# CHAPTER 109
# Brain and Spinal Cord Tumours

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## Introduction

Brain and spinal cord tumours are increasingly being diagnosed in Africa and other developing countries as more centres acquire more sophisticated and less invasive diagnostic facilities. Improvement in the number and expertise of medical personnel also plays a significant role. In addition, advances in anaesthesia and surgical techniques have allowed for increased survival of children with these otherwise dismal clinical conditions. However, the situation in Africa is still far from the ideal, that obtains in the developed nations because only very few centres have the facilities and manpower to manage these conditions. Even in developed centres, however, the poor prognoses of some tumour types have not changed despite all the advances.[1]

Brain tumours are much more common than spinal tumours; this is more so in the paediatric age group. Available data show brain tumours as the second most common tumour after leukaemia (20%), and the most common solid paediatric tumours.[2,3,4] Brain tumours comprise 40–50% of all tumours.[5] One-quarter of all childhood cancer deaths is caused by brain tumours.[1] Although paediatric brain tumours are predominantly infratentorial (60%), the location of tumours depends highly on age. Children under 6 months of age are more likely to have a supratentorial tumour (75%).

The nature of the tumour also depends on location as well as age. In the infratentorial region, there is a relatively equal incidence of primitive neuroectodermal tumour (PNET); most commonly, medulloblastoma); brain-stem gliomas; and pilocytic astrocytomas. In the supratentorial region, astrocytomas are significantly more common. Concerning age, congenital tumours presenting in neonates are more likely to be neuroectodermal in origin, with teratomas being most common, whereas in older children, astrocytomas, PNET, and ependymomas predominate. Some glial tumours, such as mixed gliomas, are unique to children. They are located more frequently in the cerebellum (67%) and are usually benign.

## Demographics

The exact incidence or prevalence of central nervous system (CNS) tumours in most African countries is largely unknown. More new cases are seen now than previously, perhaps due to a true increase in incidence, or to increased awareness by the communities and medical workers, with more people presenting now than before. In addition, improvements in diagnostic techniques may be partly responsible for this increase.

From unpublished statistics obtained from Regional Centre for Neurosurgery in Sokoto, Nigeria, paediatric (age <15 years), brain tumours accounted for 22% of all the brain tumours with a male-to-female ratio of 4:1. Another centre in Nigeria found that the most common types of tumours encountered are the astrocytomas, medulloblastomas, craniopharyngiomas, and ependymomas.[6]

## Aetiopathogenesis

Like most neoplasms elsewhere in the body, the exact cause of craniospinal tumours in humans is unknown. In animal models, however many environmental agents have been used to induce tumours. Whether this can be extrapolated to the humans remains to be verified. Associated factors include the following:

- *Genetics:* Brain tumours in general are not believed to be inherited genetic disorders, except in von Recklinghausen's disease and some gliomas. Some chromosomal abnormalities may be associated in part with specific tumour types. Astrocytomas may be associated with abnormalities of chromosomes 7, 9, 10, 17, 19 and deletion of p53 gene and amplification of the epidermal growth factor (EGF) gene.
- *Chemical agents:* Many chemicals show carcinogenic properties in animals and produce CNS tumours, especially ethyl and methyl nitrosourea and anthracine derivatives, but this is yet to be proven in humans.
- *Viruses:* There have been reports of patients with JC virus (JCV)-induced demyelination who have developed multifocal astrocytomas, but again the evidence is not firm.
- *Radiation:* There is increasing evidence that exposure to excessive radiation plays a role in causing brain tumours. There is increased incidence of brain tumours in those who have had irradiation to the head and neck for different conditions.
- *Immunosuppression:* Immunosuppression is known to increase the risk of primary lymphoma of the brain, particularly in transplant patients, but it is unlikely that it plays a significant role in the development of cerebral tumours in general.
- *Trauma:* The concept that head trauma leads to the development of meningioma has been controversial. Although epidemiological studies do not support trauma as an aetiological factor, there have been case reports of meningiomas developing at the site of substantial meningeal trauma.

## Pathophysiology

As brain tumours increase in size, they acquire new blood vessels from surrounding blood vessels. Sometimes they grow so rapidly that they overwhelm the blood supply and undergo ischaemic necrosis. Many of the tumours cause surrounding oedema, which may be responsible for the mass effect produced by the tumour. Brain tumours rarely metastasize except for some such as medulloblastoma and ependymomas, which become carried along the cerebrospinal fluid pathways to the spinal cord—a phenomenon sometimes referred to as drop metastasis.

## Classification of Craniospinal Tumours

Classification of craniospinal tumours has evolved over the years. Currently, the most widely used classification system is the World Health Organisation (WHO) system, based on the cell of origin.[7] Detailed classification is beyond the scope of this chapter, but an abridged version (Table 109.1) is given to highlight the major groups.

## Clinical Presentation

The clinical presentations of brain tumours are those of features of raised intracranial pressure from the mass effect of the tumour or obstructive hydrocephalus, focal neurological deficit referrable to the

*Table 109.1: WHO classification system (abridged).*

| | Craniospinal tumours | Examples |
|---|---|---|
| Tumours of neuroepithelial tissue | Astrocytomas | Grades i-iv with the anaplastic type and glioblastoma multiformes carrying the worst prognosis |
| | Oligodendroglial tumours | Oligodendroglioma |
| | Ependymal tumours | Ependymoma and subependymoma |
| | Mixed gliomas | |
| | Choroid plexus tumours | |
| | Neuronal and mixed neuronal-glial tumours | Ganglioglioma |
| | Pineal tumours | Pineoblastomas |
| | Embryonal tumours | Neuroblastoma and retinoblastoma |
| Tumours of cranial and spinal nerves | Schwannoma | |
| | Neurofibroma | |
| | Malignant peripheral nerve sheath tumour | Malignant schwannoma |
| Tumours of the meninges | Tumours of meningothelial cells | Meningiomas |
| | Mesenchymal, nonmeningothelial tumours | Fibrous histiocytoma, mesenchymal chondrosarcoma |
| | Primary melanocytic lesions | Malignant melanoma |
| | Tumours of uncertain origin | Haemangioblastoma |
| Haemopoietic neoplasms | | Malignant lymphomas |
| Germ cell tumours | | Germinomas, choriocarcinoma, yolk sac tumours, teratomas |
| Cysts and tumour-like lesions | | Rathke's cleft cyst, dermoid and epidermoid cysts, colloid cyst of 3rd ventricle |
| Tumours of the pituitary | | |
| Local extension from regional tumours | | Craniopharyngioma, chondroma, chordoma |
| Metastatic tumours | | |

site of the brain affected, and/or seizure. Children with brain tumours often experience a long delay between the onset of symptoms and diagnosis because the symptoms are nonspecific and the patients are treated for conditions other than the brain tumour.

Historical findings include intermittent headache and nausea that is worse in the early mornings. As the disease progresses, the headache becomes persistent and there is associated vomiting. Depending on age, the child may present with worsening irritability and progressive head enlargement due to ensuing hydrocephalus. The child may also present with drowsiness, gait disturbance, visual impairment, limb weakness, stunting of growth or precocious puberty, and seizures.

Examination findings are age specific. The infant may present with features of hydrocephalus with tense fontanelles. There may be failure to thrive, cranial nerve deficits, and limb weakness. There may be hypertonia with exaggerated deep tendon reflexes. Visual assessment may show loss of some visual fields and the presence of papilloedema.

The diagnosis of craniospinal tumours requires a high index of suspicion and meticulous examination to elicit subtle changes that may give away the diagnosis; herein lies the need for commitment and patience.

## Diagnostic Investigations

Before the advent of computed tomography (CT) and magnetic resonance imaging (MRI) in the mid 1970s and 1980s, respectively, the diagnoses of brain tumours were based largely on x-rays and angiographic findings. The findings were often nonspecific. X-ray features include erosion of the clinoid processes, widening of the sella turcica, thinning of the skull bone with a copper beaten appearance, osteolytic or osteosclerotic lesions, and calcifications, among others.

CT scans have revolutionised neurosurgery and have now made it possible to diagnose most intracranial pathologies. CT also gives very good bony definition. It is able to give the site and size of the tumour and presence of surrounding oedema.

MRI gives a far better soft tissue definition than the CT scan and clearly defines the extent of the tumour and oedema. It also allows for the image to be taken in different planes. Modifications of this technique have increased the sensitivity and specificity of this diagnostic tool, including MR angiography (MRA), MR spectroscopy, and functional MRI (fMRI).

A lumbar puncture (to be avoided in the presence of raised intracranial pressure (ICP)) could be done to sample for tumour cells in cerebrospinal fluid (CSF) in suspected tumours such as medulloblastomas.

Conventional angiography is important to assess the vascularity of a tumour, look for the feeding vessels, and plan for possible tumour embolisation prior to surgery, thus minimising blood loss at surgery and easing surgical tumour resection.

## Management

The management of brain tumours begins with an adequate history and detailed clinical examination, followed by relevant investigations, as outlined earlier. These findings not only help to establish the diagnosis but also determine the extent to which the disease has affected the anatomy and function of the individual. Further, these findings identify those patients requiring emergency intervention.

Patients present as emergencies in the following settings:

- acute raised intracranial pressure due to tumour expansion, surrounding oedema causing mass effect, or acute hydrocephalus from tumour obstruction of the CSF pathway;
- haemorrhage into a tumour cyst, leading to sudden neurological deterioration (e.g., pituitary apoplexy); and

- any of the above leading to herniation, especially of the brain stem, which could be rapidly fatal.

Raised intracranial pressure can be managed nonsurgically, surgically, or a combination of both. Nonsurgical methods include the following:

- *head elevation* to 15°–30° with the head in a neutral position, which ensures adequate venous return and reduces congestion in the brain;
- *oxygen administration and controlled hyperventilation*, which causes cerebral vasoconstriction and reduces congestion within the brain;
- *use of diuretics* (e.g., mannitol and frusemide); and
- *steroids (dexamethasone)*, which are very useful for intracranial mass lesions with surrounding oedema. The effect is often dramatic with quick resolution of symptoms, which gives the surgeon a period of time to prepare for the definitive treatment.

The surgical methods for reducing raised intracranial pressure include the insertion of a shunt for hydrocephalus, excision of the tumour, or debulking where total excision is not feasible.

When patients do not present in an emergency setting, they are optimised for the definitive treatment, which includes correction of nutritional deficiencies, blood levels, dyselectrolytaemia, and cardiovascular and respiratory problems. The definitive treatment modalities include surgery, radiotherapy, chemotherapy, and immunotherapy. These modalities are either used singly or in various relevant combinations.

### Surgery

Surgery for childhood tumours is usually done for histological confirmation, total excision or maximum cytoreduction, neural decompression, and CSF pathway restoration. Surgical resection is usually the primary treatment modality; in fact, most benign brain tumours can be cured by surgical resection only. Patients with malignant tumours also benefit from maximal tumour debulking because it allows for better response to chemotherapy and radiotherapy.

Surgical resection has been improved by advances in surgical techniques and instrumentation, such as microsurgery, surgical lasers, and ultrasonic aspirators. Stereotaxy and intraoperative ultrasonography enable the surgeon to precisely localise the tumour based on a previously taken CT scan or MRI. Despite these advances, some tumours still pose great challenges. These include brain-stem tumours, optic chiasmal tumours, and diencephalic tumours, which are operable but often not completely resectable.

Complications of surgery include postoperative haemorrhage, major or minor neurological deficits, brain swelling, and infection. Fortunately, children tend to do better with rehabilitation, even from major postoperative neurological deficit. The success of surgery depends not only on the surgeon, but also on a coordinated and dedicated team that includes the neurosurgeon, anaesthetist, paediatrician, nurses, and physiotherapist, among others.

### Radiotherapy

Radiotherapy is required for children with malignant brain tumours or unresectable benign tumours because of a microscopic residual tumour, even when there is macroscopic total tumour excision. Some tumours, such as ependymomas, medulloblastomas and other PNETs are carried along the CSF pathway to the spine necessitating irradiation of the whole neuroaxis.

External beam radiotherapy entails giving high-energy radiation that passes through the scalp and skull to the brain. Anything along its path is irradiated. Complications include the effect on rapidly dividing cells, leading to transient hair loss, bone marrow suppression with anaemia, thrombocytopaenia, and leucopaenia. Endocrine dysfunction and progressive cerebral vascular thrombosis, which may present as a stroke 10–20 years after irradiation, have all been reported. Another complication is the induction of secondary tumours in the brain, which may occur 5–15 years after the irradiation.

A very serious complication of radiotherapy is its deleterious effect on the intellectual development of the child. About 80% of the development of the child's brain occurs in the first 2 years of life. Therefore, radiotherapy is generally not recommended in children younger than 3 years of age. An alternative is the use of chemotherapy in these young children.

Stereotaxic irradiation, such as that provided by a gamma knife or a linear accelerator, delivers focally intensified radiation with minimum damage to the surrounding tissues. A single large dose is usually given.

Another method is the use of brachytherapy or interstitial radiotherapy. In this modality, radioactive seeds are implanted in the tumour intraoperatively by using a catheter or by stereotactic means. The seeds are then removed after they have delivered the calculated required dose.

### Chemotherapy

Chemotherapeutic antineoplastic agents also affect rapidly dividing cells and give rise to complications similar to those produced by radiotherapy. They are often given in combination, and act by inhibiting cellular metabolism. They could be given intraarterially through the vertebral or carotid arteries (the use in children is still being evaluated), or intrathecal, to deliver high doses of the drug at the target site. Intrathecal chemotherapy can be given by use of spinal needles. A subcutaneous Ommaya reservoir can be used for intralesional, intracystic, or intraventricular administration of cytotoxic medications.

Intraarterial injection may be complicated by blindness and stroke.

## Individual Tumours of the Brain

The general features of tumours have been given earlier. The peculiarities of some specific tumours are noteworthy and are presented here.

### Infratentorial Tumours

Infratentorial, or posterior fossa, tumours are more common in the paediatric age group than in adults. The common posterior fossa tumours include medulloblastoma, cerebellar astrocytomas, ependymomas, and brain-stem gliomas.

#### Primary neuroectodermal tumours

PNETs include medulloblastomas, medulloepitheliomas, pigmented medulloblastomas, ependymoblastomas, pineoblastomas, and cerebral neuroblastomas. These tumours originate from undifferentiated cells in the subependymal region in the foetal brain. The frequency of PNETs is similar to that of pilocytic astrocytoma.

#### Medulloblastomas

Medulloblastomas initially arise in the inferior medullary velum (possibly from remnants of the external granular layer) and grow to fill the 4th ventricle, infiltrating the surrounding structures (Figure 109.1). They are the most common malignant posterior fossa tumour in the paediatric population. They are characterised by their tendency to seed along the neuro-axis, following CSF pathways, and rarely can metastasise to extraneural tissues.[8,9] Ten to thirty percent of affected patients will have evidence of "drop-mets" at the time of diagnosis. Extracranial metastases account for 5% of cases, involving bone, liver, and lymph nodes.

Medulloblastomas are highly cellular, vascular tumours with a deeply basophilic nucleus and multiple mitoses ("small blue cell tumour"). These histological features are commonly seen in the other variants of PNET.

The male-to-female ratio is approximately 3:1. Medulloblastomas demonstrate a bimodal age distribution: a larger peak occurs at 5–9 years of age, and a smaller peak occurs at 20–30 years of age.

Gross surgical excision followed by craniospinal irradiation remains the ultimate goal in these patients. Any attempt to remove the tumour from the floor of the 4th ventricle, however, can lead to significant morbidity. Up to 30% of tumours will have invaded the floor, and care

must be taken not to damage this very eloquent region.

Children with nondisseminated medulloblastoma and old enough to have radiation therapy have a 5-year survival rate of about 70%. In the presence of dissemination, adjuvant chemotherapy (CCNU (lomustine), vincristine, and prednisolone) can be given, but the survival rates are significantly lower. Neurological, endocrine, and cognitive deficits are not uncommon in this latter group.

### Piloctyic astrocytoma

Low-grade astrocytomas occur more often in children and young adults than in adults. They are the most common astrocytic tumours in children, accounting for 80–85% of cerebellar astrocytomas and 60% of optic gliomas. They comprise about 33% of all posterior fossa tumours in children and represent about 25% of all paediatric tumours.

Pilocytic astrocytomas usually arise in the cerebellum, brain stem, hypothalamic region, or optic pathways, but they may occur in any area where astrocytes are present, including the cerebral hemispheres and the spinal cord.

There is an association of pilocytic astrocytomas with neurofibromatosis; optic nerve gliomas are common tumours in patients with this condition and may present bilaterally. Patients with optic pilocytic astrocytomas associated with NF1 usually have better outcomes than patients with juvenile pilocytic astrocytomas.

These tumours are usually discrete, indolent lesions associated with cyst formation. The cysts may be unilocular or multilocular, with an associated tumour nodule. CT scan reveals a hypodense or isodense nodular mass that homogeneously enhances with contrast with a cystic component in 60–80% of cases. Calcification is present in 10% of juvenile pilocytic astrocytomas. MRI shows a contrast enhancing hypointense lesion on T1WI and hyperintense on T2WI (Figures 109.2 and 109.3).

Histologically, the tumour contains fibrillary astrocytes with associated Rosenthal fibres (intracellular eosinophilic rod-shaped bodies). The tumour is named after the "hair-like" bipolar (piloid) astrocytes. Features more typically seen in higher grade gliomas can be seen (e.g., nuclear atypia, mitoses, endothelial proliferation, and necrosis) but they have no proven prognostic significance.

The peak incidence of pilocytic astrocytoma is in patients 5–14 years of age; it has no gender predilection.

Posterior fossa pilocytic astrocytomas are typically treated with surgery and completely resected whenever possible. Optic nerve tumours can often be managed conservatively, and surgery is usually indicated in an attempt to preserve vision in the unaffected eye. There has been some success treating these tumours with chemotherapy (particularly platinum-based regimes), but this is used only for unresectable progressive tumours, or progression of remnants.

Patients with juvenile pilocytic astrocytoma have a better prognosis than patients with most other types of astrocytomas. If gross total resection is possible, the 10-year survival rate is as high as 90%.[10] After subtotal resection or biopsy, the 10-year survival rate is as high as 45%. Morbidity is related to the location of the tumour and to the associated complications of tumour resection.

### Ependymoma

Ependymomas account for 1–8% of paediatric brain tumours. There is a female preponderance with a female-to-male ratio of approximately 2:1. The median age of presentation is in the second decade, but there is a bimodal peak with the younger peak at about age 5 years. Ependymomas are derived from ependymal cells and occur most commonly in the ependymal lining of the ventricles. These tumours can also arise in the conus (myxopapillary) and spinal canal. The majority of ependymomas are located in the posterior fossa with a predilection for the 4th ventricle; 20% arise from the floor. Extension through the foramina of Magendie and Luschka is not uncommon. Approximately 10% will have spinal metastases at the time of diagnosis, although

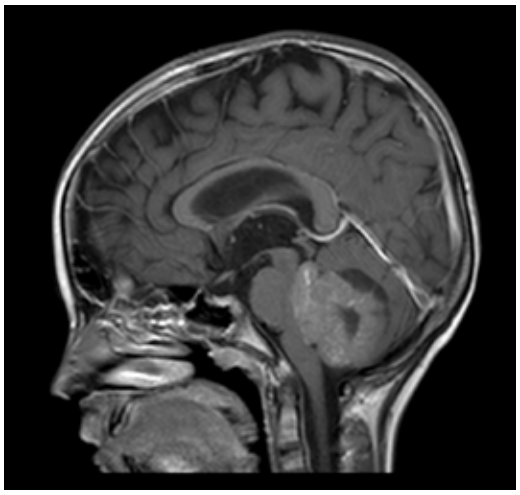

*Figure 109.1: Medulloblastoma. Note the tumour filling the 4th ventricle and extending up the cerebral aqueduct.*

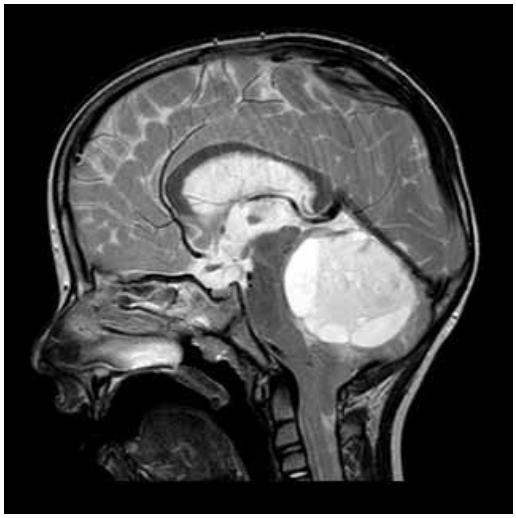

*Figure 109.2: Large posterior fossa pilocytic astrocytoma. Hydrocephalus has been treated by external ventricular drainage (EVD), but tonsillar herniation remains.*

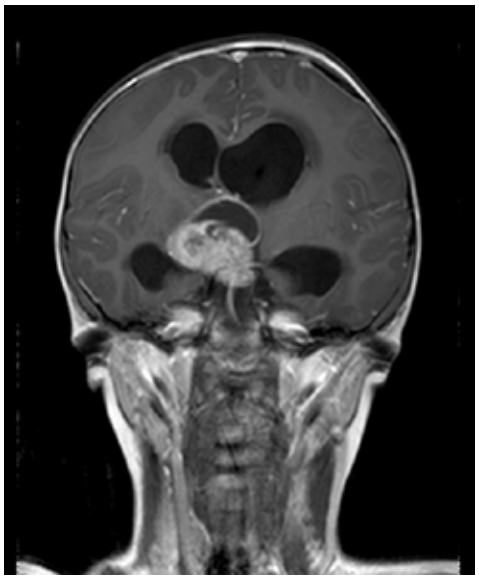

*Figure 109.3: Large suprasellar pilocytic astrocytoma encircling the basilar artery. This could not be completely removed at surgery.*

this is more common with the anaplastic variants. Tumours presenting supratentorially are more common in adults and are usually located in the trigone of the lateral ventricles. Symptoms of ependymomas are similar to those of medulloblastoma, but there may also be cranial nerve deficits.

Ependymomas of the posterior fossa present as solid mass lesions. Histologically, they are uniform ependymal cells forming true rosettes and perivascular pseudorosettes. It is not uncommon to have associated calcification, cysts, and haemorrhage. Anaplastic ependymomas tend to have histological features of a higher grade tumour with vascular proliferation and necrosis. Ependymomas of the spinal cord and conus are more likely to be low grade, and total excision may be curative.

A CT scan of the tumour shows an isodense mass that may enhance slightly but irregularly with contrast.

Gross total surgical excision, where possible, with adjuvant radiotherapy is the treatment of choice.

Combined treatment leads to a 35–60% 5-year survival rate. Although adults have an increased tendency to anaplastic variants, their survival rate is better than that of children. This is most likely due to the fact that craniospinal radiation therapy is limited to children older than the age of 5 due to the adverse effects on the developing nervous system.

### Brain-Stem Gliomas

Brain-stem gliomas require special mention because they present a peculiar challenge to the neurosurgeon. Their total resection is difficult and often impossible, except in expert hands.

In children, brain-stem gliomas represent 10–20% of all brain tumours. Most brain-stem gliomas are low-grade astrocytomas and can be conservatively managed. Surgical debulking is reserved for exophytic portions of tumour.

Ninety percent of diffuse pontine gliomas are, however, more aggressive (anaplastic astrocytoma or glioblastoma) with a very short survival. They present with rapidly deteriorating symptoms such as dysarthria, hemiparesis, ataxia, and facial and abducens nerve palsies.

MRI shows enlargement of the pons, with a lesion hypointense on T1WI and hyperintense on T2WI that is contrast enhancing. CT also shows an enhancing lesion (Figure 109.4).

Because of the danger and difficulty of obtaining a biopsy in this condition, the patient could be commenced on radiotherapy even without a biopsy. However, some authorities have completely resected such tumours.

## Supratentorial Tumours

### Craniopharyngioma

Craniopharyngiomas arise from remnants of the Rathke's pouch. They account for 2–4% of all brain tumours, occurring mostly in the age group 5–15 years. Craniopharyngiomas are slow growing and may attain a large size before manifesting. They present with visual impairment due to compression of the optic nerves; endocrinopathy, especially diabetes insipidus and obesity; and features of hydrocephalus due to extension to the floor of the 3rd ventricle.

CT scan shows a sellar or parasellar mass with cystic and solid components and sometimes calcification. A preoperative hormonal work-up is very important. Teamwork with an endocrinologist and ophthalmologist is invaluable.

Surgery is the primary treatment modality and may employ the use of endoscopic trans-sphenoidal resection or craniotomy and excision.

Radiotherapy, especially intracystic injection, has been found useful in incompletely resectable or nonresectable lesions; also effective is the intracystic injection of bleomycin. Systemic chemotherapy has not been found useful.

### Pilocytic astrocytoma

Pilocytic astrocytoma differ markedly from infilterating fibrillary or diffuse astrocytomas in terms of their ability to invade tissue and for malignant degeneration. They occur throughout the neuraxis and are more common in children and young adults than in adults. Malignant transformation has been reported after many years, even without radiotherapy. Cerebral gliomas, optic gliomas, and thalamic and hypothalamic gliomas tend to occur in young adults.

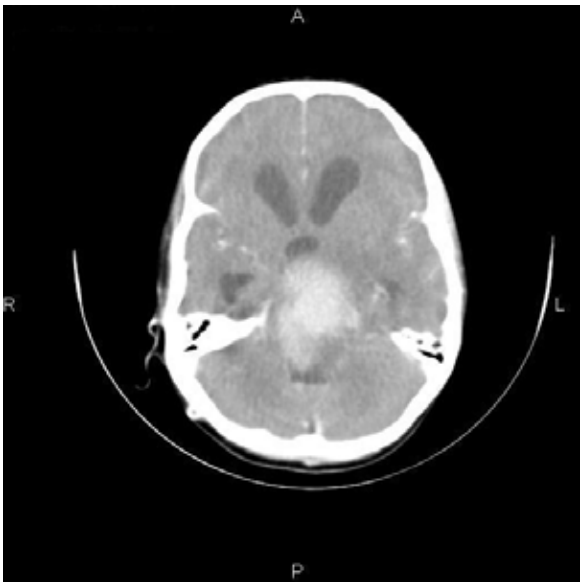


*Figure 109.4: Post contrast CT of a 12 year-old-girl with brain-stem glioma. Note the enhancing lesion with dilated lateral and 3rd ventricles.*

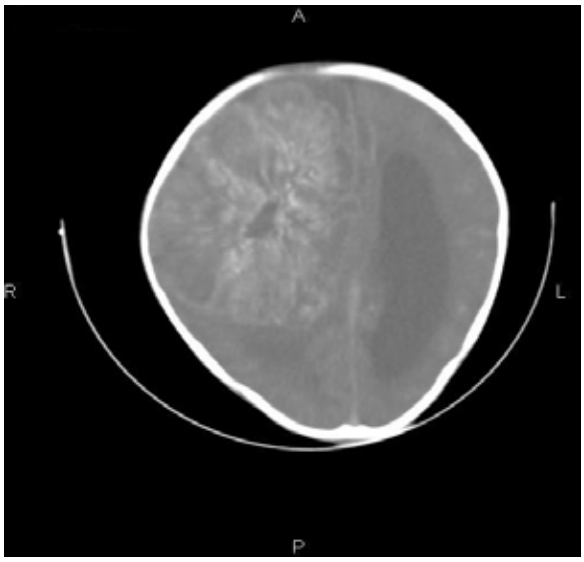


*Figure 109.5: CT scan of a 6-year-old boy showing a well-circumscribed huge right fronto-parietal contast-enhancing astrocytoma. Note the effacement of the right ventricle, mild oedema, midline shift, and left lateral ventriculomegaly.*

Hemispheric lesions give rise to headaches, seizures, and focal weakness. Chiasmal tumours present with visual deficits, endocrine dysfunction, or symptoms of hydrocephalus.

CT or MRI show these tumours as well-circumscribed, contrast-enhancing lesions (Figure 109.5) with cystic components and a mural nodule, having little or no surrounding oedema.

Surgery is the most recommended treatment modality, with an aim to total excision, including especially the mural nodule. Radiotherapy is controversial, but can be used after a subtotal resection in a patient older than 3 years of age. Chemotherapy is given to younger patients.

Complete resection yields 100% recurrence-free survival without adjuvant therapy.[11] After subtotal resection and radiotherapy, 10- and 20-year freedom from progression rates are 74% and 41%, respectively,

and 10- and 20-year survival rates are 81% and 54%, respectively.

#### Choroid plexus papilloma and carcinoma

Choroid plexus papilloma and carcinoma represent 0.4–0.6% of all intracranial tumours. They represent approximately 3% of childhood brain tumours and are more likely to occur in the lateral ventricle. Although these tumours can occur in the 4th ventricle, this feature is more commonly seen in adults. Approximately 85% of tumours present in children under 5 years.

Symptoms can be due to hydrocephalus, which is likely to be due to the presence of increased protein and xanthochromia within the CSF, causing diminished absorption. Overproduction of CSF is not uncommon, but does not explain the persistence of hydrocephalus after tumour removal. These tumours may also present with mass effect, particularly in the case of papilloma.

Although macroscopically, choroid plexus papilloma appears as a reddish solid tumour, histologically, it can be difficult to differentiate from a normal choroid. As with the malignant variants of a tumour, the presence of nuclear atypia, mitoses, and necrosis is suggestive of carcinomatous change.

Surgical resection results in a cure for benign papillomas. These tumours are very vascular, and in the presence of carcinoma, preoperative chemotherapy or embolisation may have roles in shrinking tumour size and reducing vascularity in order to aid surgical resection.

Five-year survival rates of about 80% have been described. The main late morbidity arises from persistent subdural collections due to a ventriculo-subdural fistula.

### Spinal Cord Tumours

Spinal cord tumours are relatively rare in the paediatric age group when compared to brain tumours.[12] They could be extradural or intradural. The intradural tumours can be intramedullary or extramedullary, with intramedullary tumours more common than extramedullary. Intramedullary tumours in children include astrocytomas, ependymomas, gangliogliomas, diffuse leptomeningeal tumours, and haemangiomas. The more common childhood extramedullary tumours consist of meningiomas, schwanomas, and neurofibromas; extradural tumours comprise lipomas and dermoid and epidermoid cysts.

#### Astrocytomas

Astrocytomas are by far the most common childhood spinal tumours. They are mostly benign, but can be malignant in 10–15% of cases. They may remain asymptomatic for a long time, reaching large sizes before presentation. The onset of symptoms is usually insidious and may appear at the time of a trivial injury. An early symptom is back pain, which is usually diffuse.

#### Extramedullary tumours

Radicular pain may be suggestive of an extramedullary lesion.

Children present with frequent falls, lower limb weakness or motor regression, or gait abnormalities. About a third present with kyphoscoliosis. Sphincteric involvement occurs late in the disease. A few may present with features of hydrocephalus, which is thought to be due to increased protein secretion by the tumour cells into the CSF, leading to obstruction. Examination reveals varying degrees of motor weakness and upper motor neurone features.

MRI is the diagnostic tool of choice because it clearly defines the lesion. CT myelography may be done where MRI is contraindicated or not available. Plain radiographs are mandatory for those with scoliosis. Again, extramedullary tumours may show as thinning or sclerosis of the pedicles. There may also be enlargement of the neural foramina in dumbbell tumours.

Adjuncts for spinal cord surgery include ultrasonic aspirator and lasers.

Astrocytomas are excised after performing a laminectomy or laminotomy. The outcome depends on the neurological state of the patient prior to surgery. Those with incomplete injury are likely to benefit more than those who have a complete injury. The majority of extramedullary tumours are benign, and gross total excision is possible in most cases.

## Management

### General Principles

Mass lesions in children and adults present in one (or more) of the following ways:

- raised ICP, which can be due to the mass lesion or hydrocephalus;
- seizures, which can be focal or generalised;
- focal neurological deficits, which are specific to the location of the tumour; and
- pituitary tumours and craniopharyngiomas, which may present with either a neurological deficit (in particular visual loss) or endocrine dysfunction.

Classic neurological features of raised ICP in older children and adults include headache, vomiting, and blurred vision. Papilloedema/ataxia is another feature. Nonspecific features in infants and children, as previously mentioned, include poor feeding/failure to thrive, irritability, behavioural disturbance, and deterioration in schoolwork.

Remember ABC when assessing any acutely unwell patient: airway, breathing, circulation. This is particularly relevant in children who may have a decreased conscious level, or may be dehydrated due to anorexia and vomiting.

### Work-ups

The general work-up is the same as for hydrocephalus.

Specific work-ups include the following:

- MRI of the brain and spine is preferable prior to any surgical intervention; is useful to assess suitability for endoscopic management (e.g., endoscopic third ventriculostomy (ETV) or biopsy); allows better evaluation of the tumour and its relation to eloquent structures; and allows assessment of any spinal disease (e.g., drop metastases). A CT scan can, of course, be used as an alternative—it is not quite as good, but with reconstructions in the sagittal plane, is a realistically achieved alternative.
- Tumour markers are obtainable from CSF and serum, and are essential for optimising management of pineal region tumours.
- Endocrine assessment should be used for pituitary and craniopharyngioma patients, who may need preoperative hormone replacement.
- Opthalmology and visual field assessment is most commonly required for pituitary patients, and is recommended for any child with a tumour along the visual pathway.

## Treatment Options

Previous series have described carrying out treatment for symptomatic hydrocephalus first. This has the advantage of rapidly improving such symptoms as headache and vomiting, and also allows investigation of CSF markers. In the presence of an aqueductal stenosis due to an offending tumour, an initial endoscopic approach may be used to create a third ventriculostomy and biopsy the tumour at the same sitting.

In recent years, however, in the case of a posterior fossa tumour, the pendulum has swung back towards single-sitting surgery. This option is aimed at opening up the CSF pathways and avoids the added risks of a third ventriculostomy or shunt. An EVD may be placed during the early part of the procedure if there is grossly raised ICP. A drawback is that this regime relies on an operating theatre being available at the time of presentation or deterioration, whereas the two-stage procedure allows more leeway from the organisational point of view.

Surgical excision is carried out wherever tumour location makes it possible. Unlike for adult cases, the free use of chemo- and radiotherapy is not possible for young children. Children under the age of 5 years

have a significant long-term morbidity with whole brain (with or without spine) radiotherapy. Outcomes are improved in children older than 5 years of age at diagnosis with no evidence of disseminated disease and maximal surgical resection.

Brain-stem and optic tract gliomas are usually low-grade astrocytomas and are predominantly managed conservatively.

## Follow-up Imaging

MRI within the first 24 hours after surgery provides a baseline for further follow-up and optimises any adjuvant therapy. The artefact produced from surgical intervention makes radiological assessment of any residual tumour after 48 hours exceptionally difficult; therefore, if imaging cannot be performed within this time period, it should be deferred for at least 6 weeks.

## Key Summary Points

1. Central nervous system tumours are the most common childhood tumours worldwide.
2. More cases are reporting to health facilities, but the challenges in Africa are inadequate facilities and manpower to manage these conditions appropriately.
3. Presentation commonly involves features of raised intracranial pressure, seizure, or focal neurological deficit.
4. Africans typically present at the late stage of brain and spinal cord disease.
5. CT scan and MRI are the investigations of choice, but tissue diagnosis is required for treatment (except in the few instances in which this is not feasible due to the site of the tumour).
6. Full resection is not always the aim—the histology and location of the tumour should dictate the surgical aim.
7. Pre- and postsurgical adjuvant treatment will become increasingly important as their efficacy increases.
8. Patients with incomplete neurological deficit are more likely to benefit from spinal surgery than those with complete deficits.

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## Suggested Reading

Paediatric neurosurgery is a constantly evolving science. The most up-to-date reviews are to be found in journals, many of which are available online. In the authors' opinion, the best are Child's Nervous System (published by Springer), Paediatric Neurosurgery (published by Karger), and Journal of Neurosurgery (Pediatrics).

Many textbooks are available; two that cover many aspects of both diagnostic and operative management of paediatric neurosurgical patients are:

Albright AL, Pollack IF, Adelson PD. Principles and Practice of Pediatric Neurosurgery. Thieme Medical Publishers, Inc., 2008.

McLone DG, ed. Pediatric Neurosurgery: Surgery of the Developing Nervous System, 4th ed. Saunders, 2001.