tend to present during childhood and early adulthood, usually with seizure [1]. The tumor grows slowly, if at all, and hence is not associated with features of increased intracranial pressure.

Sporadic lesions are single, intracortical, and measure up to a few centimeters. Their location has been in frontal, temporal, or parietal cortex with some reports in the third ventricle, cingulate gyrus, and pulvinar [3]. Multiple lesions tend to occur in association with neurofibromatosis, type II, and range in size from microscopic to grossly visible nodules. The lesions associated with the syndrome, however, may remain undiagnosed until autopsy [4,5]. The first case in the present study was a young man who died of an intracranial hemorrhage. He had multiple meningioangiomatosis but no other stigmata of neurofibromatosis II. No genetic studies were possible in this case.

Grossly, meningioangiomatosis is composed of well-demarcated thickening of the cortex with an overlying meningeal plaque. The gyral pattern is maintained but the surface of the brain may appear abnormal. The surgeon’s description in case 2 of the brain being boggy with widened gyri was very precise and helpful to the pathologist in interpreting this lesion at the intra-operative consult, emphasizing the importance of clinicopathological correlation in such unusual lesions.

Microscopy reveals characteristic proliferation of small caliber vessels in the cortex, accompanied by a cuff of meningotheelial cells and fibroblasts. The cortex between the vascular and meningotheelial proliferation either remains normal or shows reactive astrocytic changes. Leptomeningeal thickening is due to proliferation of meningeal cells. Calcification is usually present in the leptomeningeal and cortical portions, both in vessels and surrounding cortex. All of the 3 cases showed the typical features. In case 1, the striking palisading pattern was unusual and resembled schwannoma. The intracortical location and associated angiomaticatosis, however,
precluded that diagnosis. In case 3, meningothelial proliferation was seen outside the cortex but was restricted to the sulci.

Neurofibrillary tangles (NFTs) have been described in both the intervening and distant cortex associated with meningioangiomatoses [3,6]. Their pathogenesis is unclear but they probably indicate degenerative changes in neurons [3]. We performed Bielchowsky stain in all 3 cases but failed to demonstrate NFTs. So-called “calcified rocks,” described by several authors in long-standing lesions [7-9], are hard nodules in the superficial cortex, but they were not observed in our 3 cases.

Differential diagnoses include astrocytomas, vascular tumors, and meningiomas with cortical invasion. Mild astrocytic proliferation accompanied by reactive atypia may suggest an astrocytic neoplasm. Furthermore, prominent endothelial proliferation might result in an erroneous diagnosis of a high-grade glioma, especially on imprint cytology. The absence of significant cellularity and glomeruloid vessels and the presence of neurons within the lesion should preclude that diagnosis, however. The presence of small sized vessels, their uniform distribution, lack of hemorrhage, normal intervening tissue, associated meningothelial and fibroblastic proliferation, and superficial location should preclude diagnosis of a vascular tumor or malformation. A florid meningothelial component may raise suspicion of a menigioma with cortical invasion. Lack of meningothelial atypia, necrosis, or cortical destruction favor meningioangiomatosis.

Meningiomas with underlying meningioangiomatosis are extremely rare [10].

The pathogenesis of this condition is unclear but a hypothesis offered by Halper et al [3] appears plausible. Leptomeningeal vessels, as they penetrate the cortex, are surrounded by meningothelial cells [11] that may proliferate under the influence of an unknown stimulus [12]. Vascular malformations are often an accompaniment of phakomatoses, supporting the nomenclature of meningioangiomatosis as a hamartoma rather than a neoplasm; this also makes the association with neurofibromatosis understandable.

In summary, meningioangiomatosis is a malformative condition that presents as a mass-occupying lesion within the cortex. It should be considered in differential diagnosis of intracortical lesions, especially in children and young adults. The superficial location, plump vessels of small size, evenly spaced without destruction of adjacent cortex, cuffed by meningothelial and fibroblastic cells, and overlaid with a thick, calcified meningeal plaque, should clinch the diagnosis. Proper diagnosis is important because of the excellent prognosis and absence of recurrence in sporadic cases.

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