Extra-Axial Neoplasms, Cysts and Tumor-Like Lesions

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Introduction

Extra-axial neoplasms, cysts and tumor-like lesions account for approximately one-third of all intracranial primary neoplasms in adults and about one-quarter of brain tumors in children. The differential diagnosis of an extra-axial mass varies significantly with both patient age and geographic location [1]. In this article we consider the pathology and imaging appearance of intracranial extra-axial masses, subdividing them into supra- and infratentorial lesions by age group. Their more specific differential diagnosis by anatomic region is illustrated graphically. Special attention is given to sellar and parasellar lesions.

Supratentorial Extra-axial Tumors in Adults

Adult supratentorial extra-axial neoplasms occur in several general locations (Fig. 1): (1) The sella and parasellar region; (2) skull and meninges; (3) CSF spaces (subarachnoid cisterns and ventricles); and (4) the pineal region. In this chapter we focus on lesions that involve the sellar region and the skull and its linings.

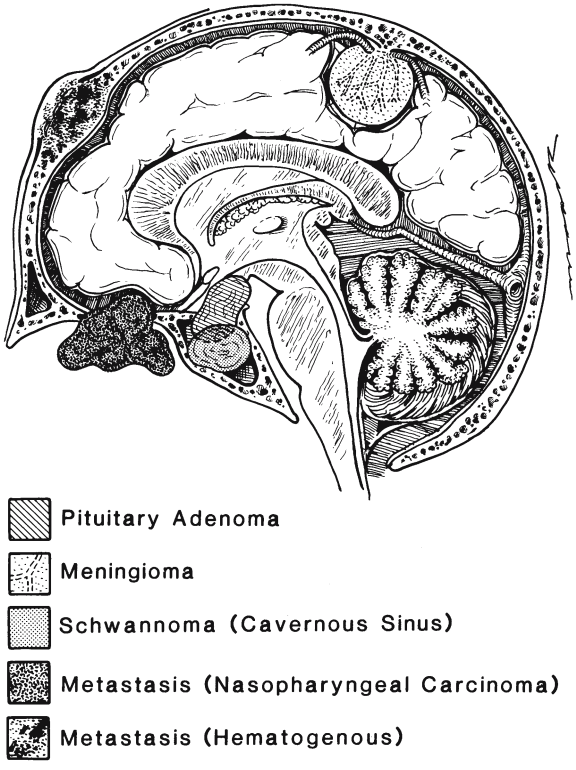

Fig. 1. Sagittal anatomic diagram depicts common locations of supratentorial extra-axial tumors in adults, showing the most important lesions found in each anatomic area

Sella and Parasellar Region

A variety of neoplasms and tumorlike lesions affect the sella and parasellar region. The differential diagnosis of a mass in this anatomically complex area is extensive, with more than thirty different pathologic entities reported in this area. However, just five lesions ("The Big Five") account for the vast majority of masses that occur in and around the sella (Table 1).

Three neoplasms account for the vast majority of sellar and parasellar tumors in adults: (1) pituitary adenoma; (2) meningioma; and (3) schwannoma (in the cavernous sinus). Less common lesions in adults include craniopharyngioma and metastasis. A spectrum of nonneoplastic cysts also can be found in this location [2].

Dividing lesions into intra-, supra-, and parasellar masses is helpful in establishing a more limited diagnosis.

Table 1. Sellar/parasellar masses in adults

Intrasellar
Most common
Microadenoma
Hyperplasia (physiologic, end-organ failure)
Less common
Rathke cleft cyst
Other nonneoplastic cysts
Metastasis to pituitary gland
Rare
Craniopharyngioma
Suprasellar
Most common
Pituitary macroadenoma
Meningioma
Aneurysm
Less common
Astrocytoma
Rathke cleft cyst
Other nonneoplastic cysts (parasitic, arachnoid, epidermoid, dermoid)
Metastasis (infundibular stalk, hypothalamus)
Lymphoma
Hypophysitis
Sarcoid
Lipoma
Rare
Craniopharyngioma
Germinoma
Parasellar (cavernous sinus)
Most common
Meningioma
Schwannoma
Metastasis
Less common
Lymphoma
Aneurysm
Rare
Chordoma
Osteocartilaginous tumor
Infrasellar (basisphenoid)
Most common
Metastasis (hematogenous)
Common
Metastasis (nasopharyngeal carcinoma)
Inflammatory disease (osteomyelitis)
Less common
Plasmacytoma
Lymphoma
Invasive adenoma
Rare
Nonfungal granuloma
Mucocele

Intrasellar Masses

Pituitary microadenomas (by definition, these are 10 mm or less in diameter) and *nonneoplastic cysts* are the most common intrasellar masses. Other lesions (e.g., craniopharyngioma, meningioma, and metastasis) are rare. Dynamic contrast-enhanced MR scans are the best imaging technique for identifying intrasellar masses. Most microadenomas enhance more slowly than the surrounding normal pituitary gland. They are seen as an area of relative hypointensity compared to the intensely enhancing pituitary gland and cavernous sinus.

Suprasellar Masses

Overall, pituitary adenoma is the most common sellar region mass, accounting for at least half of all tumors in this location. As they enlarge, small pituitary adenomas often grow upwards through the diaphragma sellae and acquire a figure of eight appearance. On MR scans, these *pituitary macroadenomas* demonstrate variable signal intensity and strong but heterogenous enhancement following contrast administration. Hemorrhage and cyst formation are not uncommon.

At surgery, microscopic dural invasion is identified with the vast majority of macroadenomas. However, identifying this finding preoperatively is difficult on imaging studies alone. Occasionally macroadenomas attain striking size, extending into the anterior and middle cranial fossae. Some adenomas exhibit frank invasion of the skull base. Despite their aggressive appearance, nearly all of these "invasive adenomas" are histologically benign; true pituitary carcinoma is exceptionally rare [3].

Meningioma is the second most common suprasellar tumor in adults (see below). It may arise from the diaphragma sellae, tuberculum, dorsum, or cavernous sinus and secondarily involve the sella itself.

Craniopharyngiomas are most commonly seen in children. However, a second peak occurs in adults between the fourth and sixth decades. *Metastases* in the

sellar/juxtasellar region most commonly involve the skull base and cavernous sinus (see below). Hematogenous metastases to the pituitary gland may cause an intrasellar mass; metastases that involve the infundibular stalk or hypothalamus are an uncommon but important cause of a suprasellar mass.

Lymphoma (almost always non-Hodgkin type) is another tumor that has a predilection for the sellar region. Any site (pituitary gland, infundibular stalk, hypothalamus, cavernous sinus) may be involved.

A variety of nonneoplastic cysts and tumor-like lesions involve the suprasellar region. These include *Rathke cleft cyst*, *dermoid cyst*, *epidermoid inclusion cyst* and *arachnoid cyst*.

Parasellar Masses

Schwannomas are comparatively rare in intracranial locations other than the cerebellopontine angle cistern (see below). The most common supratentorial site is the cavernous sinus; the trigeminal (cranial nerve V) nerve is most commonly involved. *Meningiomas* and *metastases* often also involve the cavernous sinus.

Skull and Meninges

Meningioma

Meningioma is the second most common primary brain tumor in adults, representing between 15% and 20% of these neoplasms. Most *typical meningiomas* are slowly growing neoplasms that are histologically benign although their location at the skull base may make complete resection difficult.

Three microscopic subtypes of *typical meningioma* are recognized: (1) A meningothelial (or syncytial) type; (2) a fibrous (fibroblastic) type; and (3) a transitional form. *Atypical and anaplastic (malignant) meningiomas* are biologically more aggressive tumors that account for 10%-15% of cases. The rare tumor formerly known as "angioblastic meningioma" is now designated as *hemangiopericytoma of the meninges* [4].

More than three-quarters of meningiomas are supratentorial; the most common locations are the convexity and falx cerebri. Other favored sites include the sphenoid ridges, olfactory grooves, parasellar region (tuberculum, dorsum, diaphragma sellae and the cavernous sinus), and cerebellopontine angles (see below).

Most meningiomas arise from arachnoid cap cells and are firmly attached to the dura. A circumferential collar of reactive thickening accounts for the "dural tail sign" that is seen in approximately 60% of meningiomas. Meningiomas typically invaginate into the brain, displacing the cortex and creating a surrounding cleft of cerebrospinal fluid and vessels that is easily identified on MR imaging.

Metastases

Metastases account for between one-quarter and one-third of all brain tumors in adults. Several varieties are recognized: (1) direct geographic extension of local tumors; (2) hematogenous metastasis; and (3) CSF dissemination from extra- or intracranial neoplasms.

Common regional malignant tumors that may spread directly to the adjacent skull and dura include *squamous cell carcinoma* (of the nasopharynx and paranasal sinuses) and *adenoid cystic carcinomas* (of the salivary or mucous glands). *Scalp carcinomas* (e.g., basal cell carcinoma) occasionally extend through the underlying calvarium and may even involve the dura.

Hematogenous metastases from extracranial primary tumors such as carcinoma of the lung or breast most commonly involve the brain parenchyma. The calvarial vault, central skull base, and dura are also frequently affected.

Supratentorial Extra-axial Tumors in Children

Extra-axial tumors in children are less common than their intra-axial counterparts. The most common location of a supratentorial extra-axial mass in a child is the sella, followed by the pineal region and ventricles (Fig. 2) [5]. The skull and meninges are a less common but nevertheless important site.

Sella and Parasellar Region

Pituitary adenoma, the most common sellar lesion in adults, is very rare in children. Only two sellar lesions commonly occur in the pediatric age group; one (craniopharyngioma) is truly an extra-axial mass whereas the other (an exophytic astrocytoma of the optic chiasm or hypothalamus) is considered an intra-axial neoplasm.

As with adults, dividing lesions into intra-, supra-, and parasellar locations is helpful in establishing an appropriate differential diagnosis (Table 2).

Intrasellar Masses

Craniopharyngiomas that are exclusively intrasellar are rare. Nonneoplastic cysts (such as a *Rathke cleft cyst*) occur but are relatively uncommon in this age group.

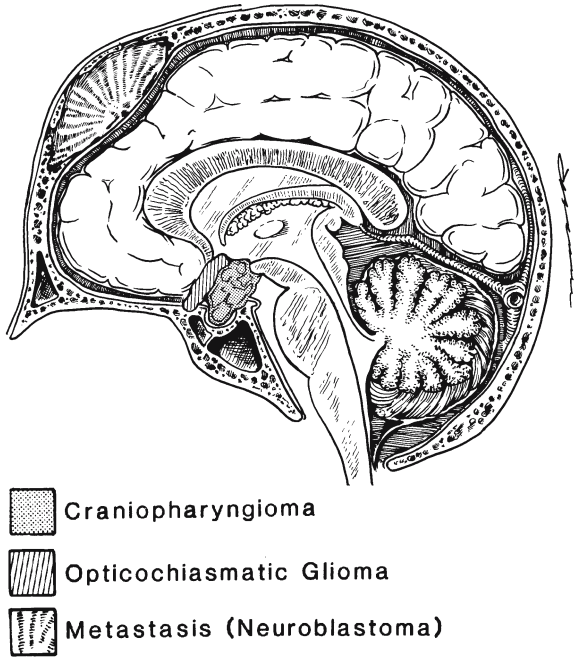

Fig. 2. Sagittal anatomic diagram depicts supratentorial extra-axial tumors in children, showing the important lesions found in the sella/ parasellar region as well as the calvarial vault

Table 2. Supratentorial extra-axial masses in children

Sellar/parasellar
Common
Craniopharyngioma
Astrocytoma (exophytic from chiasm, hypothalamus)
Less common
Germinoma
Lymphoproliferative disorder
Rare
Langerhans' cell histiocytosis (eosinophilic granuloma)
Hypothalamic hamartoma
Skull/meninges
Common
Eosinophilic granuloma
Less common
Metastasis (neuroblastoma)
Lymphoproliferative disorder
Rare
Chordoma

Suprasellar Masses

Craniopharyngiomas are the most common nonglial brain tumor in children. Approximately 80% of craniopharyngiomas are suprasellar, accounting for almost half of all pediatric tumors in this location. Craniopharyngiomas are derived from fetal ectodermal anlage, probably as incomplete involution of an upward evagination of the primitive oral cavity called the stomatodeum, also known as the craniopharyngeal duct (Rathke's pouch).

Between 85% and 90% of all craniopharyngiomas are cystic or are composed of both solid and cystic components. Two histologic types are recognized, namely, the so-called "adamantinomatous" craniopharyngioma (the most common type in children and usually cystic or mixed) and the "papillary" craniopharyngioma (the dominant variant in adults, and more often solid). Most craniopharyngiomas are calcified; their signal intensity on MR scan varies.

Although they are histologically benign tumors and most are very slow-growing lesions, craniopharyngiomas do tend to infiltrate surrounding structures. Malignant transformation is exceedingly rare.

Exophytic astrocytoma of the optic chiasm, optic nerves, and hypothalamus is the second most common suprasellar neoplasm in children. Other less common tumors in children include *germinoma* and *lymphoproliferative disorders*. Nonneoplastic masses in this area include the same spectrum of developmental cysts observed in adults (see above) as well as uncommon lesions such as *Langerhans' cell histiocytosis* and *hypothalamic (tuber cinereum) hamartoma*.

Other less common tumors such as *germinoma* also occur in the suprasellar region.

Skull and Meninges

In the absence of neurofibromatosis type 2 (NF-2), meningioma (the most common adult neoplasm in this location) is rare in children. In contrast to adults, metastases are also rare. The exceptions are *metastatic neuroblastoma* and *lymphoproliferative disorders*. *Eosinophilic granuloma* is a common nonneoplastic calvarial or skull base lesion that may show extensive bony destruction, occasionally mimicking infiltrating tumor.

Chordomas represent less than 1% of all intracranial tumors. Chordomas are derived from notochordal remnants. Their preferred intracranial location is the clivus; the cavernous sinus is a less common site. Although chordomas can occur at any age, they rarely affect individuals less than 20 years of age [3].

Infratentorial Extra-axial Tumors in Adults

Most infratentorial tumors in adults are extra-axial and their most common site is the cerebellopontine angle cistern (Fig. 3). The jugular foramen, foramen magnum, and clivus are also relatively common sites, whereas the occipital squamae are rarely involved by tumors.

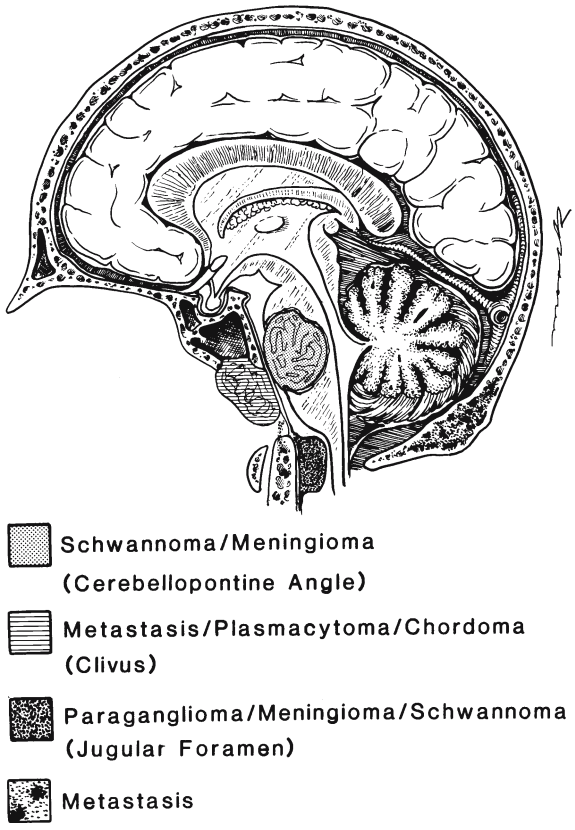

Fig. 3. Sagittal ana-tomic diagram depicts common infratentorial extra-axial tumors in adults, showing the important lesions found in specific locations within the posterior fossa

Table 3. Infratentorial extra-axial masses in adults

Cerebellopontine angle/temporal bone
Most common
Vestibulocochlear ("acoustic") schwannoma
Common
Meningioma
Vascular ectasia (vertebrobasilar)
Less common
Aneurysm
Other schwannomas (e.g., trigeminal)
Nonneoplastic cyst (parasitic, epidermoid, arachnoid)
Metastasis
Paraganglioma
Rare
Cholesterol granuloma
Malignant external otitis
Sarcoid
Idiopathic invasive cranial pachymeningitis
Jugular foramen
Most common
Meningioma
Common
Schwannoma
Paraganglioma
Metastasis
"Pseudotumor" (asymmetric jugular bulb; turbulent flow)
Clivus/foramen magnum
Most common
Metastasis
Common
Meningioma
Less common
Plasmacytoma
Invasive adenoma
Paraganglioma
Rare
Chordoma
Osteocartilaginous tumors

Cerebellopontine Angle

Most cerebellopontine angle (CPA) masses arise as extensions from lesions in the temporal bone (particularly the internal auditory canal). Others occur as primary lesions of the CPA itself (Table 3).

Temporal Bone Tumors

Schwannoma is the most common posterior fossa mass in adults. Intracranial schwannomas account for approximately 8% of all primary intracranial tumors. The vestibulocochlear nerve (cranial nerve VIII; "acoustic schwannoma") is the site of origin in approximately 80%-90% of these tumors. Most arise from the vestibular portion of the nerve.

Small vestibulocochlear schwannomas are initially intracanalicular. As they expand, they extend through the porus acusticus into the CPA cistern. An "ice cream on a cone" appearance on imaging studies is typical. Most schwannomas are isointense with brain on T1-weighted scans and hyperintense on T2-weighted scans. Strong contrast enhancement is typical. Intratumoral cysts and associated arachnoid cysts are common.

Metastases and *endolymphatic duct tumors* may involve the petrous temporal bone and cause a secondary CPA mass.

Cisternal Neoplasms

Meningioma is the second most common posterior fossa mass in adults. Most arise along the petrous temporal bone and exhibit a flat broad-based attachment to the underlying dura. Larger lesions may invaginate into the adjacent cerebellum and usually appear separated from it by an identifiable CSF-vascular cleft. Meningiomas sometimes extend into the internal auditory canal al-

though nonneoplastic dural reaction also occurs in this area.

Less common CPA cisternal masses in adults include *epidermoid, dermoid,* and *arachnoid cysts. Tumor mimics* such as malignant otitis, mastoiditis, hemangioma, and idiopathic invasive cranial pachymeningitis may cause bone destruction with thickening of the adjacent dura.

Jugular Foramen

Benign Neoplasms

Several tumors may originate within the jugular foramen and extend intracranially. A *paraganglioma (chemodectoma)* is a benign neoplasm that arises from parasympathetic ganglia adjacent to the jugular bulb adventitia. Paragangliomas are highly vascular tumors that enhance strongly but heterogeneously with contrast administration. "Flow voids" caused by high velocity signal loss often give these lesions a "salt and pepper" appearance on MR scans. Paragangliomas may extend locally into the basal cisterns and cause an extra-axial mass effect. Focal (e.g., jugular spine) as well as more striking, diffuse bone erosion may be present.

Schwannomas of glossopharyngeal, vagus, and spinal accessory nerves (cranial nerves IX, X, and XI) are much less common than their counterparts in the internal auditory canal. When present, schwannomas cause smooth, scalloped expansion of the jugular foramen. Flow voids are uncommon and contrast enhancement is comparatively homogeneous.

Jugular foramen "pseudotumors" (e.g., asymmetric jugular bulb) should not be confused with true masses in this location.

Malignant Neoplasms

Hematogenous *metastases* to the jugular foramen are uncommon. When they occur, irregular permeative bone destruction with an adjacent soft tissue mass is typical.

Clivus and Foramen Magnum

Clivus Masses

Any aggressive or slowly-growing clival tumor may break through the overlying cortex and extend extradurally. In adults, tumors that commonly affect the clivus include *chordoma*, *meningioma, plasmacytoma, metastasis* (either hematogenous or direct spread of regional extracranial neoplasms such as nasopharyngeal squamous cell carcinoma), and *invasive pituitary adenoma* (see above). Because it is formed from enchondral bone, the central skull base may also give rise to a spectrum of *osteocartilaginous tumors* (e.g., enchondroma) that may cause an extra-axial mass effect.

Foramen Magnum Masses

The differential diagnosis of a foramen magnum mass in an adult varies with specific location. Anterior intradural extramedullary masses include *meningioma*, *schwannoma*, and *glomus jugulare tumor*. Less commonly, *nonneoplastic cysts* (such as epidermoid inclusion cyst, arachnoid cyst, neurenteric cyst) are found in this location. Nonneoplastic masses include craniovertebral junction anomalies and proliferative arthropathies with associated degenerative disease (e.g., rheumatoid pannus).

Tumors that are located posterior to the medulla are less common. In adults, *subependymoma* (extending inferiorly from the fourth ventricle) and *metastasis* are the most common neoplasms that occur here. Overall, tonsillar herniation (congenital or acquired) is the most common cause of a mass behind the cervicomedullary junction.

Infratentorial Extra-axial Masses in Children

Most posterior fossa neoplasms in children are intra-axial, arising either from the cerebellum or brainstem (e.g., pilocytic and fibrillary astrocytoma) or from the fourth ventricle and its adjacent structures (e.g., medulloblastoma and ependymoma). Primary infratentorial extra-axial tumors are rare in children. Occasionally an *exophytic astrocytoma* or lateral extension from an *ependymoma* causes a significant extra-axial mass effect (Fig. 4).

Although sporadic cases do occur, *schwannoma* and *meningioma* are rare in the absence of neurofibromatosis type 2 (NF-2). Uncommon tumors such as *choroid plexus papilloma* and nonneoplastic cysts such as an *arachnoid, epidermoid, neurenteric* or *dermoid cyst* are occasionally identified as posterior fossa extra-axial masses in children [6].

■ **Note:** The author retains the copyright in Figs. 1–4.

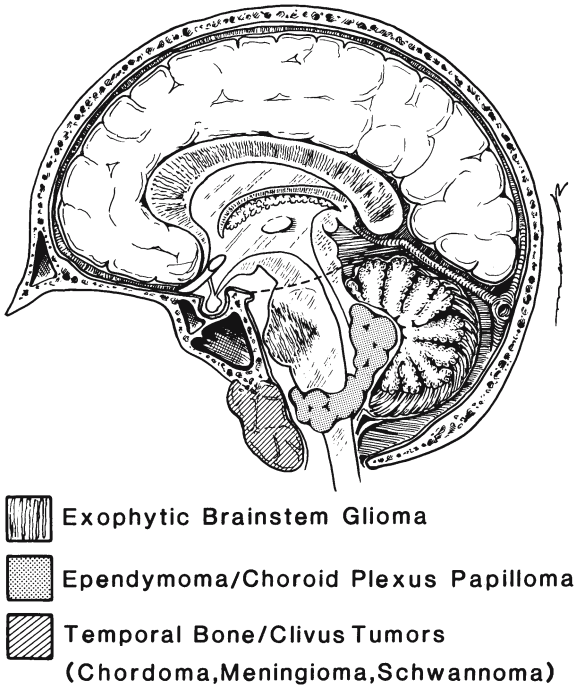

Fig. 4. Sagittal anatomic diagram depicts extra-axial posterior fossa masses in children, showing typical infratentorial lesions in each location

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