Extra-axial brain tumors

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Abstract

Extra-axial brain tumors are the most common adult intracranial neoplasms and encompass a broad spectrum of pathologic subtypes. Meningiomas are the most common extra-axial brain tumor (approximately one-third of all intracranial neoplasms) and typically present as slowly growing dural-based masses. Benign meningiomas are very common, and may occasionally be difficult to differentiate from more aggressive subtypes (i.e., atypical or malignant varieties) or other dural-based masses with more aggressive biologic behavior (e.g., hemangiopericytoma or dural-based metastases). Many neoplasms that typically affect the brain parenchyma (intra-axial), such as gliomas, may also present with primary or secondary extra-axial involvement. This chapter provides a general and concise overview of the common types of extra-axial tumors and their typical imaging features.

INTRODUCTION

The category of extra-axial brain tumors includes a large number of varied pathologic tumors grouped by their primarily extraparenchymal involvement, typically involving the meningeal layers of the brain. Extra-axial tumors are responsible for approximately half of all intracranial neoplasms in the USA (Dolecek et al., 2012). Many of these extra-axial tumors may involve anatomic sites outside the central nervous system (CNS) with similar imaging characteristics. Some of these neoplasms may also secondarily invade the brain parenchyma (e.g., meningioma, craniopharyngioma, metastatic disease).

EPIDEMIOLOGY

Based on the most recent data from the Central Brain Tumor Registry of the United States (2005–2009) (Dolecek et al., 2012), approximately 35% of all intracranial neoplasms (with more than 7.49 cases per 100 000/ year) are meningeal-based neoplasms (Table 15.1). Among these tumors, benign meningiomas represent approximately 35.5% of all intracranial neoplasms, followed next by glioblastomas (15.8%). Meningiomas also become more common with advancing age and they constitute the most common intracranial tumor in age groups older than 35 years (Dolecek et al., 2012). Tumors of the pituitary gland constitute 14.1% of all intracranial tumors (Dolecek et al., 2012). See Table 15.1 for further description of the current prevalence of different extra-axial tumors. Nerve sheath tumors are the most common extra-axial tumors during childhood (< 14 years of age), constituting approximately 4.9% of all intracranial neoplasms (Dolecek et al., 2012). Craniopharyngiomas have a slightly lower prevalence in this age group (4.1%) (Dolecek et al., 2012). Overall, vestibular schwannomas constitute the most common nerve sheath tumors (65% of these tumors), with an incidence of 1.1 per 100 000/ year, and are responsible for 5% of all intracranial neoplasms (Dolecek et al., 2012).

DIFFERENTIAL DIAGNOSIS OF EXTRA-AXIAL MASSES

The differential diagnosis of extra-axial intracranial masses varies depending on their location. The main neoplasms affecting the most common anatomic locations

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are summarized in Figures 15.1–15.5. The most frequent tumors involving the cerebellopontine angle cistern are schwannomas (typically arising from the vestibular nerves and extending into the internal auditory canals), followed by meningiomas or metastases (Fig. 15.1). Meningiomas, metastases, hemangiopericytomas, or dural-based lymphomas are responsible for most of the supratentorial extra-axial masses along the lateral cerebral convexities (Fig. 15.2). The differential diagnosis of pineal masses is broad, with pineal parenchymal tumors, germ cell tumors, and metastases being the most common neoplasms (Fig. 15.3). Intraventricular masses more often include ependymal and choroid plexus tumors, intraventricular meningiomas, central neurocytomas, or giant cell astrocytomas (Fig. 15.4). Some of these tumors have a neuroglial or neuronal histologic origin, commonly involve the brain parenchyma and are described in detail in the chapter dealing with intra-axial neoplasms. The most frequent sellar and suprasellar neoplasms are pituitary macroadenomas, meningiomas, and craniopharyngiomas (Fig. 15.5).

### MENINGEAL TUMORS

**Meningiomas**

Meningiomas are the most common intracranial and dural-based neoplasms. They are twice as frequent in women (Rees and Wen, 2010; Dolecek et al., 2012) and show progressive increase in frequency with age. Radiation exposure is a well-characterized risk factor for development of meningiomas, and hormonal receptors (estrogen and progesterone) in these tumors likely contribute to their growth. Syndromes associated with an increased risk and prevalence of meningiomas, typically...
**Fig. 15.2.** (A) Differential diagnosis of supratentorial dural-based masses. (B) Gadolinium-enhanced axial T1-weighted images of a right middle cranial fossa meningioma, (C) dural metastasis, and (D) hemangiopericytoma (with multifocal involvement of the right middle cranial fossa and left prepontine cistern).

**Fig. 15.3.** (A) Differential diagnosis of pineal masses. (B) Gadolinium-enhanced axial T1-weighted images of a pineal tumor of intermediate differentiation, (C) pineoblastoma, and (D) germinoma. Pineoblastomas are often quite large at the time of diagnosis.

**Fig. 15.4.** (A) Differential diagnosis of intraventricular masses. (B) Gadolinium-enhanced axial T1-weighted images of a central neurocytoma, (C) meningioma, and (D) choroid plexus papilloma.
presenting with multiple lesions, include neurofibromatosis type II, Gorlin syndrome, Cowden syndrome, and multiple endocrine neoplasia type 1 (MEN1), among other genetic conditions (Mawrin and Perry, 2010; Plotkin et al., 2013).

**PATHOLOGIC SUBTYPES**

Meningiomas originate from the arachnoidal cap cells in the arachnoid layer and they are currently subdivided into at least 16 neuropathologic subtypes (Louis et al., 2007; Mawrin and Perry, 2010) (Table 15.2). The vast majority, roughly 90%, are World Health Organization (WHO) grade I neoplasms. Based on their biologic behavior, these subtypes can be clustered into three different WHO grades (Table 15.2).

**Table 15.2**

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<td>Anaplastic (malignant)</td>
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**IMAGING FEATURES**

Meningiomas typically present as homogeneously enhancing dural-based masses, often exhibiting smooth tapering extensions along the adjacent dura mater (“dural tails”) (Figs 15.6 and 15.7). The most common locations for meningiomas include the cerebral convexity (up to 34% of cases), parafalcine region, sphenoid wings, parasellar compartment, posterior fossa (including the tentorium), and the olfactory groove (Drevelegas, 2005; Rockhill et al., 2007; Raizer, 2010). On plain radiographs and computed tomography (CT) studies, meningiomas often demonstrate hyperostosis of the underlying adjacent bone (18–50% of benign cases) and intratumoral calcifications (up to 25–27% of meningiomas) (Drevelegas, 2005; Rockhill et al., 2007) (Fig. 15.6). Focal osteolysis is unusual and only seen in less than 3% of meningiomas (Drevelegas, 2005). Most meningiomas appear as well-defined hemispheric or lobulated masses with increased attenuation on plain CT images. On magnetic resonance imaging (MRI), meningiomas show avid homogeneous gadolinium enhancement, and are T1 iso- to mildly hypointense and T2 iso- to mildly hyperintense (in relation to gray matter) (Drevelegas, 2005). The presence of cerebrospinal fluid (CSF) clefts (crenscietic between tumor and brain), white-matter buckling, or a signal void pseudocapsule (between tumor and brain) is very helpful in confirming an extra-axial location (Drevelegas, 2005). Gradient echo (T2*) sequences demonstrate the presence of

![Fig. 15.5. (A) Differential diagnosis of sellar/suprasellar masses. (B) Gadolinium-enhanced axial T1-weighted images of a pituitary macroadenoma (sellar/suprasellar heterogeneously enhancing mass), (C) meningioma (homogeneously enhancing suprasellar mass), and (D) craniopharyngioma (large predominantly cystic suprasellar lesion).](image-url)
intratumoral foci of susceptibility artifact compatible with mineralization and/or intratumoral vascularity. Dural tails are seen in approximately 60% of meningiomas (Drevelegas, 2005; Rockhill et al., 2007). The correlation between direct dural invasion around the meningioma and the presence of dural tails is controversial and some authors have attributed the presence of dural tails to reactive dural thickening, dural hyperemia, and/or tumoral invasion (Drevelegas, 2005; Rokni-Yazdi et al., 2009; Sotoudeh and Yazdi, 2010). The extent of parenchymal vasogenic edema, irregular contour, and local osteolysis may be helpful for the differentiation between benign and atypical/malignant meningiomas (Chernov et al., 2010) (Fig. 15.8). The role of diffusion-weighted imaging in the identification of atypical and malignant subtypes is very controversial, with some reports showing no significant difference, while other reports suggest lower apparent diffusion coefficient values in these subtypes (Toh et al., 2008; Santelli et al., 2010; Sanverdi et al., 2012; Yin et al., 2012; Watanabe et al., 2013) (Fig. 15.8).

Meningiomas are highly vascular tumors with intense intratumoral blushes on conventional cerebral angiography and markedly elevated relative cerebral blood volume (rCBV)/cerebral blood flow values in dynamic susceptibility contrast and arterial spin labeling MR perfusion (Kimura et al., 2006) (Fig. 15.8). Intratumoral rCBV values within angiomatous meningiomas and in the surrounding brain parenchyma around anaplastic meningiomas appear to be statistically higher than with other subtypes (Zhang et al., 2008a, b) (Fig. 15.8). MR spectroscopy can be helpful in the distinction of meningiomas from other extra-axial or dural-based masses due to the presence of alanine peaks in well-differentiated meningiomas (Demir et al., 2006, 2008a; Chernov et al., 2010). Meningiomas are considered “en plaque” when they appear as broad elongated flattened lesions infiltrating the dura and inner table of the skull, often associated with local hyperostosis (Kim et al., 1987; Drevelegas, 2005).
Intradiploic or intraosseous meningiomas are rare meningiomas that develop within the diploic space of the skull, often expanding the diploic space and infiltrating the pericranial spaces (Arana et al., 1996; Changhong et al., 1997). Secondary osseous involvement is also seen commonly with meningiomas (up to 26% of cases) (Gasparetto et al., 2007). Involved bone should be resected to prevent recurrence. Hyperostosis may occur with or without bone involvement. Meningiomas often exhibit prominent intraxial vasogenic edema in the surrounding brain parenchyma (up to 90% of cases) (Gasparetto et al., 2007), that appears to correlate with intratumoral vasculature (Pistolesi et al., 2003; Schmid et al., 2010) and also with a close apposition of the tumor to brain parenchyma. More extensive areas of peritumoral vasogenic edema have been described with meningothelial (Ide et al., 1994; Tamiya et al., 2001) and other uncommon subtypes (including angiomatous, microcystic, lymphoplasmacytoma-rich, and secretory meningiomas (Tamiya et al., 2001; Paek et al., 2005; Osawa et al., 2013), as well as with tumors with higher proliferation indexes (Osawa et al., 2013) (Fig. 15.7).

Additional features described with meningiomas include the presence of cystic changes within (intratumoral) and around these tumors (“acquired arachnoid cyst”) (Demir et al., 2007; Zhang et al., 2009), metaplasia with intratumoral fat (Sacher et al., 1985; Okamoto et al., 1996), and intracranial hemorrhage (Latchaw et al., 1981).

**Mesenchymal nonmeningothelial tumors**

This category includes a broad spectrum of mesenchymal pathologies, ranging from fat-containing tumors, chondroid or osseous-matrix tumors, to highly vascularized dural-based mesenchymal tumors with variable biologic behavior and aggressiveness. Hemangiopericytoma is the most common mesenchymal...
nonmeningothelial tumor. Sarcomas represent less than 0.1–0.2% of intracranial neoplasms (Louis et al., 2007) and can occur intracranially as a delayed sequela of craniofacial irradiation (particularly fibrosarcoma, malignant fibrous histiocytoma, chondrosarcoma, and osteosarcoma) (Louis et al., 2007). Primary sarcomas are rare and most of the intracranial sarcomas are secondary to metastatic CNS involvement (Al-Gahtany et al., 2003).

**FAT-CONTAINING TUMORS**

These tumors represent approximately 0.4% of intracranial neoplasms and include lipomas (Fig. 15.9), osteolipomas, angiolipomas, hibernomas, complex lipomatous lesions, epidural lipomatosis, and intracranial liposarcoma. Fat-containing tumors are, in general, benign (except for the rare intracranial liposarcoma) (Jabot et al., 2009). Osteolipomas are characteristically seen in the tuber cinereum region (Bognar et al., 2002; Moschopulos et al., 2006; Louis et al., 2007).

**HEMANGIOPERICYTOMA**

Hemangiopericytoma is classically described as a highly cellular and vascularized neoplasm, almost always dural-based, with a high recurrence/metastatic rate and variable pathologic grading (WHO II or III) (Uttley et al., 1995; Louis et al., 2007; Sibtain et al., 2007) (Fig. 15.10). There is recent debate regarding the origin of these tumors, and many soft-tissue pathologists call them “solitary fibrous tumors,” while neuropathologists retain the nosology of hemangiopericytoma (Penel et al., 2012). These tumors are uncommon (when compared to meningiomas) and represent approximately 0.4% of all primary intracranial tumors (Chiechi et al., 1996; Louis et al., 2007). Primary hemangiopericytomas commonly present as a solitary dural-based mass, more frequently in the occipital region near the sinus confluence (Louis et al., 2007) and typically without calcifications (Osborne et al., 1981; Akiyama et al., 2004) (Fig. 15.10). On CT studies, the presence of focal osteolysis of the adjacent bone (instead of hyperostosis) may be suggestive of hemangiopericytoma (Rusalleda et al., 1994;
Angiographic studies depict the marked vascularity of these tumors, occasionally with intratumoral “corkscrew-like” vessels (Akiyama et al., 2004).

OSTEOCARTILAGINOUS TUMORS
Most commonly, these tumors arise from the skull base osseous structures – especially the synchondroses – and extend intracranially, often involving the adjacent dura. They include chondrosarcoma and osteosarcoma.

VASCULAR TUMORS
The most common intracranial vascular tumors include benign hemangiomas (subdivided into capillary and cavernous, involving the skull and/or dura) (Perry et al., 1993; Ito et al., 2009; Joshi et al., 2009; Patnaik et al., 2012), hemangioendothelioma (intermediate grade) (Parajon and Vaquero, 2008), and more malignant subtypes, particularly angiosarcoma (Choi et al., 2008) and Kaposi sarcoma (Louis et al., 2007; Pantanowitz and Dezube, 2008). Intracranial involvement by these tumors is, in general, very rare.

OTHER MESENCHYMAL TUMORS
Meningeal sarcomatosis and Ewing sarcoma-peripheral primitive neuroectodermal tumor (EWS-pPNET) are included in this group. Both pathologies are very rare and present as diffuse leptomeningeal involvement (in the case of leptomeningeal sarcomatosis) (Grier and Yachnis, 2004; Uluc et al., 2004) and as a focal dural-based mass (for EWS-pPNET) (Antunes et al., 2001).

CHORDOMA
Chordoma appears to originate from notochordal remnants and is considered to have low-to-intermediate biologic aggressiveness (Gangadhar and Santhosh, 2012), although systemic hematogenous metastases may occur in more aggressive lesions. This tumor most commonly presents as an extra-axial and extradural lobulated, locally infiltrating and destructive mass in the sacrococcygeal (up to 60% of cases) and skull base (up to 40%) regions (Gangadhar and Santhosh, 2012) (Fig. 15.11). Rarely, chordoma can also present as a primarily intradural extra-axial mass (Roberti et al., 2007). CT may be the study of choice to demonstrate the osteolytic changes and presence of internal calcifications (Erdem et al., 2003). However, MRI is more sensitive for detection of tumoral invasion and extension into the adjacent soft tissues and intracranial spaces. The tumor is composed of physaliferous (having bubbles or vacuoles) cells with intracytoplasmic fluid, making them typically T2 hyperintense/T1 hypointense with variable enhancement (Erdem et al., 2003) (Fig. 15.11). Some chordomas can exhibit internal foci of T1 hyperintensity from the physaliferous cells that are characteristic of this neoplasm (Bonneville et al., 2006). Steady-state sequences such as CISS/FIESTA sequences are very helpful for better evaluation of intracranial extension. Previous studies have suggested that clival chordomas often originate more centrally while chondrosarcomas involving the skull base are more lateral, and most commonly develop near the petroclival fissure and foramen lacerum (Weber et al., 1994; Chapman et al., 2011). However, other studies have concluded that there are no specific imaging features that could help with their accurate diagnosis and there is significant anatomic overlap between these two skull base tumors (Pamir and Ozduman, 2006).

Fig. 15.11. Chordoma. (A) Gadolinium-enhanced axial, (C) coronal, and (D) sagittal T1-weighted images, as well as (B) axial T2-weighted image. Large irregular and infiltrative central skull base mass extending to the cavernous sinuses, medial aspects of the middle cranial fossa, and prepontine cistern. Biopsy confirmed the clinical diagnosis of chordoma.
TUMORS OF THE CRANIAL NERVES

Schwannomas

These are benign encapsulated neoplasms arising from Schwann cells that surround the peripheral portions of the cranial and spinal nerves. Schwannomas are the second most common extra-axial intracranial tumor and constitute approximately 8% of intracranial neoplasms (Drevelegas, 2005; Louis et al., 2007). These tumors are well-defined, typically encapsulated, tumors (Drevelegas, 2005) (Fig. 15.12) that may often be dissected free from the parent nerve. There are two histologic components: Dense Antoni A and looser Antoni B tissue patterns. Schwannomas most commonly affect the inferior vestibular division of the eighth nerve and, less often, the fifth cranial nerve (Fig. 15.12). Because they grow slowly, secondary arachnoid cysts are often seen around large tumors. In addition, intratumoral benign cystic degeneration may occur in larger tumors. Signal intensity varies depending on the relative proportions of tissue, with the predominantly Antoni type A lesions appearing more homogeneous and hypointense of T2-weighted images, while those with more Antoni B tissue are T2 hyperintense and are more heterogeneous, often with internal foci of cystic change or hemorrhage (Drevelegas, 2005). Schwannomas, particularly in the cerebellopontine-angle cistern region, often have associated arachnoid cysts. Dural tails are also occasionally seen with schwannomas, and they are the second most common tumor to show this feature, after meningiomas. The presence of calcifications within the tumor is rare and statistically argues against the diagnosis of schwannoma (Drevelegas, 2005).

Neurofibromas

These WHO grade I neoplasms typically involve the spine and, sporadically, skull base in patients with neurofibromatosis type 1, but they only rarely arise intracranially from cranial nerves (Rodriguez and Berthrong, 1966; Drevelegas, 2005).

LYMPHOMAS AND HEMATOPOIETIC NEOPLASMS

Lymphoma

Primary CNS lymphomas (PCNSLs) are defined as extranodal lymphomas involving CNS structures without evidence of extension outside the CNS (different from the secondary CNS involvement seen in cases of peripheral lymphoma). Most of the PCNSLs are classified as diffuse large B-cell lymphomas. PCNSLs involving immunocompetent individuals are more frequent in older male patients but are seen in younger immunocompromised individuals (e.g., posttransplant, acquired immunodeficiency syndrome (AIDS), inherited immunodeficiencies) (Louis et al., 2007). PCNSL presents more commonly with intraparenchymal involvement, often as an ill-defined hyperdense mass on CT (due to high cell density and high nuclear-to-cytoplasm ratio) involving the subependymal and periventricular regions or corpus callosum. Meningeal involvement by primary CNS lymphoma is occasionally seen (up to 30–40% of cases) (Louis et al., 2007). Primary dural involvement is very unusual and pathologically is usually classified as low-grade marginal-zone B-cell type (Mneimneh et al., 2013). Dural-based lymphoma typically presents as a homogeneously enhancing dural-based mass with abnormal restricted diffusion, somewhat similar to a meningioma (Iwamoto and Abrey, 2006; Kulkarni et al., 2012; Tandon et al., 2012) (Fig. 15.13). There have been also reports of dural lymphoma confused with epidual hematomas (Iaccarino et al., 2013). Steroid treatment has a dramatic effect in these cases and lesions can disappear very rapidly after treatment (Louis et al., 2007). For this reason, they are often called “ghost tumors.”

Fig. 15.12. Bilateral schwannomas. (A) Gadolinium-enhanced axial and (B) coronal T1-weighted images; (C) axial T2-weighted image. Multiple large bilateral enhancement masses along the trajectories of multiple cranial nerves in a patient with neurofibromatosis type II, compatible with schwannomas.
Plasmacytomas

These tumors can develop in patients with multiple myeloma as intra- or extraosseous lesions and rarely involve the CNS (Cerase et al., 2008). Plasmacytomas can present as an intraosseous masses involving the central skull base with secondary intracranial involvement or as a dural-based mass in cases of solitary intracranial plasmacytomas (Cerase et al., 2008; Cao et al., 2010). Most of the solitary intracranial plasmacytomas occur in older patients (fifth and sixth decades) and are iso- to hypodense on CT (Cerase et al., 2008; Cao et al., 2010). They show nonspecific MRI findings, variable T1/T2 signal, variable enhancement, and abnormal restricted diffusion (Cerase et al., 2008; Cao et al., 2010). Their imaging features can very similar to other intracranial pathologies, such as meningiomas, lymphomas, metastases, and nonneoplastic processes (Cerase et al., 2008).

Myeloid sarcoma

Previously called chloroma or granulocytic sarcoma, this is a rare tumor composed of immature granulocytic cells and seen in patients with acute myeloid leukemia (or other myeloproliferative disorders) that can involve intracranial structures in 2–3% of cases (Hakyemez et al., 2007; Grier et al., 2008). The intracranial cases more often present as dural-based masses and are considered to originate from the bone marrow in the skull, extending intracranially (Hakyemez et al., 2007). Differentiation from lymphoma can be difficult, even in histologic analysis (Hakyemez et al., 2007; Grier et al., 2008). Myeloid sarcomas are typically iso- to hypointense in T1- and T2-weighted images, with homogeneous enhancement and often with abnormal restricted diffusivity (Hakyemez et al., 2007).

GERM CELL TUMORS

Germinomas (formerly called “atypical teratoma,” and identical to testicular seminoma and ovarian dysgerminoma) are rare tumors, probably arising from totipotent germ cells. These tumors constitute less than 3% of all primary intracranial neoplasms and usually present in or near the midline, in children or young adults (Takeshima et al., 2012; Wang et al., 2010). These neoplasms can be extra- or intra-axial and often involve the suprasellar and pineal regions (Wang et al., 2010). Germinomas often demonstrate secondary leptomeningeal dissemination following the CSF pathways (up to 10% of cases) but rarely can also disseminate as primarily dural-based lesions (Takeshima et al., 2012). Teratomas, which also arise from pluripotent cells, more often present as midline lesions, often located in the pineal and suprasellar regions (Fig. 15.14). Teratomas may show heterogeneous components, including sebaceous lipid, watery fluid, and solid enhancing tissues.

Germ cell tumors are subdivided in several histopathologic subtypes, including germinoma, teratoma, mature teratoma, yolk sac tumor, embryonal carcinoma, and choriocarcinoma (Louis et al., 2007).

The CT and MR features of germ cell tumors are predominantly nonspecific, particularly with extra-axial involvement. Nonteratomatous germ cell tumors often are T1/T2 iso- to hypointense and show homogeneous enhancement (Wang et al., 2010). Because germinomas are typically hypercellular and often contain a lymphocytic component (probably reactive), they may be homogeneously hyperattenuating on plain CT and have restricted diffusivity on MRI (Ogiwara et al., 2015). These can be very useful differentiating imaging features. Calcification and necrosis can be seen occasionally (Wang et al., 2010). Teratomas classically
present as mixed lesions with cystic, calcified, and fatty components (Fig. 15.14). Neoplasms with chorocarcinomatous or syncitiotrophoblastic components often show intratumoral hemorrhage (Louis et al., 2007) that will cause T1 shortening. Leptomeningeal and dural metastases of germ cell tumors are difficult to differentiate from other neoplastic conditions.

**TUMORS OF THE SELlar REGION**

**Craniopharyngioma**

These are intracranial cystic tumors that are considered benign (WHO grade I), typically suprasellar in location, and may be derived from Rathke’s pouch remnants. They represent up to 4.6% of all intracranial tumors (5–10% in children) (Louis et al., 2007). Craniopharyngiomas demonstrate a bimodal distribution with two age peaks: one during the first and second decades and a second during the fourth and fifth decades. Despite their benign pathologic designation, craniopharyngiomas often recur locally because of adherence to surrounding brain and depending on the extent of surgical resection. There are two histopathologic subtypes: adamantinomatous (mostly younger patients) and papillary craniopharyngiomas (mostly older patients).

Adamantinomatous craniopharyngiomas are typically mixed cystic and solid masses with internal or ring calcifications and occasional hemorrhagic foci (Fig. 15.15A–C). Papillary craniopharyngiomas are more solid and intratumoral calcifications are seen less frequently but may be present in this subtype (Fig. 15.15D–F). Most of the papillary subtype cases have been reported in adults. Vascular encasement, lobulated contour, and
T1 hyperintense cysts are more often seen with the adamantinomatous subtype, while hypointense cysts and a predominantly solid appearance are more commonly seen with the papillary tumors (Sartoretti-Schefer et al., 1997) (Fig. 15.15).

Pituitary adenomas

Pituitary adenomas represent the most frequent intrasellar masses and are typically classified based on their size into micro- and macroadenomas (< or > 10 mm respectively). The vast majority of pituitary adenomas are considered functional (up to 75%) (Kumar et al., 2007), and, the most common functional subtype are the prolactin-secreting adenomas. Adenomas can manifest clinically due to endocrinologic disturbances (which vary by hormone production) or due to local mass effect (usually with visual disturbance due to chiasmatic compression or other cranial nerve palsies). Microadenomas may appear as areas of relatively decreased enhancement within the pituitary gland during early postcontrast images (dynamic scanning), often hypointense on precontrast T1-weighted images (Kumar et al., 2007). Macroadenomas, by definition, will expand the sella and/or extend into the suprasellar cistern. They often appear as T1 isointense, T2 hyperintense, with heterogeneous enhancement, and often exhibit intratranonal necrotic/cystic changes (in up to 18% of macroadenomas) (Kumar et al., 2007) (Fig. 15.16). Macroadenomas frequently show intratumoral hemorrhage (up to 30% of cases) with fluid levels. Absence of vascular narrowing of the adjacent cavernous internal carotid arteries is a frequently described sign favoring a macroadenoma (Fig. 15.16).

Rathke’s cleft cysts

These cystic lesions are also originated from remnants of the Rathke’s pouch and are more commonly intrasellar, located at the level of the pars intermedia (but can also be occasionally present along the infundibular stalk and suprasellar region). Their T1 and T2-weighted MR appearance is variable and depends on the amount of intracystic debris/proteinaceous content. Mild peripheral enhancement can be present and intracystic nodules have been reported in the literature as a useful imaging finding to differentiate them from other cystic pathologies (Binning et al., 2005).

Metastatic Extra-Axial Tumors

Dural metastases develop by direct/contiguous extension or via hematogenous seeding. Pelvic malignancies may disseminate via the Batson retrovertebral plexus, which communicates intracranially with the retroclival veins. The most prevalent primary neoplasms presenting as dural-based masses in adults include breast cancer, prostate cancer (Fig. 15.17), lung adenocarcinoma, renal cell carcinoma, and multiple myeloma (Maroldi et al., 2005). For children, neuroblastoma and sarcoma represent the most common tumors presenting as dural-based metastases. Leptomeningeal metastases can also have a hematogenous mechanism (arterial seeding or venous hematogenous access) (Maroldi et al., 2005).

Other Nonneoplastic Cystic Lesions

Dermoid and epidermoid cysts

These are considered to be slowly growing developmental pathologies, representing up to 1% of all intracranial tumors (Gelabert-Gonzalez, 1998). Dermoid and epidermoid cysts are thought to be the result of abnormal inclusion of ectodermal tissue intracranially during development (Orakcioğlu et al., 2008), probably from incomplete separation of the surface ectoderm from...
the neural tube or related to the invaginations of the otic capsule or during development of the eye and orbit. Dermoid cysts are generally benign sebaceous lipid-containing lesions, often located near the midline, that can occasionally rupture, producing chemical meningitis, vasospasm, and hydrocephalus (Orakcioglu et al., 2008). They are often T1 hyperintense, T2 hypointense but occasionally can show CSF-like signal similar to epidermoid cysts (Orakcioglu et al., 2008). Dermoid and epidermoid cysts increase in size due to desquamation and secretions related to the epithelial components lining the cysts. Epidermoid cysts often present as heterogeneously fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted imaging (DWI) hyperintense extra-axial lesions involving the basilar cisterns (Hakyemez et al., 2005) and in the cerebellopontine angle (Fig. 15.18). Atypical appearances have been reported in up to 5.6% of epidermoid cysts (Ren et al., 2012) due to intralesional hemorrhage.

**ARACHNOID CYSTS**

Arachnoid cysts are considered nonneoplastic intra-arachnoid cysts constituting approximately 1% of all intracranial mass-like lesions (Cincu et al., 2007), often involving the middle cranial fossa. These cysts are likely developmental in etiology and related to focal splitting/duplication of the arachnoid membrane (Cincu et al., 2007). Arachnoid cysts follow the appearance of the cerebrospinal fluid in CT and MR images, with variable local mass effect on the adjacent structures (Fig. 15.19).

**NEURENTERIC CYSTS AND ECCHORDOSIS PHYSALIPHORA**

Neurenteric cysts are rare cystic endodermal-derived developmental lesions typically located in the spinal canal and posterior fossa (most commonly within the cerebellopontine angle and prepontine cisterns (Preece et al., 2006). These cysts are typically CSF isointense on T1- and T2-weighted images, without internal enhancement, and often show increased FLAIR signal and variable appearance of DW images (Preece et al., 2006) (Fig. 15.20). Ecchordosis physaliphora are also rare cystic endodermal-derived developmental pathologies, typically located in the prepontine cistern and often associated with an osseous spicule or small stalk in the adjacent clivus (Mehnert et al., 2004) (Fig. 15.21).

**CONCLUSION**

There are many different types of intracranial neoplasms and nonneoplastic conditions that can present as extra-axial intracranial masses. The differential diagnosis can often be narrowed if specific imaging findings (such as focal hyperostosis for meningiomas or presence of abnormal DWI hyperintensity in cases of epidermoid cysts) are present. The ultimate diagnosis often requires pathologic sampling or long-term follow-up to confirm stability. The challenge of the radiologic assessment of these lesions is to develop new, more accurate noninvasive imaging techniques, decreasing the need for pathologic sampling and accelerating their treatment.

**DISCLAIMER**

The views and opinions expressed herein are those of the authors and should not be construed as being official, nor as representing the Uniformed Services University of the Health Sciences, the Department of Defense, nor the federal government.
Fig. 15.18. Epidermoid cyst. (A) Axial fluid-attenuated inversion recovery (FLAIR), (B) axial T2-weighted, (C) axial, and (D) coronal postcontrast T1-weighted images, as well as axial diffusion-weighted (B = 1000) (E) and axial apparent diffusion coefficient (ADC) (F) images. There is a large T2 hyperintense mass-like lesion in the basilar cisterns and medial aspect of the left temporal lobe, producing local mass effect, without abnormal nodular enhancement but with increased diffusion-weighted imaging signal (E) (isointense on the ADC map), compatible with a large epidermoid cyst.

Fig. 15.19. Arachnoid cyst. Axial, coronal, and sagittal T2-weighted images. There is a large arachnoid cyst (isointense to cerebrospinal fluid on all sequences) centered on the basilar and left middle cerebral artery cisterns.
Fig. 15.20. Neurenteric cyst. (A) Sagittal and (B) axial postcontrast T1-weighted; (C) axial T2-weighted and (D) axial apparent diffusion coefficient images. There is an intrinsically T1 hyperintense tubular structure in the preoptic and premedullary cisterns, compatible with a neurenteric cyst (confirmed by pathology).

Fig. 15.21. Ecchordosis physaliphora. (A) Axial T1 – post. (B) axial computed tomography – bone window. (C) axial CISS, and (D) sagittal reformatted CISS images. A nonenhancing cystic structure in the preoptic cistern associated with a small midline spicule along the clivus is compatible with ecchordosis physaliphora.

REFERENCES


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