Skull and Scalp Anomalies

4

Donna C. Wallace and Lindsey N. Weak

4.1 Introduction

Anomalies of the pediatric skull and scalp can be thought of as congenital or acquired. Though most lesions in the pediatric population are benign, there are a few malignant types as well.

Typically, an anomaly may be found upon palpation by a parent (Fig. 4.1) or the primary care provider. The child may then be referred to the pediatric neurosurgery practice for a more thorough evaluation. Identification and examination of a lesion would include the size and location, color, whether it is mobile or fixed, and whether it is pulsatile or painful to touch. A lesion with clear drainage may be leaking CSF.

D.C. Wallace, MS, RN, CPNP (✉)
L.N. Weak, DNP, RN, CPNP
Division of Pediatric Neurosurgery,
Banner Children Specialists at Cardon Children's Medical Center, 1432 S Dobson Road Suite 403, Mesa, AZ 85202, USA
e-mail: Dcwallace001@gmail.com; Lindsey.weak@gmail.com

4.2 Diagnostic Testing

Once the child is referred to pediatric neurosurgery, the provider may order a computed tomography (CT) scan or a magnetic resonance imaging (MRI). It is not uncommon for newborns to undergo ultrasonography (US); however, ultrasounds are usually considered preliminary tests, and more eloquent testing is usually needed. CT scans are ordered judiciously because of radiation exposure to the growing cranium. Most pediatric facilities limit the amount of radiation by using specific formatting. CT scans are ordered to assess bony lesions, whereas MRIs are ordered to assess soft tissue lesions, as well as possible intracranial involvement. Swift and Trumble (1999) note that up to one third of patients with solitary nontraumatic lumps on the head have some degree of intracranial extension.

Lesions that are noted to be in the midline or pedunculated (a stalk-like structure) should be evaluated with an MRI to assess the degree of probable intracranial extension. Midline structures that are pulsatile should be considered vascular and may be connected to a dural sinus (e.g., sinus pericranii). Further testing of possible vascular lesions may require specialized magnetic resonance imaging studies that include venous and arterial structures, known as MRI/MRVs. Arteriograms may also be performed (Table 4.1).

C.C. Cartwright, D.C. Wallace (eds.), *Nursing Care of the Pediatric Neurosurgery Patient*,
DOI 10.1007/978-3-319-49319-0_4

Fig. 4.1 Finding a new bump while washing hair

4.3 Neoplastic

The term neoplasm refers to an abnormal growth of tissue caused by the rapid division of cells that have undergone some form of mutation. Benign neoplasms are usually localized and do not invade other tissues, while malignant neoplasms are usually invasive.

Benign skull lesions are typically seen as a single lesion on radiographs, and CT scans or skull X-rays are typically performed as the bone is more clearly identified in these studies. Malignant lesions tend to have ragged margins and usually occur in multiples.

Benign

Examples of benign tumors of the skull include epidermoid and dermoid cysts, fibrous dysplasia,

Table 4.1 Lesions of the skull and scalp overview

Lesion	Histology	Imaging features	Signs/symptoms	Treatment
Cutis aplasia	Varies depending on depth of defect. Fibrovascular stomas and/or edematous stroma	N/A	Well-demarcated, noninflammatory ulceration	Smaller lesions will re-epithelialize without tx, larger lesions require surgical repair (skin grafting)
Cranial dermal sinus tract	Stratified squamous epithelium	Inferiorly directed tract, enlarged foramen cecum may be present with nasal cranial dermal sinuses	Dimple usually located along the middle, with/without clear or yellow drainage	Surgical resection if intracranial
Epidermoid/dermoid cyst	Squamous epithelium and keratin, dermoids also include hair and sebaceous/sweat glands	Lytic with sclerotic margins and bone erosion	Soft or hard, nonmobile nodule. Usually appears over suture lines	Surgical resection
Ewing's sarcoma	Compact and uniform cells with distinct nuclei. Sheets of round blue cells with increased nucleus-to-cytoplasm ratio	Bone destruction with irregular and poorly defined borders, "onion-peel" arrangement. Associated soft tissue mass	Intermittent at site, worse at night. Size of tissue mass can fluctuate. Leukocytosis, anemia, fevers	Local radiation with systemic chemotherapy. Surgical resection depending on the site of the disease
Fibrous dysplasia	Fibroblastic collagen mixed with immature woven bone	Sclerotic, cyst-like, or "ground glass" appearance	Painless, boney skull deformity	Conservatively followed or surgical excision if symptomatic
Neuroblastoma	Sheets of neuroblasts with small round blue cells, dark nuclei, and little cytoplasm	"Sunburst" appearance of bone spicules with elevation in periosteum seen on CT. Often present near cranial suture lines in children	Headache, pain, fever. "Raccoon eyes" with orbital bone involvement	Radiation and chemotherapy

Table 4.1 (continued)

Lesion	Histology	Imaging features	Signs/symptoms	Treatment
Neurofibromas	Proliferation of all aspects of peripheral nerves. Wavy serpentine nuclei with pointed ends. Stromal mucin deposition and fibroplasia	Sphenoid wing dysplasia, pseudarthrosis	Axillary freckles, café au lait spots, dermal fibromas	Surgical excision
Langerhans cell histiocytosis	Large cells with eosinophilic cytoplasm and irregular nuclei	Lytic or "punched-out" appearance, with/without soft tissue mass	Asymptomatic or with localized pain over a raised, soft, and tender area	Low-dose radiation, surgical excision, or conservative tx with immobilization (lesions of the spine and long bones)
Osteoma	Similar to normal bone, decreased marrow	Hyperdense and expansile	Asymptomatic or with pain over lesion	Biopsy and/or surgical excision
Osteoblastoma	Large epithelioid osteoblasts	Well-demarcated, mixed lytic/sclerotic lesion with enlarged diploe	Asymptomatic or with pain over lesion	Biopsy and/or surgical excision
Sinus pericranii	Endothelial lining with congenital etiology. Connective tissue with traumatic etiology	Soft tissue mass with extracranial and intracranial venous communication seen with venogram. Associated skull defect	Compressible, painless mass. Enlarged in recumbent position and diminished in erect position	Surgical resection

histiocytosis (eosinophilic granuloma), osteomas and chondromas, as well as neurofibromas. Benign vascular lesions include hemangiomas, sinus pericranii, encephaloceles, and aneurysmal bone cyst.

Acute neoplasms of the scalp are unusual in children; congenital lesions of the scalp are more common. Lesions that are not attached to the cranium may be referred to a pediatric plastic surgeon for evaluation. An example of an interesting and rare benign lesion of the scalp is cranial fasciitis (Halder et al. 2012).

Malignant

Malignant lesions of the skull include chordomas, sarcomas, neuroblastomas with inner osseous involvement, and some leukemias.

Malignant lesions of the scalp are rare, but there are a few examples of congenital lesions that could become malignant. These include nevus sebaceous lesions that could lead to basal cell carcinoma. Some moles or nevi could also become malignant.

4.3.1 Neurofibromas

Craniofacial neurofibromas can involve peripheral nerves of the face, orbit, and cranial base. Facial lesions can be disfiguring and are usually taken care of by plastic surgeons. Neurofibromas are seen in patients who have neurofibromatosis (NF), which is classified into two distinct clinical groups based on the pattern of clinical presentation. These are multisystem disorders.

4.3.1.1 Epidemiology

NF 1 is one of the most commonly occurring genetic disorders and is seen frequently in the neurology and neurosurgery clinic. It occurs in 1 in 3,000–4,000 people (Pollack 2008). It is an autosomal dominant condition with the effected gene on chromosome 17 (17q11.2). Spontaneous mutation is high, with 30–50% of cases representing new mutations.

NF2 is one tenth as common, affecting only 1 in 50,000 patients (Pollack 2008).

4.3.1.2 Evaluation

Children with NF1 often have lesions of the skin such as freckles in unusual places (i.e., axillary), multiple café au lait spots, or dermal fibromas. Eye lesions can include glaucoma, optic pathway gliomas, and Lisch nodules. Sometimes pulsatile exophthalmos is also present.

Children with NF 2 can present with either unilateral or bilateral cranial nerve VIII lesions. They may also present with a meningioma.

Neurofibromas of the scalp may be seen in NF 1 and sometimes NF 2.

Patients who have neurodermatoses are often screened with CT scan or MRIs. It is during the screening processes that lesions of the skull and scalp may be found. The Committee on Genetics of the American Academy of Pediatrics has published guidelines on how these children should be screened and monitored (Hersh and Committee on Genetics 2008).

Bony lesions of the skull in children with NF include sphenoid wing dysplasia seen on head CT scan. Other bony lesions include tibial bowing as well as pseudarthrosis.

4.3.1.3 Treatment

Focal, resectable, or symptomatic lesions are surgically removed. Other lesions are usually followed with routine radiographs. The nurse is mindful to observe for lesions that are growing or changing in characteristics. Clinical coordination includes making sure the children are seen by the appropriate disciplines including neurology, genetics, orthopaedics, and neurosurgery.

4.3.2 Fibrous Dysplasia

4.3.2.1 Etiology and Pathophysiology

Fibrous dysplasia is another example of a lesion that though benign can become malignant (Greenberg 2014). Normal bone is replaced with fibrous connective tissue. The abnormal tissue is composed of fibroblastic collagen, and it is mixed with immature woven bone.

The etiology is not clear. There are three types: (1) cystic, though not an actual cyst, there is widening of the diploe with thinning of the outer table and little involvement of the inner table; (2) sclerotic, usually seen in the skull base and facial bones; and (3) mixed.

Fibrous dysplasia accounts for approximately 2.5% of all bone tumors (Swift and Trumble 1999). It is most commonly seen at puberty and has equal distribution between boys and girls. As is seen with other bone tumors, the disease becomes more prominent during periods of growth.

4.3.2.2 Evaluation

Initially, the child presents with a progressive and painless deformity of the skull. Various parts of the craniofacial skeleton may be involved including the orbit and foramina of other cranial nerves. Thus, there is concern of visual impairment when it involves the orbit because of distortion of the globe. These children are usually seen by a craniofacial team (Fig. 4.2).

As this is a bony lesion, the most appropriate test would be a CT. On plain radiographs and on CT, the lesion has a "ground glass" appearance due to the spicules of woven bone. An MRI may be performed to rule out intracranial involvement, including the involvement of cranial nerves.

4.3.2.3 Treatment

These can be slow-growing lesions and may be followed conservatively. If the lesion is impairing vision or impedes other cranial nerves, surgical excision is planned.

A meta-analysis of surgery versus watchful waiting by Amit et al. (2011) looked at several trials, studies, and individual case presentations. It was determined that surgeons prefer "watchful waiting" if the patient was asymptomatic, and surgery would be preferred for those who were symptomatic (i.e., visual changes). Asymptomatic patients with lesions of the optic nerve who underwent optic nerve decompression usually had some loss of vision. Thus, it was determined that watchful waiting would be more appropriate for those patients without impaired vision.

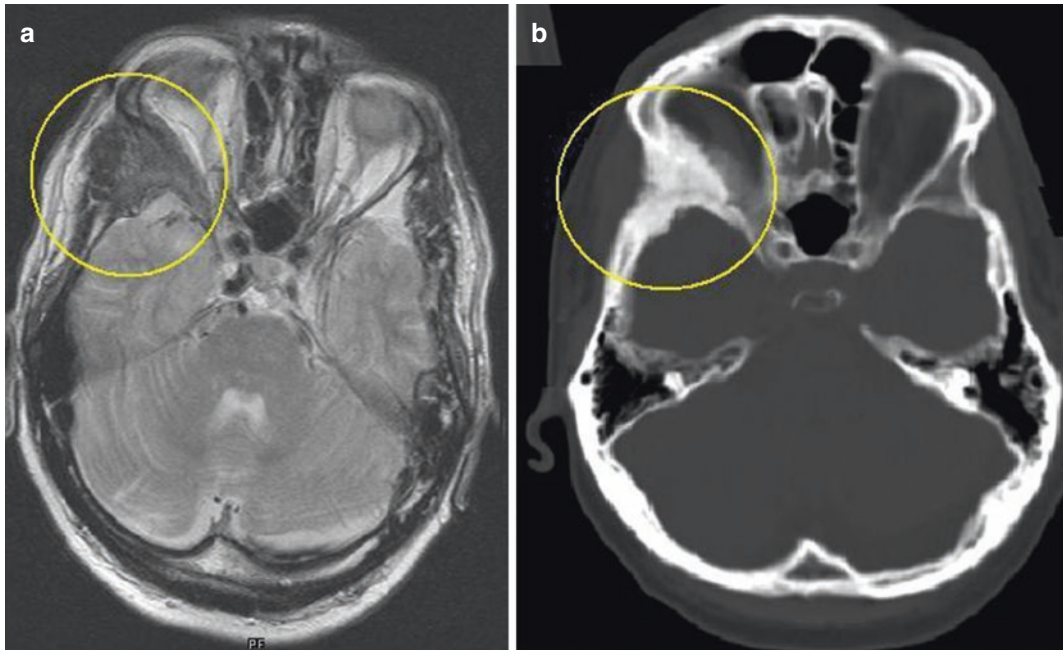


Fig. 4.2 Fibrous dysplasia seen on (**a**) MRI and (**b**) CT with bone windows

4.3.3 Langerhans Cell Histiocytosis

Langerhans cell histiocytosis (LCH) is an entity that has been referred to as histiocytosis X or eosinophilic granuloma. When seen as a single lesion, it is classically known as eosinophilic granuloma.

LCH is a nonneoplastic type of histiocytic disorder that is most commonly characterized by a single lesion but also can be seen as multiple osteolytic bone lesions. Multiple lesions are more frequently seen in children under age 3 years.

LCH can be seen as a systemic entity, disseminating into the CNS and through the viscera. Lesions can present as skin lesions, pulmonary infiltrates, hepatosplenomegaly, and lymphadenopathy.

There is a subgroup of malignant histiocytic disorders, and these include the monocyte-related leukemias (acute monocytic leukemia, acute myeloid/myelomonocytic leukemia) and other tumors.

4.3.3.1 Etiology

LCH is a rare histiocytic disorder that accounts for 7–10% of all reported skull lesions. It is seen in all age groups but it is most common in children from 1 to 3 years of age. The incidence appears to be three to five cases per million children, which is the same as pediatric Hodgkin's lymphoma, and in one to two cases per million in adults.

The genetics of LCH is not well defined at this time. There is no current evidence that relatives of the patients with this condition are at increased risk of developing it.

4.3.3.2 Pathophysiology

LCH is named because of a presumed derivation of the morphology similar to Langerhans cells, which are specialized dendritic cells found in the skin and mucosa.

Gene expression array data has shown that Langerhans cell is not the actual cell origin for LCH. Rather it is a myeloid dendritic cell that expresses the same antigens (CD1a, CD207) as the actual Langerhans cell.

A few studies reported evidence of viruses within the LCH lesions, but at this time, viral etiology debate is ongoing. It is now thought to be an inflammatory myeloid neoplasia because of

two mutations found in up to 75% of all biopsies. Some studies support that LCH is a reactive condition due to immunological dysfunction stemming from regulatory T-cell expansion and a lack of mutations and tumor suppressor genes. However, there is also support that LCH is a neoplasm which comes from finding cancer-associated proto-oncogene mutations seen in a high percentage of LCH biopsies (McClain 2016).

4.3.3.3 Clinical Presentation

The areas of involvement vary with age. Acute disseminated multisystem disease is most commonly seen in children less than 3 years of age, while a more indolent disease involving the single origin is more common in older children and adults. Presenting symptoms in patients depend on the organ system involved. Most patients with bone marrow involvement are young children with diffuse disease in the liver, spleen, lymph nodes, and skin. LCH patients under 2 years of age with two or more "risk organs," such as hematopoietic organs, have a poor prognosis.

4.3.3.4 Lytic Bone Lesions of LCH

Bone involvement occurs in the majority of patients with this condition. The most frequent sites of bony involvement are the skull (40–80%), femur, ribs, vertebrae, and humerus.

It is interesting to note that some lesions may be asymptomatic, while other patients may complain of pain in a localized area of the bone. Examination of the area usually reveals a lesion that is raised, soft, and tender to palpation. Skull radiographs typically demonstrate a lytic or "punched out" appearance with unequal involvement of the outer and inner tables. Typically there is a sclerotic rim. Sometimes there is an accompanying soft tissue mass (Fig. 4.3a–c).

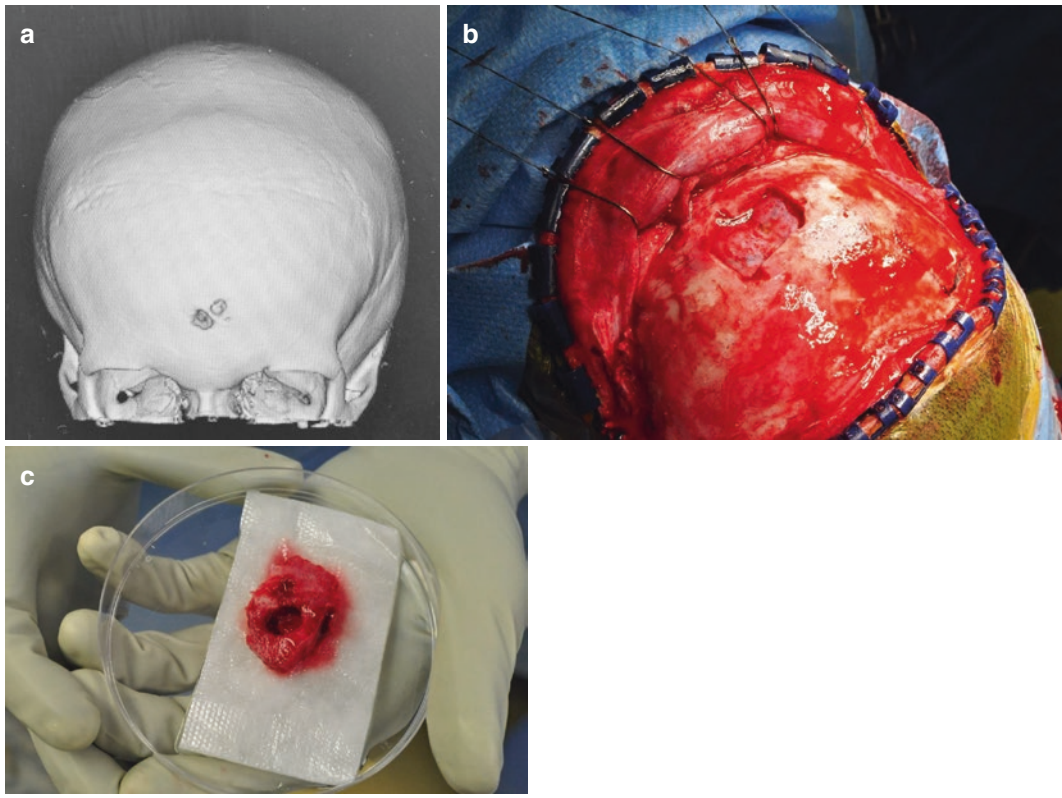


Fig. 4.3 (**a**) A 12-year-old girl with recurrent frontal skull lesions from histiocytosis. (**b**) Intraoperative view of excision of skull lesions. (**c**) Excised lesion with the surrounding bone

4.3.3.5 Treatment

Treatment depends on presentation. For example, in cases where children have pain without neurologic deficit, immobilization may be tried (such as in lesions of the spine or long bones). Studies indicate that younger children may spontaneously regress with arrest of further bony destruction. The pain usually resolves when the disease arrests. Low-dose radiation is sometimes used for these lesions with good result.

Parent and Shiflett (2011) state that management of LCH skull lesions usually requires wide surgical resection. These lesions may be attached to dura and subcutaneous tissues and can be friable. These lesions can be persistent and recur. Chemotherapy or low-dose radiation may be considered.

4.3.4 Osteoma and Osteoblastoma

Osteomas are more commonly seen in young adults and rarely in children. These lesions have a distinctive appearance on CT of being hyperintense and expansile. If the lesions are essentially asymptomatic, they are followed. If surgery is performed, the entire lesion is removed and is repaired with a split-thickness graft (Swift and Sacco 2008).

Osteoblastomas are more commonly seen in children than adults and are considered benign. These account for about 1% of benign tumors and about 10–20% of tumors of the skull.

Testing with radiographs may be inconclusive, and a biopsy is usually required. This is an example of a benign lesion that can grow and cause pain or discomfort. As with other such lesions, the entire lesion is removed and replaced with a split-thickness cranial bone graft.

4.3.5 Ewing's Sarcoma

This childhood bone cancer was described as early as 1866 and has been associated with Dr. James Ewing because of his extensive work. This lesion is rarely a primary lesion of the skull, but it frequently metastasizes to the calvaria.

4.3.5.1 Epidemiology

Ewing's sarcoma is the second most common bone cancer found in children, but it is also relatively uncommon. It accounts for about 1% of all childhood cancers. Though seen at any age, it rarely occurs in adults over age 30 years.

4.3.5.2 Evaluation

The most common presenting symptom is pain, which can be intermittent. It is usually worse at night, and the size can fluctuate as well, enlarging when it is more painful. It is associated with leukocytosis, anemia, and fevers.

Radiographic appearance shows destruction of the bone with irregular and poorly defined borders. There is usually an associated soft tissue mass. It may appear to be inflammatory in nature, yet bone destruction is a hallmark feature (Figs. 4.4 and 4.5).

4.3.5.3 Treatment

Identification of the cell type is imperative. The cellular structure is composed of compact and uniform cells with distinct nuclei. The neuropathologist must differentiate this tumor from other sarcomas, as the 5-year survival rate with Ewing's sarcoma is less than 5% where as other sarcomas have better prognoses. The lesion is radiosensitive, so initial treatment includes local radiation therapy along

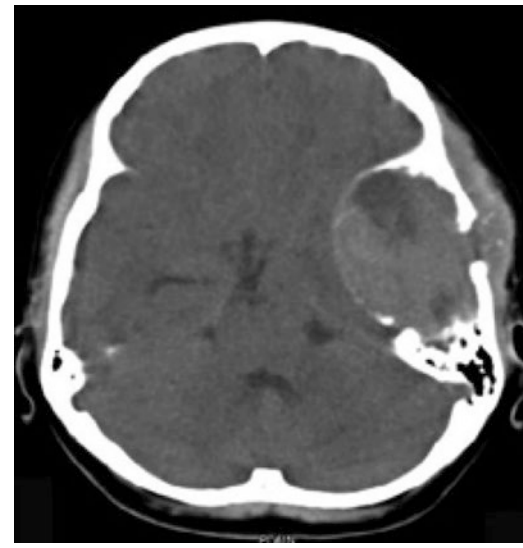


Fig. 4.4 Ewing's sarcoma on CT showing destruction of the bone and painful scalp mass

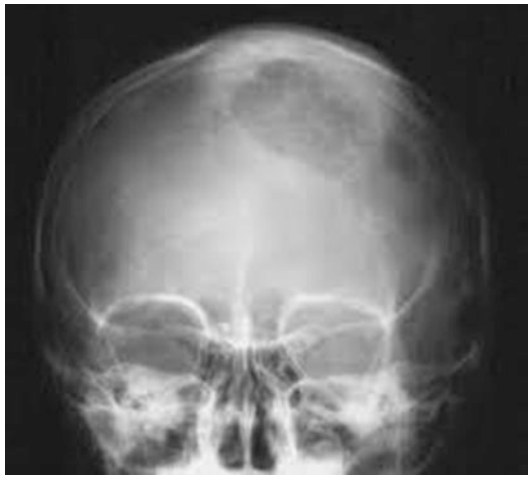


Fig. 4.5 Ewing's sarcoma on skull X-ray showing bony destruction

with systemic chemotherapy. Recurrences are common, and prognosis is poor with any type of treatment (Kieffer et al. 1989). Surgical resection is considered depending on the site and may be performed after chemotherapy.

4.4 Congenital

4.4.1 Cutis Aplasia

4.4.1.1 Epidemiology and Pathophysiology

Cutis aplasia (also known as aplasia cutis congenita) is a rare, congenital, localized skin defect that can occur on any part of the body but is most commonly found on the scalp. Although the exact incidence is unknown, it is believed to occur in approximately 1 in 10,000 births (US National Library of Medicine 2016). There are several etiologic causes for this defect that can be categorized into non-syndrome related and syndrome related.

Non-syndrome-related causes include intrauterine exposure to teratogenic drugs such as methimazole or misoprostol, viral infections in the perinatal period, intrauterine vascular ischemia, and amniotic adhesions. Familial cases have also been reported and are mostly inherited through an autosomal dominant pattern. Mutations in the ribosomal GTPaseBMS1 gene have been linked to cutis aplasia (Wan 2016).

While cutis aplasia is usually isolated, it has also been noted to appear with other malformations and syndromes including Adams-Oliver syndrome, Johanson-Blizzard syndrome, trisomy 13, and Wolf-Hirschhorn syndrome.

Due to the various, multifactorial causes of cutis aplasia, Frieden (1986) proposed a classification system consisting of nine groups characterized by the number, location, and pattern of the defect and the presence of associated malformations and/or genetic components.

4.4.1.2 Evaluation

Cutis aplasia occurs most frequently on the scalp (80% of cases) along the vertex but can also appear on the face, trunk, and limbs. It appears as a well-demarcated ulceration or open wound and sometimes involves the underlying tissue and bone. It varies in size from <1 to >10 cm, as well as in shape. It can appear circular, linear, oval, or stellate in configuration. Defects that occur early in gestation may heal before birth and be covered with a thin, smooth, and atrophic membrane, fibrous tissue, or a parchment-like scar with associated alopecia. Defects that occur later in gestation appear more ulcerative (Wan 2016).

A comprehensive physical examination should be performed to evaluate for any associated malformations or syndromic manifestations. Medical imaging is not usually indicated, unless evaluation of structural lesions that may require neurosurgical intervention is desired (Fig. 4.6).

4.4.1.3 Treatment

Smaller lesions (<3 cm) will usually re-epithelialize spontaneously and sometimes heal with an atrophic scar or an area of alopecia. With these smaller lesions, the treatment plan may include an antibiotic ointment and wet gauze dressing. However larger lesions, or lesions with associated bone and dural defects, are at risk for infection and hemorrhage and require surgical repair. Surgical intervention may include skin rotation flaps, tissue expanders, or skin grafts (Parent and Shiflett 2011). There have been literature reports where the defect can expose the sagittal sinus, in which case immediate surgical intervention is warranted due to the high risk of fatal hemorrhage (Kim et al. 2001).

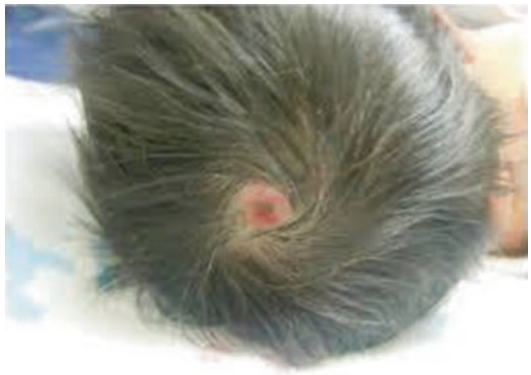


Fig. 4.6 Cutis aplasia of the scalp

4.4.2 Epidermoid and Dermoid Cysts

4.4.2.1 Epidemiology and Pathophysiology

Epidermoid and dermoid cysts are common findings in the pediatric neurosurgical clinic, accounting for 50–60% of all pediatric scalp lesions (Parent and Shiflett 2011). Although epidermoid and dermoid cysts have similar presentations, they differ in their cell makeup. They are both ectoderm-lined inclusion cysts that develop when the surface ectoderm fails to properly separate during neural tube closure. Epidermoid cysts are primarily made up of squamous epithelium and keratin, while dermoid cysts contain hair, sebaceous and sweat glands, as well as squamous epithelium. They can present intradurally or extradurally, but for the purposes of this chapter, we will focus on the extradural lesions.

Epidermoid and dermoid cysts are mainly unilocular and slow growing (Smirniotopoulos and Chiechi 1995). They typically grow to be 1–2 cm by the time of discovery but will continue to grow if not surgically removed. As they grow, they tend to involve more and more of the inner table of the bone and can erode through the bone if not removed.

4.4.2.2 Evaluation

Epidermoid and dermoid cysts are among the most common masses of the scalp and skull. Most commonly, they form along sites of fusion with 50% of them involving the frontozygomatic suture of the orbit (Veselinovic et al. 2010). Upon palpation, they can feel soft and partially compressible, or they can feel hard. This is dependent on the amount of bone bordering the edges of the cyst. They are most commonly nonmobile masses because they are fixed to the underlying bone periosteum. Although diagnosis can usually be made upon physical examination, a skull X-ray or head CT scan may be ordered by the provider as additional preoperative workup. This is helpful with surgical planning to understand how much of the inner

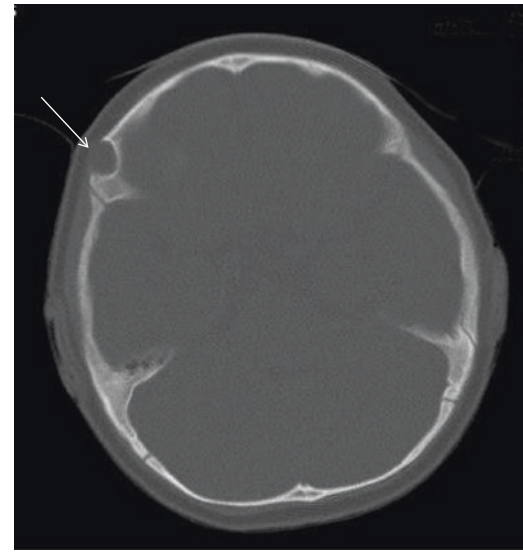


Fig. 4.7 Erosion of the bone by dermoid cyst seen on CT with bone windows

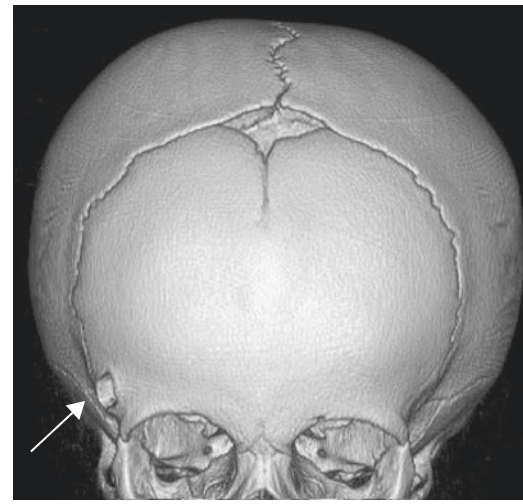


Fig. 4.8 Erosion of the bone by dermoid cyst seen on 3D CT

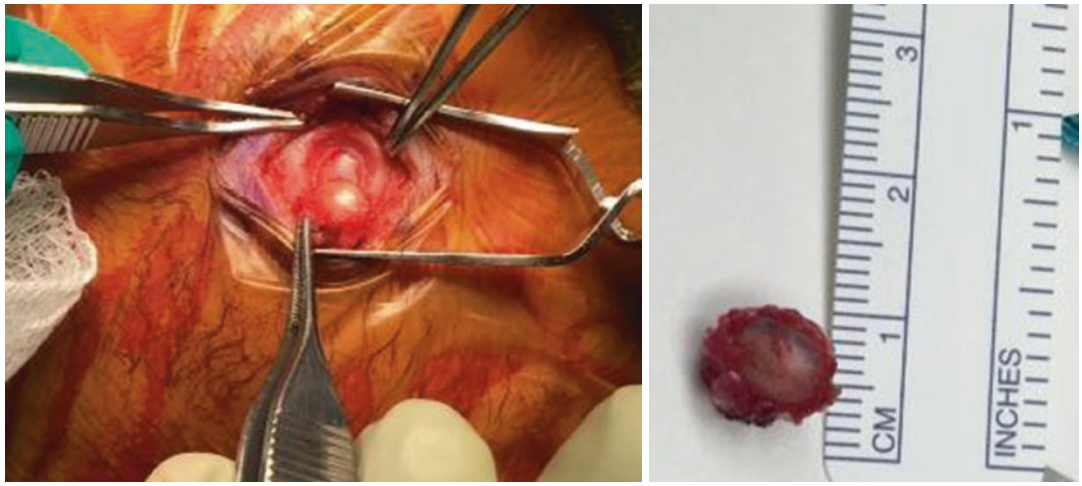


Fig. 4.9 Intraoperative excision of epidermoid

table of the bone is involved if any and if there is intracranial involvement (Figs. 4.7 and 4.8).

4.4.2.3 Treatment

Surgical removal is the treatment of choice with epidermoid and dermoid cysts. If left untreated, they can potentially erode through the skull, and because of their superficial location, they are at risk for injury or rupture. Surgery is done on an outpatient basis and involves little postoperative pain or recovery time. If the cyst is removed intact without rupture, recurrence is rare (Fig. 4.9).

4.4.3 Cranial Dermal Sinus

4.4.3.1 Etiology and Pathophysiology

Dermal sinus tracts are a common pediatric finding that can be present anywhere along the neural axis. Although they are most commonly found in the lumbosacral area, they can also be found on the cranium. Of those that present on the cranium, 85% are found along the midline in the occipital region, while the remainder can be found in the nasofrontal area or, very rarely, the posterior parietal area (Jimenez and Barone 1999) (Fig. 4.10a, b).

Dermal sinuses are formed when there is an interruption in the separation of the neural ectoderm and the epithelial ectoderm during the time of normal midline fusion at 3–5 weeks of gestation. This interruption results in a focal segmental adhesion lined with stratified squamous epithelium. Cranial dermal sinus tracts have variable depth depending on their degree of separation. They may end subcutaneously or extend deeper to include central nervous system structures that reflect its ultimate embryologic level. This would include the fourth ventricle for those found in the occipital level or the crista galli for those found in the nasofrontal area. Of those found in the nasofrontal area, 90% end extracranially (Dias and Partington 2011).

An associated dermoid or epidermoid cyst may form anywhere along the dermal sinus tract but most commonly occur at its terminus. Although rare, if an inclusion cyst does occur intracranially, mass effect and local compression could lead to serious complications including hydrocephalus and signs and symptoms associated with increased intracranial pressure (headache, vomiting, ataxia, nystagmus, papilledema, unsteady gait, etc.).

The most commonly associated risk with cranial dermal sinuses is infection as the tract serves as a constant portal for pathogens to enter. For sinuses that end subcutaneously, an infection may present with localized erythema, edema, or purulent drainage. However, sinuses that extend intracranially are at risk for much more serious infections that lead to meningitis or abscesses, with *Staphylococcus aureus* being the most common pathogen.

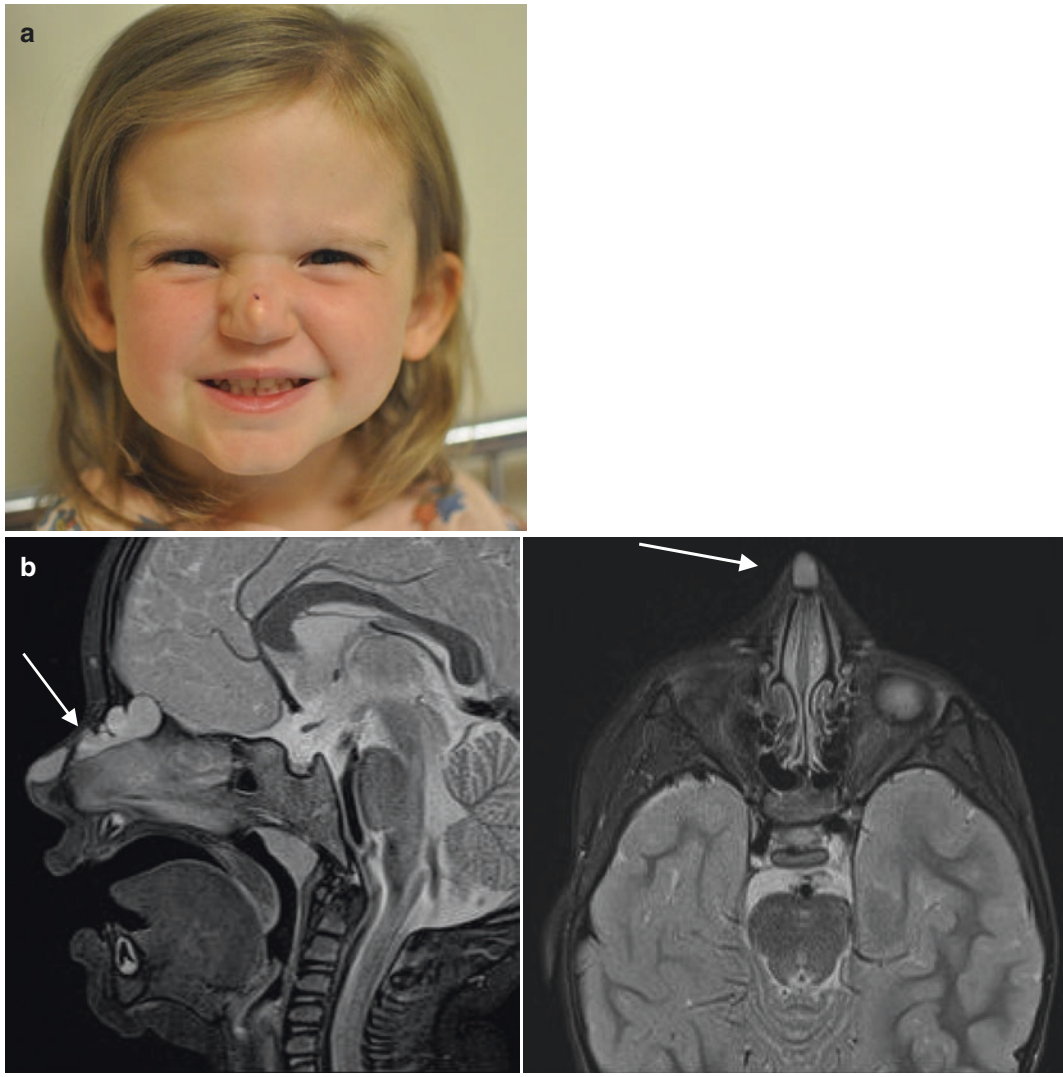


Fig. 4.10 (**a**) A 2-year-old girl with nasal dermoid marked for surgery. (**b**) Sagittal and axial T2 images of nasal dermoid. Note sagittal view of dermoid extending intracranially

4.4.3.2 Evaluation

Cranial dermal sinuses are easily seen when they present on the nasofrontal area and appear as a small dimple. Sometimes there is an associated abnormal hair pattern surrounding the tract and/or clear or yellow drainage. Cranial dermal sinuses located in the occipital or parietal area are more disguised by normal hair growth and may not be discovered on initial routine exam. In fact, it is not uncommon for occipital dermal sinuses to be found only after recurrent unexplained episodes of meningitis have presented. In this case, close examination of the midline of the scalp should be done and may require shaving of the head in order to completely visualize the sinus. Anytime there is a report of an area of hair that is wet without explanation; the area should be shaved and examined for a cranial dermal sinus tract.

A brain MRI is recommended for evaluation of the depth of the dermal sinus tract. If meningitis is suspected, a lumbar puncture is performed.

4.4.3.3 Treatment

Cranial dermal sinuses that end subcutaneously, if able to be kept properly clean, do not require surgical excision. However, sometimes these sinuses become infected or form abscesses. In these cases, surgical excision is recommended to avoid recurrent infections. Cranial dermal sinuses that extend intracranially are surgically explored, and the tract is excised, including any associated cysts. In the presence of meningitis, the patient usually completes a course of antibiotic therapy before surgery is recommended, given there is not declining neurological status.

4.5 Acquired Lesions

4.5.1 Hematomas

A cephalohematoma is a collection of blood between the skull and the pericranium, confined within the borders of cranial sutures. These are sometimes referred to as subperiosteal hematomas. Bleeding elevates the periosteum, and at the time of the injury, the site is more firm. It becomes more ballotable after a few days (Fig. 4.11).

A calcified cephalohematoma is one that and has not resorbed, but rather has hardened in its original form. A calcified hematoma has an inner and outer layer of the bone. The inner layer consists of the fetal inner and outer table of the intramembranous calvarial bone, while the outer layer is made up of sub-pericranial bone (Figs. 4.12 and 4.13).

The most common cause of the cephalohematoma that becomes calcified is the result of trauma associated with an instrument-assisted vaginal birth, occurring in 1–2% of spontaneous vaginal deliveries. These injuries are usually noticed within the first 24 h but can be found up to 72 h after birth. Most of these hematomas spontaneously resorb within 1 month, with 80% of them absorbing within 2–3 weeks (Greenberg 2010). Other causes of cephalohematomas include trauma in childhood (Fig. 4.14).

A caput succedaneum is edema at the neonate's presenting part of the head as a result of pressure against the mother's cervix during labor. The edema in caput succedaneum crosses suture lines. It may involve wide areas of the head or may be the size of a large egg (Fig. 4.15).

A subgaleal hematoma may occur without any bony trauma or may be associated with a linear skull fracture especially in a small child under 1 year of age. It is caused from bleeding into the loose connective tissue that separates the scalp from the periosteum. This type of hematoma typically does cross suture lines. It is not uncommon for the care provider or examiner to not notice much of a hematoma right after the injury, because it is still solid at this point. Within a day or 2, the lesion is noted to be a soft fluctuant mass similar to a "water balloon." These lesions do not calcify, because they occur above the bony layers.

4.5.1.1 Treatment

The nurse caring for an infant or small child with a hematoma needs to be mindful of several things.

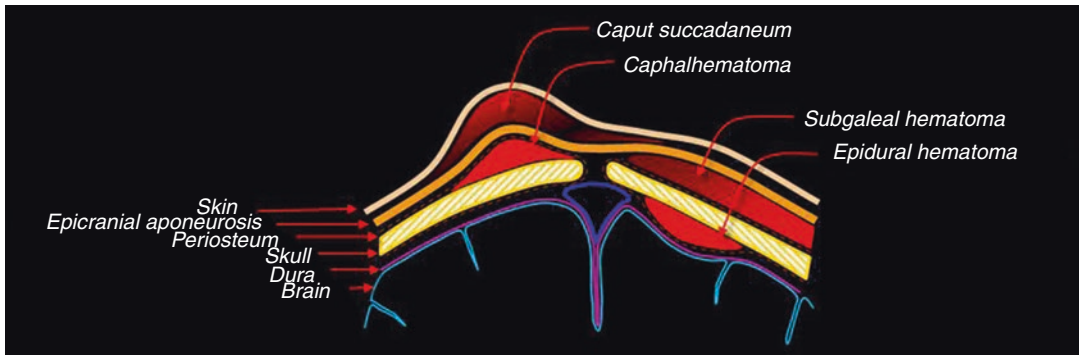


Fig. 4.11 Diagram of various skull injuries and their locations

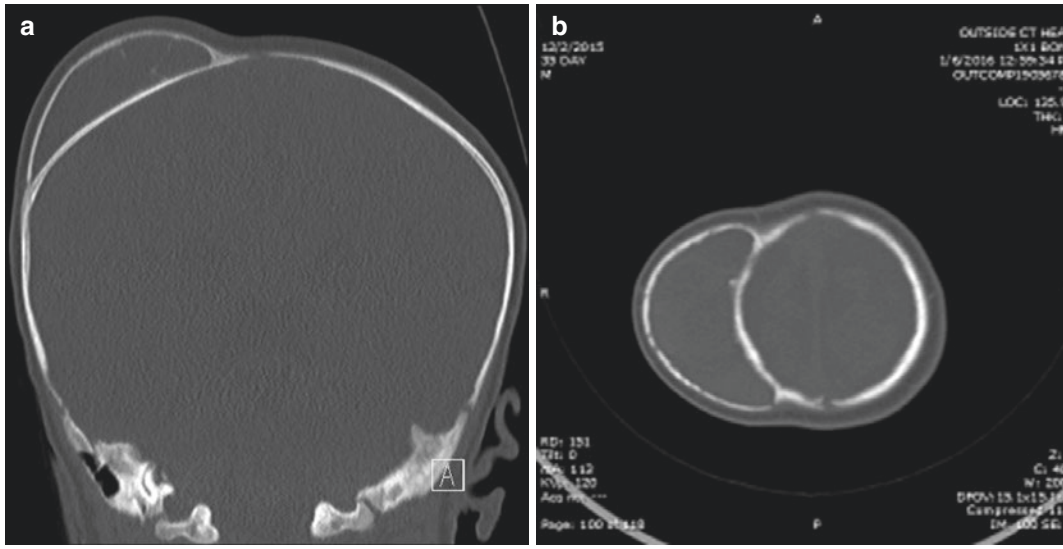


Fig. 4.12 (**a**) Coronal CT with bone windows showing right parietal calcified cephalohematoma. (**b**) Calcified hematoma on CT scan

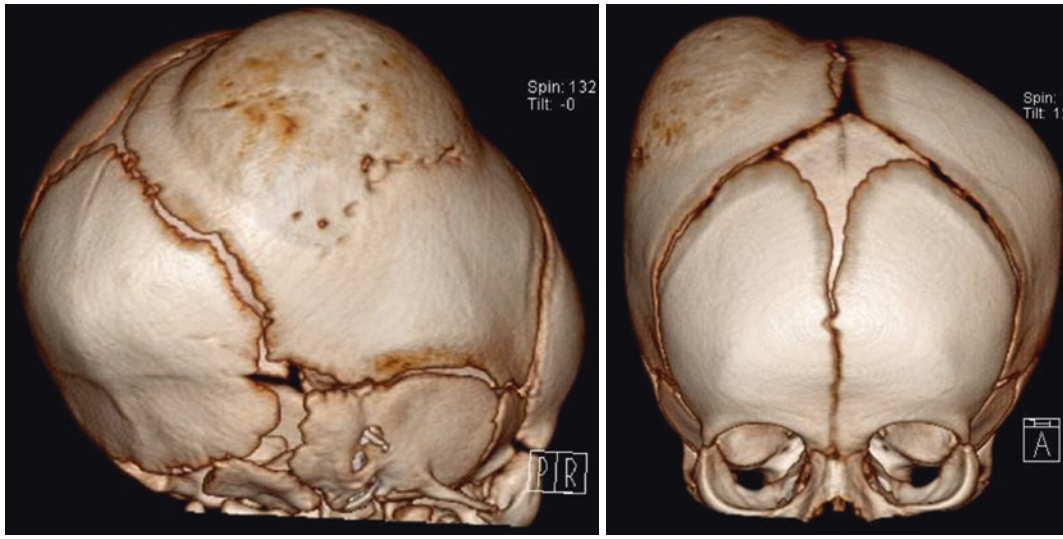


Fig. 4.13 CT head with 3D recon showing calcified right parietal cephalohematoma

First of all, small children have small circulating volumes, so large hematomas can cause hypovolemia and later anemia. In an infant with a large hematoma, one must be mindful of hyperbilirubinemia as the blood is resorbed. This can occur up to 10 days after an injury. Occasionally an infant may require a blood transfusion.

Infection of a hematoma is rare but has been documented. In one case, a 1-month-old female presented with febrile illness and was found to have *E. coli* sepsis. She had a cephalohematoma that was fluctuant, and it also grew *E. coli.* (Weiss et al. 2009). It was presumed the hematoma became infected via hematologic seeding (Fig. 4.16).

One review suggests that there may be an elevated incidence of intracranial hemorrhage (ICH) in babies with cephalohematomas. It was found that 7/19 infants who underwent neuroimaging

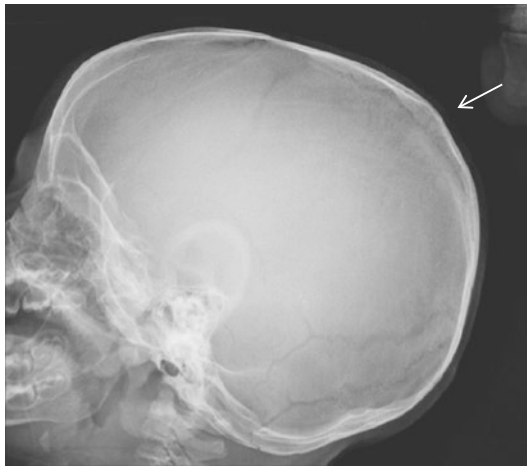


Fig. 4.14 Small calcified hematoma due to trauma

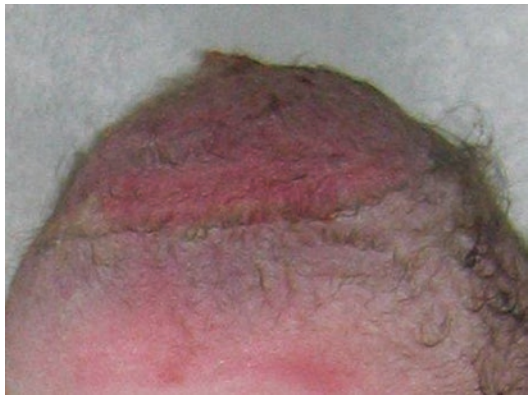


Fig. 4.15 Caput succedaneum

(36.8%) had ICH including two epidural hematomas (Kim et al. 2014).

Sometimes a calcified hematoma can be quite large and unsightly. These children are often referred to the pediatric neurosurgery office. Typically a low-dose CT scan is ordered to make sure that there are no underlying intracranial injuries as well as to document the condition of the skull. The pediatric neurosurgeon then meets with the parents. Many times these lesions need to be surgically excised, and hearing that your baby needs surgery can be quite traumatic. Other lesions are small and do not require surgical intervention.

Occasionally the pediatric office will see an older child with a calcified hematoma that was not operated on in early childhood or infancy. Sometimes the calcified hematoma remains quite large, and the surgeon must decide along with the parents whether or not it is beneficial to have it removed. If small enough and the hair covers it, most likely it will not be removed. However, larger bony defects may require surgery.

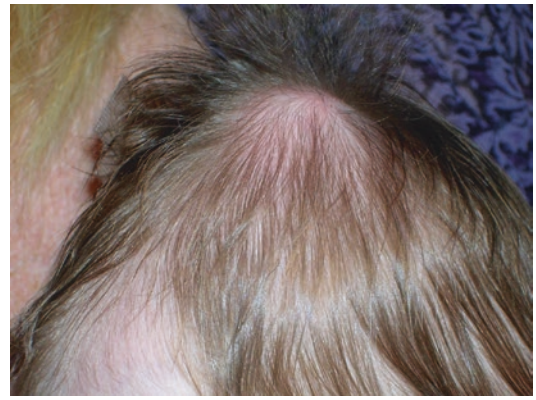
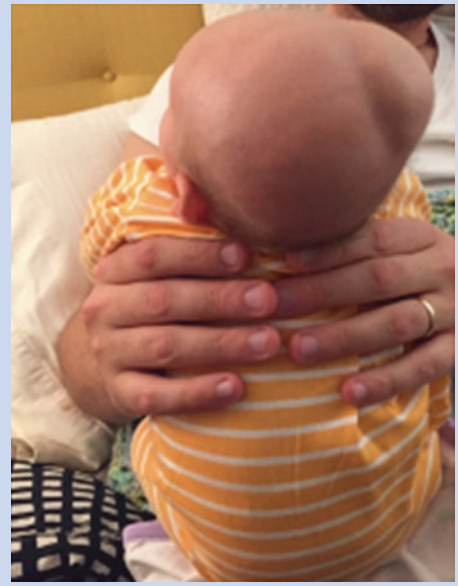


Fig. 4.16 Infected calcified cephalohematoma (Courtesy of Dr. Weiss)

Box 4.1

Baby with calcified cephalohematoma



4.5.2 Growing Fracture of Childhood

An interesting and unusual late complication of skull fractures seen small children is what is known as a growing fracture of childhood. These are also known as growing skull fractures and posttraumatic leptomeningeal cysts (Fig. 4.17). It was first described in 1816 and has been reported to occur in less than 0.05–1.6% of cases. It occurs more commonly in children under age 12 months, but 90% of patients with these complications are under the age of 3 years (Dutcher et al. 2001; Liu et al. 2012).

There are several theories as to the cause. It is known that in some skull fractures, there may be dural tearing and entrapment of the arachnoid membrane or brain tissue within the fracture margin. Some experts also feel that as the rapid growth of the brain and skull occurs, the dura adheres more tightly to the bone and thus is more easily torn when the skull is fractured in a small child. The protruding intracranial contents may prevent the fracture from healing.

4.5.2.1 Treatment

Treatment requires surgery. This includes resection of the leptomeningeal cyst or protruding tissue. The surgeon then performs a meticulous watertight repair of the dura, followed by a cranioplasty. The surgeon is mindful that the child may have increased intracranial pressure as one of the causes of intracranial contents protruding through a fracture. Thus, once the defect is closed, the patient is monitored closely in the pediatric intensive care unit postoperatively.

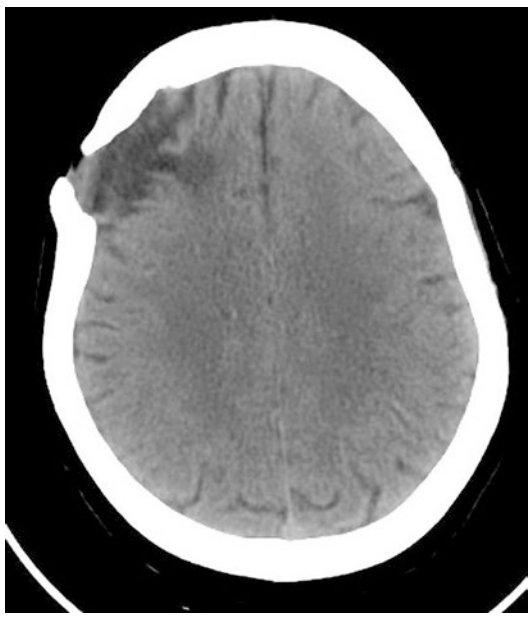


Fig. 4.17 Leptomeningeal cyst

Nursing care for these children would be the same for any craniotomy patient, with close monitoring of vital signs, mindful for signs and symptoms of increased intracranial pressure. Neuro checks are performed regularly, and any change should be brought to the neurosurgeon's attention right away.

4.6 Vascular

Vascular Lesions of the Scalp

Vascular lesions are actually quite common in children, occurring in up to 75% of newborns. We commonly see small pink to red lesions over the face, head, and neck, and these are sometimes known as "stork bites" (nevus flammeus). They are usually small, less than 2 cm, and are not raised. Most of these small lesions disappear within the first or second year of life.

Lesions known as port wine stains contain abnormal blood vessels. These lesions can be quite large and disfiguring. They also do not involute, but rather can continue growing.

Clinical Concerns

As noted above, the smaller lesions usually disappear without intervention. Larger lesions may indicate intracranial abnormalities that include lesions of the cortex. One such diagnosis is Sturge-Weber syndrome. These children can have devastating seizures and developmental delays. Thus, it is imperative that larger lesions are evaluated by the neurology or neurosurgery team. Evaluation would include appropriate radiographs such as MRI and MRA/MRV (Fuchs and Tomita 2001).

4.6.1 Sinus Pericranii

4.6.1.1 Etiology and Pathophysiology

Sinus pericranii is a rare, slow-flow venous anomaly that presents when there is an abnormal communication between extracranial and intracranial venous pathways. More specifically, the communication is usually seen between the sagittal sinus and dilated transosseous emissary veins. The nature of this vascular abnormality may be congenital, spontaneous, or acquired by trauma. Congenital cases are present at birth and reveal an endothelial lining on close pathological examination. Traumatic sinus pericranii usually present after a head injury which results in a skull fracture, a torn emissary vein, or a direct injury to the sagittal sinus. Spontaneous sinus pericranii occur after an intrinsic defect in the skull itself causes erosion through the skull table. These are also thought to be congenital with a late diagnosis in some cases (Bollar et al. 1992; David et al. 1998; St. Clair and McCutcheon 2011).

Although the majority of cases present in young childhood, they can also present in adolescence or adulthood, especially with posttraumatic etiology. Sinus pericranii can present as an isolated incidence or in association with a syndrome such as Crouzon's syndrome, Apert's syndrome, or Hunter's syndrome. Association with craniosynostosis has also been documented in the literature (Nobuyuki et al. 2007) (Fig. 4.18).

4.6.1.2 Evaluation

Sinus pericranii are usually found on the cranial vault at the midline or just off center, proximal to the sagittal sinus. They present as a soft mass or scalp swelling that is easily compressible and painless. They are noted to swell and appear engorged while lying in the recumbent position or during a Valsalva maneuver (such as crying in the infant child) and decrease in size in the upright or erect position. Over time they tend to increase in size; however, there have been some reports of spontaneous regression (Rozen et al. 2008).

The underlying skull defect associated with sinus pericranii is best visualized on a head CT, but a CT venogram is also required to evaluate the communication between the venous pathways. An MRI and MR venogram are also useful in evaluation.

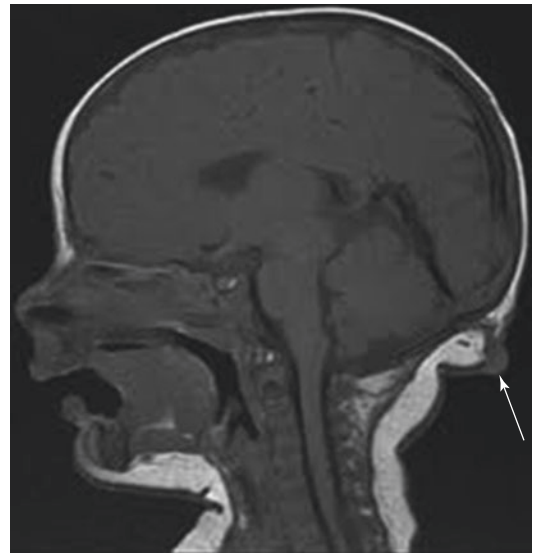


Fig. 4.18 Sinus pericranii

4.6.1.3 Treatment

In the case of progressive disease, surgical resection is indicated. Because of the risk for serious complications such as hemorrhage, infection, or air embolism, some surgeons recommend prophylactic resection (Bollar et al. 1992; David et al. 1998; St. Clair and McCutcheon 2011). Although surgical resection is the most common treatment approach, endovascular embolization has also been utilized in some cases.

4.7 Inflammatory/Reactive

4.7.1 Cranial Fasciitis

4.7.1.1 Etiology and Pathophysiology

This is a rare, benign lesion that is thought to be the result of possible trauma. It is considered a reactionary process. There may be a history of a delivery by forceps or vacuum extraction. The most common site is the temporal bone, followed then by the frontal, parietal, and temporoparietal

area. There is no family history, and the lesion does not return once removed.

The lesion arises from the deep fascia (hence the name), periosteum, or the fibromembranous layer covering the sutures and fontanels. Though most lesions are found in the scalp, there have been a few reported cases of intracranial lesions. The intracranial lesions can arise from the dura without scalp involvement.

4.7.1.2 Evaluation

Most patients, up to 95%, present with an asymptomatic lesion in the scalp. There is almost always a history of trauma, and the lesion can grow rapidly. Radiographs showing erosion of the outer table is a common finding; however, there are a few reported cases where cranial fasciitis has penetrated the calvarium through to the epidural space. There are also a few reports of intracranial lesions found exclusively intracranially after head trauma or tumor removal (Halder et al. 2011). These tend to be slow-growing lesions and are thought to be the result of altered blood supply to lesion within the cranium.

4.7.1.3 Treatment

These scalp and skull lesions are totally resected, and intracranial lesions require surgical planning depending on the location.

Conclusion

Unusual new masses found on a child's scalp or skull usually eventually referred to the Pediatric Neurosurgery service. Though only a few may be malignant, even benign lesions can grow and should be evaluated by the neurosurgeon for definitive treatment.

Pearls

- Unresolving cephalohematomas should be referred to neurosurgery for early treatment to prevent calcification.
- Cranial dermal sinus should be included in the differential diagnosis of any patient with unexplained meningitis and/or an area of wet hair that cannot be explained. Sometimes shaving of the head must be done before the sinus is visualized.
- Any lesion or mass that is noted to progress or grow in size should be referred to the neurosurgical clinic for further evaluation.
- Neuroscience nurses caring for infants with scalp hematomas should be mindful of possible anemia and hyperbilirubinemia.

References

Amit M, Collins MT, FitzGibbon EJ, Butman JA, Fliss DM, Gil Z (2011) Surgery versus watchful waiting in patients with craniofacial fibrous dysplasia: a meta-analysis. Database of abstracts of reviews of effects, 2015 Issue 2. University of New York, Wiley. http://onlinelibrary.wiley.com/o/cochrane/cldare/articles/DARE-12011006500/sect0.html

Bollar A, Allut A, Prieto A, Gelabert M, Becerra E (1992) Sinus pericranii: radiological and etiopathological considerations. J Neurosurg 77(3):469–472

David L, Argenta L, Venes J, Wilson J, Glazier S (1998) Sinus Pericranii. J Craniofac Surg 9(1):3–10

Dias MS, Partington MD (2011) Normal and abnormal embryology of the brain. In: Winn HR (ed) Youmans neurological surgery. Elsevier Saunders, Philadelphia, pp 1883–1897

Dutcher A, Sood A, Ham S, Canady A (2001) Skull fractures and penetrating brain injury. In: McLone, Marlin, Scott, Steinbok, Reigel, Walker, Cheek (eds) Pediatric neurosurgery. Saunders, Philadelphia

Frieden IJ (1986) Aplasia cutis congenital: a clinical review and proposal for classification. J Am Acad Dermatol 4(14):646–660

Fuchs HE, Tomita T (2001) Neurocutaneous syndromes and meningiomas of childhood. In: David M (ed) Pediatric neurosurgery. 4th edn. Saunders, Philadelphia, p 778

Greenberg MS (2010) Handbook of neurosurgery. Thieme, New York, p 918

Greenberg MS (2014) Handbook of neurosurgery. Thieme, New York, p 701

Halder A, Greene C, Rivard D, Shao L (2012) Cranial fasciitis presenting as an intracranial mass in a 10 -year-old girl. Pediatr Dev Pathol 15:146–150

Hersh JH, Committee on Genetics (2008) Health supervision for children with neurofibromatosis. Pediatrics 121:633

Jimenez D, Barone C (1999) Encephaloceles, meningoceles and dermal sinuses. In: Albright, Pollack,

Adelson (eds) Principles and practice of pediatric neurosurgery. Thieme, New York, pp 202–206

Kim CS, Tatum SA, Rodziewicz G (2001) Scalp aplasia cutis congenita presenting with sagittal sinus hemorrhage. Arch Otolaryngol Head Neck Surg 127(1): 71–74

Kim HM, Kwon SH, Park SH, Kim YS, Oh KW (2014) Intracranial hemorrhage in infants with cephalohematoma. Pediatr Int 56:378–381 (Official journal of the Japan Pediatric Society)

Liu X, You C, Lu M, Liu J (2012) Growing skull fracture stages and treatment strategy. J Neurosurg Pediatr 9:670–675

McClain KL (2016) Clinical manifestations, pathologic features, and diagnosis of Langerhans cell histiocytosis. In: Boxer L, Park J (eds). Wolters Kluwer, UpToDate. Retrieved from http://www.uptodate.com/home

Nobuyuki M, Kaneshige S, Takashi H, Yoshihiko F, Tadayuki S, Tetsuji U, Yoshiaki H (2007) Sinus Pericranii associated with craniosynostosis. J Craniofac Surg 18(1):78–84

Parent AD, Shiflett JM (2011) Skull tumors and fibrous dysplasia. In: Winn HR (ed) Youmans neurological surgery. Elsevier Saunders, Philadelphia, pp 2136–2143

Pollack I (2008) Neurofibromatosis 1 and 2. In: Albright, Pollack, Adelson (eds) Principles and practice of pediatric neurosurgery. Thieme, New York, pp 735–736

Rozen W, Joseph S, Lo P (2008) Spontaneous involution of two sinus pericranii- a unique case and review of the literature. J Clin Neurosci 15(7):833–835

Smirniotopoulos JG, Chiechi MV (1995) Teratomas, dermoids, and epidermoids of the head and neck. Radiographics 6(15):1437–1455

St. Clair EG, McCutcheon IE (2011) Skull tumors. In: Winn HR (ed) Youmans neurological surgery. Elsevier Saunders, Philadelphia, pp 1667–1691

Swift D, Sacco D (2008) Scalp and Skull neoplasms. In: Albright, Pollack, Adelson (eds) Principles and practice of pediatric neurosurgery. Thieme, New York, p 484

Swift D, Trumble E (1999) Scalp and skull neoplasms. In: Albright, Pollack, Adelson (eds) Principles and practice of pediatric neurosurgery. Thieme, New York, p 450

U.S. National Library of Medicine (2016) Genetics home reference: nonsyndromic aplasia cutis congenita. Retrieved from https://ghr.nlm.nih.gov/condition/nonsyndromic-aplasia-cutis-congenita#statistics

Veselinovic D, Krasic D, Stefanovic I, Veselinovic A, Radovanovic Z, Kostic A, Cvetanovic (2010) Orbital dermoid and epidermoid cysts: case study. Serbian Arch Med 138(11–12):755–759

Wan J (2016) Aplasia cutis congenita. Retrieved from http://emedicine.medscape.com/article/1110134-overview#a4

Weiss KJ, Edwards MS, Hay LM, Allen CH (2009) *Escherichia coli*-infected cephalohematoma in an infant. Clin Pediatr 48(7):763–766