Pathologic Quiz Case

Progressive Headaches in a 50-Year-Old Woman

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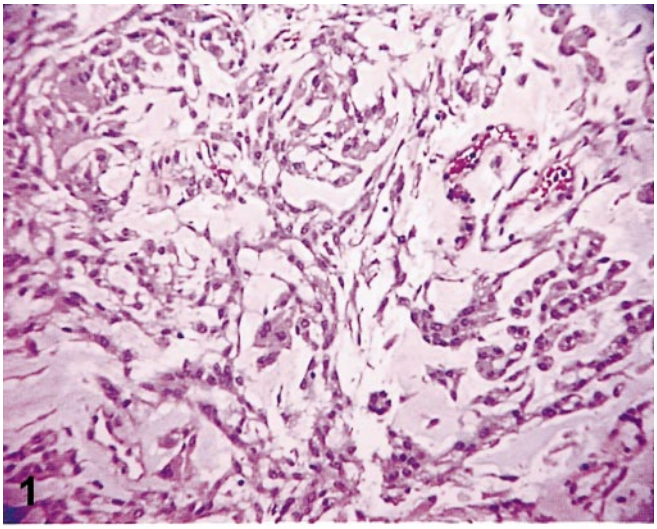
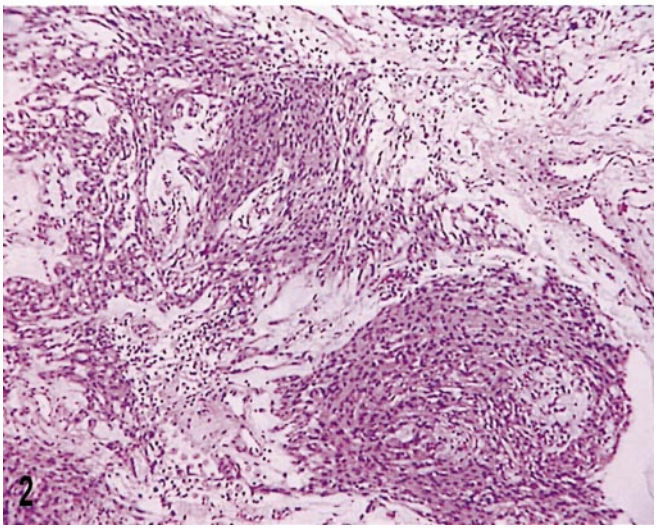
A 50-year-old woman presented with progressive headaches, unstable gait, short-term memory deficit, and mood swings. Bladder and bowel function were unaffected. No other focal deficits were noted. The patient had no history of trauma. Past medical history was significant for rheumatoid arthritis treated with piroxicam and hydroxychloroquine sulfate. Serum electrolytes and white blood cell count were within normal limits. She was mildly anemic. Hemoglobin and hematocrit values were 9.5 g/dL and 28%, respectively. A computed tomographic scan of the brain demonstrated a large, enhancing left lateral sphenoid wing tumor, measuring 5 to 6 cm in greatest dimension with surrounding edema. A follow-up magnetic resonance imaging/magnetic resonance angiography study confirmed a large, left frontotemporal, extra-axial tumor with generalized enhancement and evidence of hemorrhage within the tumor. No large vessels were noted to feed into the tumor; however, the left middle cerebral artery was markedly displaced. The patient was treated with phenytoin and a craniotomy was performed.

Surgery yielded a 3.0 × 2.5 × 1.0-cm aggregate of tan-pink, mucoid, focally hemorrhagic soft tissue fragments. Histopathology revealed abnormal trabeculae composed of vacuolated eosinophilic cells in a myxoid background (Figure 1). Also identified were small areas composed of whorled epithelial cells (Figure 2).

What is your diagnosis?





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Pathologic Diagnosis: Chordoid Meningioma

Meningiomas are generally benign, slowly progressive neoplasms, which originate from the meningothelial cells of the arachnoid membrane.[1] They are common intracranial tumors that typically manifest in middle-aged and elderly patients, with a peak incidence during the sixth and seventh decades of life and a female predominance. Meningiomas may occur in children and tend to be more aggressive.

Most meningiomas are located within the intracranial, orbital, and intervertebral cavities, although rare meningiomas have been reported in almost every organ.[2] They precipitate clinical signs and symptoms by compressing nearby structures. Therefore, specific deficits are dependent on the location of the tumor. Multiple meningiomas are associated with neurofibromatosis type 1, but usually occur as a single neoplasm.[2]

Meningiomas manifest in a variety of histologic patterns. Common features of meningiomas are syncytial arrangement of cells, nuclear pseudoinclusions, whorled architecture, and psammoma bodies, best demonstrated in the meningothelial type.

The first known case of chordoid meningioma was reported in 1980 by Connors.[3] This unusual variant was first named in 1988 by Kepes et al,[1] who studied 7 patients ranging from 8 to 19 years of age. The patients had meningeal neoplasms with a myxoid-chordoid pattern. These patients also developed systemic symptoms of iron-refractory, hypochromic, and microcytic anemia. One patient developed a dysgammaglobulinemia and retarded growth. These findings were consistent with Castleman syndrome.[1,4] More recent studies have shown chordoid meningiomas to occur primarily in adults; no sex predilection or systemic manifestations have been noted.[2]

Chordoid meningiomas feature a mixture of epithelioid and spindled cells within a myxoid matrix.[1–5] The histologic appearance closely resembles a chordoma.[1–5] The tumor exhibits cytoplasmic vacuolation and clustering or cords of tumor cells.[2,5] Meningothelial foci are also usually present. In addition, these tumors are often surrounded by a heavy lymphocytic infiltrate, often showing follicles and germinal centers; however, this feature is not diagnostic.[1,4] Most lymphocytic infiltrates in all meningioma types are composed of T cells; however, chordoid meningiomas of childhood are strongly associated with B lymphocytes and plasma cells.[4]

Ultrastructurally, meningiomas have intercellular desmosomal junctions, intermediate filaments, and complex interdigitating cell processes, which at times may be difficult to appreciate in the chordoid variant.[2,3] The vast majority of the tumor cells exhibit positive immunostaining for vimentin and epithelial membrane antigen, but are negative for glial fibrillary acidic protein, cytokeratins, carcinoembryonic antigen, and S100 protein.[4,5] MIB-1 labeling may be helpful in predicting recurrence and survival. More than 5% to 10% positivity suggests a greater propensity for recurrence.[2]

Chordoid meningiomas have a high rate of recurrence (42% in one series), which may be due to the mucoid quality of its stroma, which, similar to chordomas, facilitates the spread and recurrence of tumor.[4] Based on this clinical behavior, it is classified as World Health Organization grade II.[2]

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