Dural Lesions Mimicking Meningiomas

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Recently, a number of neoplastic and nonneoplastic entities have been reported that radiographically and clinically mimic meningiomas. Because these lesions occur infrequently and may resemble a meningioma during intraoperative analysis, they may not be considered in the differential diagnosis. This review (and case illustrations) considers some of the newly recognized and notable lesions that can mimic meningiomas, including solitary fibrous tumors, gliosarcoma, leiomysarcoma, hemangiopericytoma, melanocytoma, Hodgkin’s disease, plasmacytomas, inflammatory pseudotumors, neurosarcoïdosis, plasma cell granulomas, Rosai-Dorfman disease, Castleman’s disease, xanthoma, rheumatoid nodules, and tuberculosis. Awareness that these lesions involve the dura may facilitate intraoperative recognition and, in some cases, preclude unnecessary additional surgery.

The occurrence of neoplastic and nonneoplastic dural-based masses that mimic meningiomas has received little attention despite several recent reports. Because these occur infrequently, such lesions may not be considered during intraoperative analysis. Because their histologic features resemble variants of meningiomas (Table 1), postoperative recognition may be challenging as well. This review (and case illustrations) considers some of the newly recognized and notable lesions typically mistaken clinically or radiographically for meningiomas. Awareness that they involve the dura may facilitate intraoperative recognition and, in some cases, prevent unnecessary additional surgery.

SOLITARY FIBROUS TUMORS

On rare occasions, solitary fibrous tumors (SFTs) may develop in the leptomeninges, where they mimic a meningioma. At least 13 cases have been reported. Typically they present in the same age group as meningiomas (mean age, 57 years) and show a strong predilection for females (a 5:2 female: male ratio). They have been observed at the falx, occipital and spinal dura, tentorium, and cerebellopontine angle. Other characteristics similar to meningioma are revealed by computed tomography (CT) and magnetic resonance imaging (MRI) showing a circumscribed dural-based tumor occasionally associated with hyperostosis. Typically these lesions enhance uniformly after administration of gadolinium.

Macroscopically, SFTs are firm, gray-white, and circumscribed but not encapsulated. Reported cases range in size from 1 to 7 cm in diameter. In 1 case, an SFT exhibited underlying invasion of the brain.

Microscopically, SFTs superficially resemble fibrous meningiomas. However, unlike fibrous meningiomas, SFTs usually exhibit different histologic patterns throughout the tumor. Spindle cells in fascicles or individually enveloped in collagen may dominate parts of the tumor. Other zones exhibit a capillary pattern reminiscent of a hemangiopericytoma (HPC) (Fig 1). Whorls, storiform patterns, and psamomma bodies are absent in dural lesions, although the latter have been noted in tumors at other sites. The spindle cells have elongated or round nuclei but less conspicuous nucleoli and none of the pseudoinclusions characteristic of meningiomas. SFT cells, in contrast to fibrous meningiomas, do not stain with periodic acid-Schiff (PAS). In hypocellular areas, tumor cells encircle densely hyalinized capillaries (Fig 1).7,8,12 As in pulmonary tumors, capillary proliferation may be prominent and resemble an HPC in regions with numerous elongated channels. Nonetheless, in our experience capillaries tend to be more frequently ensheathed in collagen and less commonly branched than HPCs.6,8,12

SFTs exhibit diffuse CD34 immunoreactivity in stromal spindle cells as well as endothelia in nearly all cases, in contrast to the patchy immunostaining seen in 30% of HPCs and in 60% of fibrous meningiomas. Epithelial membrane antigen (EMA) and S-100 protein immunoreactivity are not usually seen—a feature that helps distinguish them from fibrous meningiomas.

Key words: dura, meningioma, solitary fibrous tumor, gliosarcoma, hemangiopericytoma, inflammatory pseudotumors, neurosarcoïdosis, Hodgkin’s disease, plasmacytoma, plasma cell granuloma, leiomysarcoma, rheumatoid nodules, Rosai-Dorfman disease, Castleman’s disease, xanthoma, melanocytoma.

Abbreviations: CNS, central nervous system; CT, computed tomography; EBV, Epstein-Barr virus; EMA, epithelial membrane antigen; GS, gliosarcoma; HD, Hodgkin’s disease; HPC, hemangiopericytoma; HPF, high-power field; MRI, magnetic resonance imaging (MRI); PAS, periodic acid-Schiff; PGC, plasma cell granuloma; RDD, Rosai-Dorfman disease; R-S, Reed-Sternberg; SFT, solitary fibrous tumor.
which exhibit diffuse immunoreactivity for these antigens 80% of the time.2,7,9,12 (Fig 2).

Case Illustration

A 57-year-old male presented with right facial weakness. MRI revealed an inhomogenous 7.5-cm mass extending from the dura over the petrous bone that compressed the right pons and midbrain, with evidence of erosion of the superior petrous apex and right petrous carotid canal (Fig 1A). Intraoperatively, the tumor appeared encapsulated and attached to the trigeminal nerve. Near-total resection was achieved. The sample consisted of multiple large, white, firm, nodular fragments of tissue measuring 7.5 × 4.0 × 1.3 cm. Microscopically, the tumor appeared unencapsulated and populated by fascicles of elongated spindle cells organized in hypercellular zones. In other areas, spindle cells were separated by dense bands of collagen, which also surrounded elongated, occasionally branched blood vessels (Fig 1B, C, and D). Also present were hypocellular myxoid zones with lymphoid cells and hyalinized blood vessels. Zones populated by dense collagen surrounding round tumor nuclei cut in cross-section were also present (Fig 1C). Tumor cells exhibited no EMA or S-100 protein immunostaining, but did exhibit extensive CD34 immunoreactivity (Fig 1E).

HEMANGIOPERICYTOMAS

Representing about 0.4% of primary brain tumors,13 HPCs were once classified as a variant of meningioma. Like meningiomas, the majority of intracranial HPCs are supratentorial.12,13 HPCs are typically lobular in MRI; in 2/3 of cases, this tumor has a broad-based attachment to the dura. On T1-weighted images, HPCs appear isointense with gray matter and enhance heterogeneously after administration of gadolinium. They may show bony erosion but do not show the hyperostosis or intratumoral calcification commonly associated with meningiomas (Fig 3).14 Their histologic similarity to vascular meningiomas and SFTs has been reviewed in detail.12-15 In most cases, these highly cellular tumors are populated by sheets of cells with ovoid or round hyperchromatic nuclei separated by reticulin and numerous, often inconspicuous capillaries. Mitoses are readily found. More prominent, often branching capillaries produce a characteristic “staghorn” vascular pattern, but the hyalinized vessels of a meningioma or collagen-sheathed vessels of an SFT are not usually encountered. Fibrous areas with collagen and elongated cells, particularly in a fragmented sample, may superficially resemble a fibroblastic meningioma, although the smaller nuclear size and absence of nuclear inclusions argue against a diagnosis of meningioma. Other meningioma variants might also be suggested in a fragmented biopsy, but true whorls and large hypovascular meningotheelial areas are not seen. Recognition of the numerous fine capillaries and increased mitotic activity usually establishes the diagnosis (Fig 2). Differentiating HPCs from SFTs and meningiomas is critical, because their treatment and prognosis differ.2,12,15 A number of tumors may exhibit vascular patterns similar to HPCs, but with dural-based tumors the differential diagnosis includes vascular meningiomas, hemangioblastomas and SFTs. When the histology is insufficient to make a diagnosis, immunohistochemistry with antibodies to factor XIIIa, EMA, S-100 protein, and CD34 may help differentiate these tumors.9,15,16 In contrast to SFTs, HPCs exhibit CD34 immunostaining in stromal cells in only 33% of cases, and is often patchy and less intense than immunoreactivity in endothelial cells.2,12 Factor XIIIa immunoreactivity, seen in 78% of HPCs, is noted in 100% of SFTs and 65% of meningiomas. In contrast to fibroblastic meningiomas, 80% of which exhibit EMA and S-100 immunoreactivity,17 these epitopes are essentially not seen in HPCs. Vimentin immunostaining is widespread in all 3 entities and is not diagnostic2,12 (Fig 2).

GLIOSARCOMAS

Gliosarcomas (GSs) represent 1.7% to 2.3% of glioblastomas.18-20 Occasionally, a GS developing superficially in the temporal lobe may extend into the leptomeninges, eliciting a desmoplastic response that produces an encapsulated, firm lesion both radiographically and grossly suggestive of a meningioma (Fig 4). In 1 series, approximately 12% of cerebral GSs presented as dural-based tumors.20 An extraaxial dural GS of the thoracic spinal cord has also been reported.21 Intraoperatively, the firmness and circumscription of GSs that have invaded the leptomeninges may further suggest a meningeal origin. In tumors with a predominately fibrosarcomal pattern, intraoperative differentiation between GS and a malignant fibrous meningioma may be challenging because both are firm and have a prominent spindle cell component. Although
identification of an astrocytic component or sarcoma associated with blood vessels facilitates the diagnosis of GS, these features may not be apparent on frozen sections. The true identity of the tumor is readily apparent on permanent sections demonstrating the presence of sarcoma, usually arising from the adventitia of blood vessels, associated with islands of malignant glioma. Any concern that the tumor represents a malignant meningioma may be dispelled with immunohistochemical studies demonstrating nests of glial fibrillary acidic protein (GFAP) immunoreactive anaplastic cells, factor VIII immunoreactive sarcoma around blood vessels, and lack of widespread EMA immunostaining\(^22\) (Fig 2).

Case Illustration

A 49-year-old male presented with progressive left leg weakness. On examination, he was also found to have homonymous hemianopsia with intact extraocular movements and cranial nerves. He exhibited upgoing

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**FIGURE 1.** Solitary fibrosis tumor (SFT). (A) A T1-weighted MRI image obtained after gadolinium administration demonstrating an enhancing mass attached to the dura over the petrous bone, compressing the pons and midbrain with extension into the foramen magnum. (B) Spindle cells are separated by collagen. (C) In cross-sections of areas with dense collagen, tumor cells appear round. (D) Elongated blood vessels flanked by collagen are prominent elsewhere. (E) Spindle cells exhibit extensive, diffuse CD34 immunoreactivity. (Hematoxylin and eosin; original magnification ×60 (B), ×120 (C), ×120 (D), ×60 (E).)
toes and decreased proprioception on the left side. Strength was 4/5 in the left leg and 5/5 in the right extremities. MRI revealed a 4.0 × 4.0–cm mass extending from the dura into the temporal lobe and a sylvian fissure compressing the posterior right frontal lobe and the anterior horn of the right lateral ventricle (Fig 4A). The mass enhanced irregularly with gadolinium administration.

Intraoperatively, the tumor was firm and was shelled out of the sylvian fissure. Sections through the mass revealed a firm, white, solid tumor without necrosis or hemorrhage. Intraoperative smears and frozen sections revealed a biphasic neoplasm populated by spindle cells with nuclear atypia and occasional mitoses flanked by collections of neoplastic astrocytes, suggesting a sarcomatoid glioma. Permanent sections of the tumor demonstrated large zones of spindle cell sarcoma (Fig 4B and C). In some areas, this extended from blood vessels (Fig 4D and E). GFAP-immunoreactive neoplastic astrocytes (Fig 4E) were present in other zones of the tumor. Tumor cells exhibited no EMA immunoreactivity.

LEIOMYOSARCOMAS

On rare occasions, leiomyosarcomas can present as primary intracranial tumors. These tumors arise primarily in males and are more commonly associated with human immunodeficiency virus-1 infection and immunosuppression.\(^23\)\(^{-26}\) However, a dural leiomyosarcoma has been reported in an immunocompetent 14-year-old female.\(^27\) Leiomyosarcomas develop as solitary masses involving the dura of the sphenoid wing, transverse or cavernous sinus, or occipital or temporal lobe. A rare metastatic leiomyosarcoma presenting as a meningioma-like mass has also been reported.\(^28\) Angiography may reveal contrast pooling, suggesting a cavernous hemangioma.\(^24\) MRI shows a signal intensity on T1 and T2 of the gray matter\(^25\) that is enhanced with
gadolinium administration. Histologically, these tumors tend to be low-grade spindle cell lesions containing elongated nuclei with delicate chromatin and scattered mitoses (Fig 5B). As with leiomyosarcomas at other sites in immunocompromised patients with human immunodeficiency virus-1, Epstein-Barr virus (EBV) ER1 mRNA and EBV nuclear antigen, or EBV latency-associated protein, has been demonstrated in tumor cells, suggesting a potential role in the pathogenesis of these neoplasms24,27 (Fig 2).

METASTATIC CARCINOMAS

Metastatic carcinoma can produce isolated dural lesions mimicking a meningioma. These most commonly result from breast cancer, adenocarcinomas, squamous cell carcinoma of the lung, and renal cell carcinomas.29-31 Advances in chemotherapy may be altering the current incidence, but in the last decade, solitary metastasis to the dura in breast cancer occurred in approximately 8% of autopsied patients with this
primary.32 Rarely, metastasis from carcinoids, adenoid cystic carcinomas, prostatic adenocarcinomas, and dermatofibrosarcomas have also produced meningioma-like masses.32-36 Typically, these tumors produce MRI images with increased signals on T2-weighted images and often with an enhancing dural tail mimicking a meningioma (Fig 6A). Histologic features usually reveal the metastatic nature of the tumor (Fig 6B); however, lobular growth may mimic whorls or a syncytial growth pattern, raising the possibility of a malignant meningioma (Fig 6C). Because metastatic carcinomas usually exhibit more intense cytokeratin and no S-100 protein expression, immunohistochemical analysis generally excludes meningioma17,22 (Fig 2).

Case Illustration

A 61-year-old woman presented with blurred vision and supraretro-orbital pain in her right eye. Three months earlier, an ophthalmologist had detected a cataract in her right eye. The patient had a history of carcinoma of the breast (stage II, N1, MO) with a mastectomy and chemotherapy 8 years earlier and had recently completed radiotherapy and chemotherapy for recurrent metastases to the spine. The patient’s extraocular muscles were normal on physical examination, but she reported pain with movement. Tenderness was also noted over the lumbar spine. Because of persistent symptoms, MRI was ordered; this revealed a 4 × 1 × 1-cm extraxial mass of the middle cranial fossa. Intraoperatively, the tumor was found to be attached to the dura and infiltrating the lateral wall of the orbit. Grossly, the tumor was tan soft tissue. Microscopically, the lesion appeared to be a carcinoma infiltrating the dura, organized largely in nests. Tumor cells exhibited modest EMA, intense AE1/AE3, cytokeratin-7 HER2/neu, and estrogen and progesterone immunoreactivity consistent with metastatic breast carcinoma (Figs 2 and 6).

HODGKIN’S DISEASE

Hodgkin’s disease (HD) spreads hematogenously to the leptomeninges and dura in 0.2% to 0.5% of patients with stage II or III disease, producing a firm, circumscribed dural-based tumor with radiographic and intraoperative features of a meningioma.38,39 Rarely, HD may also present as a primary central nervous system (CNS) tumor; 3 of the 9 reported cases were originally considered meningiomas.40-44 HD must be differentiated from lymphomas, lymphoplasmacytic meningiomas, inflammatory pseudotumors, neurosarcoidosis, or infectious processes. The presence of Reed-Sternberg (R-S) cells with extensive or limited membrane or cytoplasmic CD15 and CD30 immunoreactivity in a lesion with polytypic T and B lymphocytes and eosinophils supports a diagnosis of HD.45 In contrast to lymphoplasmacytic meningiomas, HD displays no identifiable meningothelial component with EMA immunostaining. The tumor has no granulomas asteroid or Schaumann bodies of neurosarcoidosis. In contrast to Rosai-Dorfman disease, this lesion lacks a histiocytic component and emperiploise. Plasma cell granulomas do not contain R-S cells or significant eosinophilic infiltrates. Like its systemic counterpart, CNS HD exhibits EBV antigens in R-S cells in approximately 1/2 of cases and was recently described in a case of primary CNS HD (Fig 2). The diagnosis of primary CNS HD is predicated on the exclusion of HD elsewhere in the body, because metastatic lesions are far more common.46 A thorough staging should include bone marrow biopsy, MRI of the chest and abdomen, and ophthalmologic examination.

Case Illustration

A 55-year-old woman presented to the emergency room complaining of a 1-week history of worsening headaches and difficulty walking. She reported having had headaches “for years.” On MRI, T1-weighted im-
ages demonstrated a hypointense posterior fossa mass arising from the inferior aspect of the tentorium. The 2.3 × 3.2 × 3.2–cm mass was enhanced markedly after gadolinium administration and had a dural tail (Fig 7A).

Intraoperatively, the mass was attached to the tentorium and was firm. Frozen sections revealed an inflammatory lesion. A gross total resection was achieved.

Microscopically, the lesion was composed of numerous small lymphocytes, histiocytes, scattered eosinophils, occasional large dysplastic cells with prominent inclusion-like nucleoli, and scarce binucleate lacunar cells (Fig 7B). The small lymphocytes were predominantly UCHL-1– and CD3-immunoreactive T cells, with occasional CD20-immunoreactive B cells. The large dysplastic R-S cells showed strong membranous and Golgi immunostaining with CD30 and focal staining with CD15 (leu M-1). The R-S cells did not react with antibodies to EMA, UCHL-1, CD3, CD20, or S-100. Numerous R-S cells showed immunoreactivity for EBV latency-associated protein (Fig 7G). The small plasma cells were polytypic in terms of IgG, IgM, IgA, and kappa and lambda light chain reactivity and were EMA immunoreactive. There was rare S-100 protein staining in scattered histiocytes. Neither emperipoleses nor granulomas were found, and special stains revealed no bacteria, fungi, spirochetes, or acid-fast bacilli. Meningothelial cells were not seen. A diagnosis of HD, nodular sclerosing type, was made.

Subsequent evaluation, including bone marrow biopsy; CT scans of the chest, abdomen and pelvis; and ophthalmologic evaluation, revealed no evidence of systemic disease.
Plasmacytomas

Plasma cell neoplasms rarely involve the CNS as dural-based lesions. Two reported cases have been reported in older adults as masses attached to the frontal dura or falx. On T1-weighted images, the tumor exhibits an intermediate signal compared to that of brain tissue, with marked enhancement after gadolinium administration. On T2-weighted images, plasmacytomas are isointense with gray matter. Histologically, they are identical to plasmacytomas elsewhere. Plasmacytomas must be differentiated from plasma cell granulomas, which also may present as dural-based lesions. Because plasmacytomas may contain leptomeninges, they must also be differentiated from lymphoplasma cytic meningiomas. Usually, their homogeneity and monoclonal immunophenotype differentiate them from both granulomas and meningiomas (Figs 2 and 8). Their recognition and differentiation from surgically managed plasmacytic meningiomas is important, because radiation and chemotherapy after surgical resection offer a potential cure.

Rosai-Dorfman disease (RDD), or sinus histiocytosis and massive lymphadenopathy, is a reactive condition typically associated with painless cervical lymphadenopathy, fever, and hypergammaglobulinemia. At least 6 cases of solitary dural-based RDD, mimicking meningioma and without nodal involvement, have also been described. These occur predominantly in adult males and may involve the epidural and/or subdural space at parasagittal, suprasellar, or cerebellar pontine angle sites. CT scans typically reveal a contrast-enhancing hypodense tumor. Histologically, these lesions are populated by foamy histiocytes (Fig 9B) with S-100 protein, CD68, α-1-antitrypsin, and α-1-antichymotrypsin immunoreactivity but no CD1a immunostaining. The infiltrate also contains B and T lymphocytes, plasma cells, and occasionally eosinophils (Fig 9C). Central necrosis may be present. The hallmark of

FIGURE 6. Dural Hodgkin’s Disease. (A) A T1-weighted MRI image obtained after gadolinium administration demonstrating an enhancing mass with a dural tail arising from the inferior aspect of the tentorium cerebelli. (B) The tumor is composed of an infiltrate of lymphocytes, plasma cells, eosinophils, and R-S-like cells in fibrotic stroma. (C) R-S cell variants exhibit EBV latency-associated protein. (D) PAS, original magnification ×120; (E) hematoxylin, original magnification ×250.)

FIGURE 7. Plasmacytoma. The tumor is populated by a monomorphic population of dysplastic plasma cells without lymphocytes or histiocytes. (Hematoxylin and eosin; original magnification ×140.)

FIGURE 8. Rosai-Dorfman disease. (A) An MRI image demonstrating an extra-axial lesion over the left parietal and occipital lobes thought to be a meningioma en plaque. (B) Foamy histiocytes are present in varying numbers. These histiocytes may exhibit emperipolesis. (C) Other areas of the mass contain a mixed inflammatory infiltrate of lymphocytes and plasma cells (Hematoxylin and eosin; original magnification ×300.)
RDD is emperiplolesis, primarily of lymphocytes, which may be inconspicuous (Fig 9B).

Dural RDD can be managed with surgical resection and has a good prognosis.90-64 It must be differentiated from more ominous lesions requiring aggressive therapy, such as Langerhans cell histiocytosis, HD, malignant histiocytic lymphoma, and plasma cell granuloma. In contrast to histiocytes in Langerhans cell histiocytosis, histiocytes in RDD are CD1a negative and lack striking nuclear grooves and Bierbeck granules ultrastructurally. Moreover, emperiplolesis is not seen in Langerhans cell histiocytosis. In contrast to HD, large R-S-like cells immunostain for S-100 protein but are negative for CD15, CD30, and EBV latent membrane protein. As described later, plasma cell granulomas are typically more fibrotic, contain histiocytes with no S-100 protein immunostaining, and lack emperiplolesis61,62,64 (Fig 2).

NEUROSARCOIDOSIS

Sarcoidosis involves the CNS in 5% of cases, accounting for approximately 5 per 1 million population.65 It usually develops in the basal leptomeninges and may involve the hypothalamus, infundibulum, or cranial nerves. Lesions seen on CT scan and operative features of meningioma have been reported at the cribriform plate, sphenoid wing, optic nerve, and cerebellar pontine angle.66-72 In rare parasagittal,68 convexity,69 parafalcine,70 and cerebellopontine angle examples,71-73 there has been no evidence of systemic disease, and angiotensin-converting enzyme levels were normal. Tu- mor-related disability due to lumbar disc disease for some time. He had a history of hypertension, hypercholesterolemia, and depression. Physical examination revealed no neurologic or ophthalmologic deficits or lymphadenopathy.

MRI of the head demonstrated a parietal 2.5 × 2.5–cm dural-based mass thought to be a meningioma associated with vasogenic edema of the underlying brain (Fig 11A). A second 1.25 × 1.25 × 0.75–cm lesion was identified at the cervical–medullary junction. MRI of the cervical spine revealed no abnormalities.

Surgery was directed at the parietal lobe lesion. Intraoperatively, the lesion was dural based, with small satellite lesions. This was resected without complications. Microscopically, the lesion was composed of dura with nodules of plasma cells and lymphocytes separated by hyalinized zones. Rare binucleate plasma cells and Russell bodies were present, but overtly dysplastic plasma cells were not found. Scattered histiocytes with no S-100 protein immunoreactivity were also present. Immunohistochemical analysis revealed a polytypic plasma cell infiltrate. No meningioma-like histology or EMA immunoreactivity was identified. Giant cells, foamy histiocytes, emperipolesis, and zonal necrosis were not present (Fig 11B and C). A diagnosis of hyalinizing plasma cell granuloma was made.

Two years postoperatively, there has been no recurrence of the parietal lesion, the foramen magnum lesion is stable, and the patient is in good health.

PLASMA CELL GRANULOMAS

On rare occasions, plasma cell granuloma (PCG) may present as a dural-based mass clinically indistinguishable from meningioma. In 4 cases previously reported and 1 case described herein, these reactive lesions occurred primarily in adults, usually presenting as discrete tumors arising from the leptomeninges or dura. MRI revealed little evidence of parenchymal invasion,72-74 although parenchymal involvement has been described.75 Histologically, they are populated by a polytypic population of mature plasma cells, plasmacytoid, and small nontransformed lymphocytes in a background of variable fibrosis, fibroblasts, and entrapped leptomeningeal cells (Fig 11B). Russell bodies are usually present.71-74 PCGs with broad bands of fibrosis may represent a variant sometimes described as “hyalinized”74 (Fig 11C). The reactive nature of these lesions is documented using immunohistochemistry for light chains, IgG, IgM, and IgA, which establishes the polyclonal nature of the process. PCGs must be differentiated from the other inflammatory lesions mentioned earlier as well as from rare lymphoplasmacytic meningiomas. Although focal EMA immunostaining may be seen within entrapped arachnoid, PCGs lack the focal areas of overt meningiomas and the extensive EMA immunoreactivity common to meningiomas.17 In contrast to RDD, histiocytes in PCG are S-100 negative and devoid of emperiplolesis. R-S cells and eosinophils are absent71-73 (Fig 2).

Case Illustration

A 42-year-old male body shop worker was evaluated for unremitting headaches. He had been on disability due to lumbar disc disease for some time. No neurologic or ophthalmologic deficits or lymphadenopathy.

MRI of the head demonstrated a parietal 2.5 × 2.5–cm dural-based mass thought to be a meningioma associated with vasogenic edema of the underlying brain (Fig 11A). A second 1.25 × 1.25 × 0.75–cm lesion was identified at the cervical–medullary junction. MRI of the cervical spine revealed no abnormalities.

Surgery was directed at the parietal lobe lesion. Intraoperatively, the lesion was dural based, with small satellite lesions. This was resected without complications. Microscopically, the lesion was composed of dura with nodules of plasma cells and lymphocytes separated by hyalinized zones. Rare binucleate plasma cells and Russell bodies were present, but overtly dysplastic plasma cells were not found. Scattered histiocytes with no S-100 protein immunoreactivity were also present. Immunohistochemical analysis revealed a polytypic plasma cell infiltrate. No meningioma-like histology or EMA immunoreactivity was identified. Giant cells, foamy histiocytes, emperipolesis, and zonal necrosis were not present (Fig 11B and C). A diagnosis of hyalinizing plasma cell granuloma was made.

Two years postoperatively, there has been no recurrence of the parietal lesion, the foramen magnum lesion is stable, and the patient is in good health.
XANTHOMAS

A xanthoma or xanthogranuloma may rarely present as a dural-based mass mimicking a meningioma. This may be primary or associated with histiocytosis X. To date, 3 primary cases have been reported, which presented as spinal lesions in children.76-78 After gadolinium administration, T1-weighted MRI demonstrates a homogeneously enhancing circumscribed mass. Histologically, these are encapsulated lesions populated by lipid-filled, CD68-positive, S-100-negative histiocytes with small nuclei accompanied by small non-transformed lymphocytes. In contrast, Langerhans cell histiocytosis of long duration may contain foamy histiocytes encircling a zone of necrobiosis flanked by lymphocytes, plasma cells, and a rare giant cell (seen in A). (Hematoxylin and eosin; original magnification ×60 (A) and ×300 (B).)
cytes, but these are S-100 protein positive and contain Birbeck granules ultrastructurally (Fig 2).

INFLAMMATORY PSEUDOTUMORS ASSOCIATED WITH SYSTEMIC DISEASE

Castleman’s disease has been associated with inflammatory pseudotumors of the dura that resemble giant lymph node hyperplasia associated with this disease.\textsuperscript{77,82} Most cases with CNS involvement have systemic manifestations with lymph node hyperplasia in the mediastium, abdomen, or neck. However, isolated CNS Castleman’s presenting as a meningioma-like mass attached to the tentorium cerebelli has recently been reported.\textsuperscript{82} These lesions produce insidious symptoms attributable to the site of origin. MRI reveals an isointense dural-based lesion that enhances uniformly with gadolinium.\textsuperscript{82} Intraoperatively, Castleman’s disease appears to be a typical meningioma.\textsuperscript{82} Histologically, this lesion resembles changes seen in lymph nodes with hyperplastic follicles of mature B and T lymphocytes surrounded by a polyclonal plasmacytic infiltrate with Russell bodies. Castleman’s disease has 2 variants: the hyaline/vascular and plasmacytic types. The former may be more common in the brain.\textsuperscript{79,82}

Castleman’s disease must be differentiated from a small-cell lymphoma, plasmacytoma, infectious process, or lymphoplasmytic meningioma. Demonstration of follicles with a nontransformed polytypic population of lymphocytes and plasma cells rules out the former. The absence of clinical evidence of infection and failure to demonstrate organisms in cultures and special stains argues against a mycobacterium infection. No meningial proliferation is seen in these lesions, excluding a diagnosis of lymphoplasmytic meningioma.

Rarely, rheumatoid arthritis is associated with the development of rheumatoid nodules that can mimic small meningiomas. Like their counterparts in joints, these lesions are composed of fibrous nodules with a polyclonal lymphocytic infiltrate around zones of necrosis with giant cells and epitheloid histiocytes\textsuperscript{83,84} (Figs 2 and 12).

LEPTOMENINGEAL MELANOCYTIC NEOPLASMS

Leptomeningeal melanocytes, present primarily in the basal and ventral spinal leptomeninges, occasionally give rise to a spectrum of melanocytic neoplasms ranging from low-grade melanocytomas to malignant melanomas.\textsuperscript{84,86} Those that are circumscribed may be radiographically indistinguishable from a meningioma. Leptomeningeal melanocytic neoplasms occur primarily in adults in the leptomeninges of the brainstem or spinal cord as solitary tumors.\textsuperscript{84,86} Grossly, they appear black or gray, and, depending on the grade, they may com-
press or invade the nervous system. Histologically, most melanocytomas are spindle cell neoplasms, although epithelioid cytology may dominate in some cases.85,86 Tumor cells may grow in sheets, nests, or fascicles. The round or oval nuclei are relatively uniform, sometimes with small nucleoli. In melanocytomas, mitotic activity may be minimal or up to 1 per 10 high-power fields (HPFs) with a MIB-1 labeling index of 0% to 2%. However, an intermediate-grade variant is now recognized that exhibits less nesting, more diffuse growth, minimal cytologic atypia, small nucleoli, a mitotic rate of 1 to 3 per 10 HPFs, and MIB-1 labeling of 2% to 4%.86 Recognition of this variant is important because of the reported increased risk of recurrence.86-89 In contrast, malignant leptomeningeal melanoma exhibits cytologic anaplasia, prominent nucleoli, and often brisk mitotic activity (2 to 15 per 10 HPFs), along with a MIB-1 labeling rate of 3 to 15 per 10 HPFs. Necrosis may be seen in occasional cases. Despite these features, the prognosis for malignant tumors appears to approximate that of uveal melanomas with less risk of metastasis and occasional apparent cure with complete resection.90-92

Melanocytic neoplasms must be differentiated from metastatic melanoma and other pigmented lesions that occur in the leptomeninges, including the rare melanotic schwannoma and its psammomatous variant. Metastatic melanoma is usually multifocal and typically involves the nervous system parenchyma. Epithelioid histology is more common; more bizarre forms, mitoses (7 to 35 per 10 HPFs), higher MIB-1 labeling (17% to 38%), and necrosis are seen.86 Pigmented schwannomas may occur intradurally at nerve root entry zones, but most commonly occur in the dorsal leptomeninges, although ventral development has been reported.85 Histologically, these tumors exhibit more elongated spindleled nuclei and cytoplasm but usually lack myxoid areas and Verocay bodies. Immunohistochemical demonstration of intercellular collagen IV, laminin, or reticulin surrounding cells may differentiate these tumors from melanocytomas, which exhibit only perifascicular deposition. Psammomatous melanotic schwannomas are associated with 60% of cases with Carney’s syndrome, exhibit psammoma bodies in more than 50% of cases, and usually occur in posterior nerve roots, skin, or the upper gastrointestinal tract (Fig 2).

Case Illustration

A 35-year-old woman presented with a 6-month history of decreased lower extremity sensation followed by progressive weakness, greater in the right leg than in the left leg, for 2 weeks. On the day of admission, she
was unable to stand or walk. MRI demonstrated a 3.0 × 2.0 × 1.0–cm homogeneously enhancing solid tumor ventral to the spinal cord at T4-T5 with compression of the spinal cord (Fig 13A). The mass had an enhancing dural tail. Radiographically, the mass was thought to be a meningioma.

The patient underwent posterior resection of the tumor. Intraoperative analysis suggested a low-to-intermediate-grade melanocytic neoplasm.

Grossly, the tumor was encapsulated, black, and soft. Microscopically, the tumor was composed of spindle cells with varying amounts of pigmentation throughout. Occasional melanophages were present. Tumor cells grew in a diffuse pattern populated by relatively uniform cells with small round and ovoid nuclei in areas with small nucleoli, but no significant cellular atypia or bizarre forms (Fig 13B and C). Up to 3 mitoses per 10 HPFs were seen focally (Fig 13D). Histology suggestive of a schwannoma or meningioma was not seen. The MIB-1 labeling rate was approximately 5%. Necrosis was not present. A diagnosis of melanocytoma, intermediate grade, was made.

Postoperatively, the patient was able to walk unassisted and experienced normal sensation in both lower extremities.

**INFECTION OF LESIONS MIMICKING MENINGIOMAS**

Rarely, infections limited to the leptomeninges may produce a dural-based mass that mimics a meningioma. In the United States, a tuberculosis from either mycobacterium tuberculosis or mycobacterium avium complex infection may produce such a lesion in an immunocompromised individual. This has recently been described in a patient with systemic lupus erythematosus and a mycobacterium avium complex infection. Histologically, due to the leptomeningeal reaction, such lesions may resemble a fibroblastic meningioma if the histiocyte component is not recognized and giant cells are lacking. Nonetheless, clinical features may suggest an infectious etiology in most cases.

**CONCLUSIONS**

A growing number of neoplastic and nonneoplastic dural lesions are now recognized and have expanded the differential diagnosis of meningiomas. Awareness of these lesions may facilitate their recognition intraoperatively and during postoperative study.

**REFERENCES**


