Dermoid Tumors

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52.1 Introduction

Dermoid tumors are congenital, nonneoplastic, inclusion lesions which have epidermal capsule and contain dermal appendages such as hair and sebaceous glands. They can occur anywhere along the neuraxis. In the literature, they are often referred to as "cysts" which refers to their histological structure and the presence of secretions (sebum) within the tumor, as well as indicates a nonneoplastic nature of these lesions. Although benign biologically, they can have malignant behavior due to their location causing mass effect and compression of vital structures or having potentially fatal complications such as abscess, spontaneous rupture, and hydrocephalus [1–4]. Remak [5] was the first to suggest displacement of epithelial rests and defective closure of the neural tube as the cause for development of these lesions. The first description of an occipital dermal sinus connected to a dermoid tumor is attributed to Olge [6] in 1865. Later in 1897, Bostroem [1] suggested ectopic inclusion of dermis and epidermis during embryonic development as the cause for formation of dermoids and epidermoids, respectively. This theory was further supported by Bailey in 1920 and later Citchley and Furguson in 1928 who further proposed that dermoids and epidermoids were the result of fetal inclusion of epidermal cells, depending on the depth of the layer or according to embryonic age [1].

Logue and Till [7] classified posterior fossa dermoids into four groups depending on extradural and interadural location with absent, partial, or complete dermal sinus connection. The significance of this classification was the risk of infection. This classification is not in common use.

52.2 Epidemiology

Dermoid cysts in pediatric age group are often reported in association with dermal sinuses and also in larger series are collectively reported with epidermoids. These tumors are generally rare accounting for 0.1–0.7 % of all intracranial tumors [8]. Epidermoids and dermoids together account for approximately 0.7–1.8 % of all intracranial masses, with epidermoid 4–10 times as common [1, 9–11]. Dermoids are usually reported in younger age, in the first decade of life, while epidermoids classically present later in adulthood [10, 12–14]. Dermoids are reported to affect both sexes equally or with slight male predominance [9, 13, 15]. Twelve out of 19 pediatric patients in

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Caldarelli et al. series were male [2]. This group reported the first large pediatric series of intracranial dermoids and epidermoids. Of the 19 patients they treated over 20-year period, 16 were dermoids and only 3 were epidermoids and the mean age of 5.5 years [2]. On the other hand, Fornari et al. [5] also reported on 36 cases of CNS dermoids and epidermoids in patients younger than 20 years of age and found only 3 dermoids out of 17 intracranial lesions of which one was infratentorial. The mean age was 16 years, with slight male predominance (20–16) [5]. Lunardi et al. [13] reported on dermoids and epidermoids of posterior fossa exclusively. In their series, they had 16 patients, 9 of whom had dermoids, with average age of 6.4 years, while the average for epidermoids was 40 years and no gender difference. These findings are summarized in Table 52.1.

The incidence of congenital dermal sinus is reported approximately 1 in 2,500–3,000 births. These are mostly in the lumbosacral region, and only a small percentage is found in the occipital region [16].

52.3 Embryology

Embryologically dermoid tumors are thought to result from inclusion of ectodermal elements at the time of the closure of the neural groove during the third to fifth embryonic life [1, 5, 10, 17, 18]. This is the same as epidermoid as opposed to teratomas where mesoderm also is involved.

In terms of time profile for development of dermoid tumors in relation to epidermoids, the current theory suggests that these lesions occur earlier in embryonic life than epidermoid tumors. Therefore, displaced cells lie near midsagittal plane and as sometimes noted are associated with defect in closure of overlying skin [5, 10]. This is in contrast to epidermoids which most often occupy a more lateral position, in particular in intracranial location. This is postulated to result either from inclusions of ectoderm at a later stage of embryogenesis or displacement during formation of secondary cerebral vesicles, i.e., the otic and optic vesicles [1]. On the other hand, lateral displacement of the primitive ectodermal and mesenchymal cells caused by the developing cerebral vasculature along Virchow-Robin spaces gives rise to laterally positioned dermoids [2, 6, 12].

The congenital dermal sinus that often is found in association with dermoids suggested to represent abnormal adhesion between the skin and neural ectoderm during development [9, 15] (Fig. 52.1a). This tract, depending on its degree of incomplete separation, may end up in subcutaneous tissue, bone, dura, subdural space, or extend at any length into the parenchyma and ultimately within the fourth ventricle. The dermal sinuses can be blind or connect to dermoid [15] (Fig. 52.1c). Rarely there may be more than one dermal sinus present [15].

In recent years, concurrent finding of a rare phenomenon of vertebral anomalies, namely, Klippel-Feil syndrome, with posterior fossa dermoids, has led some authors to propose theories on common embryological process, which suggests dermoid tumors being a late embryonic developmental anomaly [19–21]. It is proposed that this may be related to under-expression of *pax* gene that leads to segmentation failure of cervical somites that alters tissue tension at craniocervical junction leading to entrapment of dermal elements and development of posterior fossa dermoids [20, 21]. On the other hand, some have suggested that segmentation failure affects cervical flexure formation which in turn reduces fetal movements at rhombencephalo-cervical junction ultimately trapping ectodermal elements and leading to formation of dermoid tumors [19]. These theories remain unproven.

Other rare concurrent anomalies such as Dandy-Walker syndrome, callosal agenesis, and diastematobulbia in association with posterior fossa dermoid cyst have also been reported, but the significance of these anomalies remains to be explained [2, 22].

Table 52.1 Major series of mixed posterior fossa lesions

Series	Number of patients (M:F)	Age range (mean)	Pathology	Location in PF	Outcome/complication	Recurrence/reoperation	f/u period (mean)
Schijman et al. [15]	7 2:5	2–6 years	2 dermoids 5 epidermoids (1 + DS)	PF	2 deaths (1 dermoid, 1 epidermoid), 1 severe am	–	–
Yasargil et al. [12]	35 21:14 8(6:2)	18–62 years (37) 19–53 years (36)	Epidermoid Dermoid	10 ST, 22 CPA, 3 IV vent 7 ST, 1 in IV ventricle	8 am, 2 bm, 7 transient deficits 2 deaths (epidermoids)	No symptomatic recurrences	1 month–20 years
Fornari et al. [5]	17 9:8 (of 36 total)	6 months–20 years (16 years)	3 dermoids 14 epidermoids	5 PF	1 death (CPA)	2 reoperation	6–29 years (13)
Lunardi et al. [13]	16	(6.4 years) (40 years)	9 dermoids (3 with DS) 7 epidermoids	2 in IV vent 6 in IV vent	3 deaths (all epidermoid), 1 am (epidermoid)	1 recurrence (epidermoid)	(17.3)
Martinez-Lage et al. [6]	3 (0:3)	6 months–2 years	Dermoid	Extradural	Severe sinus hemorrhage	nil	1–12 years
Caldarelli et al. [2]	19 12:7	3 months–16 years (5.5 years) 6 (<1 year)	16 dermoids (12 + DS) 3 epidermoids	5 CM, 2 IV vent. 1 PMJ	No deaths	2 recurrences (1 dermoid, 1 epidermoid)	8–237 months (92 months)
Zuccaro et al. [37]	30 12:18	1 month–18 years (13 years)	8 acoustics 6 meningiomas, 2 astrocytomas, 3 arach. cysts, 2 epidermoids, others	CPA	7 deaths (6 with recurrences), 3 VPS	6 recurrences	1–12 years

CPA cerebellopontine angle, *am* aseptic meningitis, *PF* posterior fossa, *ST* supratentorial, *PMJ* pontomedullary junction

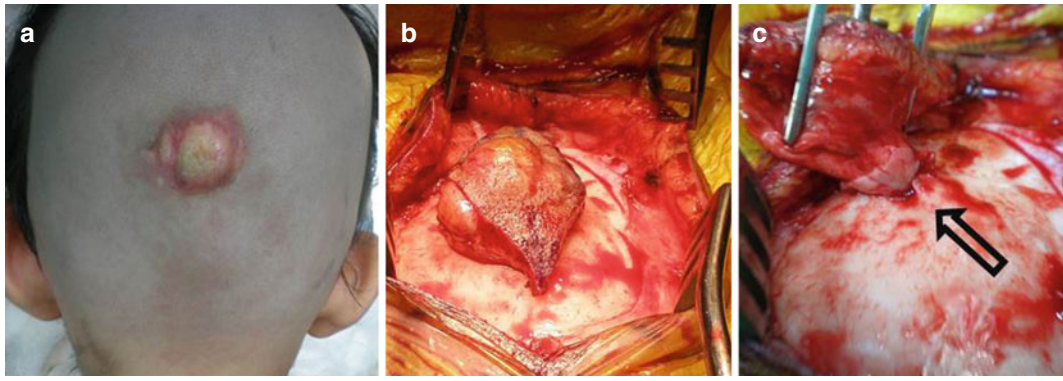

Fig. 52.1 (**a**) An infected occipital dermal sinus, (**b**) Intra-op image showing skin flap incision around the dilated dermal sinus dilatation, (**c**) Entrance of the dermal sinus into the cranium (*arrow*)

52.4 Pathology

Most pediatric intracranial dermoids arise in the posterior fossa, in midline position, i.e., cerebellar vermis and adjacent meninges being more common locations [1, 2, 18]. These tumors are also found in the cavity of the fourth ventricle [2, 13, 18], although this is not the most common location within the posterior fossa. Of the 16 pediatric dermoids in one study, 8 (50 %) were in the posterior fossa with the cisterna magna as the most common location [2] (see Table 52.1). Intra-axial location, cerebellum or brainstem, is exceptionally rare [2, 23, 24]. Intracranial extradural dermoids are also reported as rare [6]. The significance of this location becomes apparent in the review by Martinez-Lage et al. [6] where four out of nine reported cases were closely related to venous sinuses with three intraoperative hemorrhages. The other favored intracranial location is in the parasellar region [10]. Interestingly in contrast to teratomas, dermoids rarely are reported in the pineal region [10].

Grossly well-circumscribed, opaque, oval, or round multi-lobulated masses are generally well demarcated from surrounding tissues. The wall is formed by a fibrous material that varies in thickness and degree of adherence to the surrounding structures [1, 10]. Often there is a layer of reactive gliosis in the adjacent brain tissue. Calcification may be seen in the wall of the cyst [10]. The material within the cyst can vary between a dense (sometimes described as "cheesy") substance and a more brown, mucoid consistency fluid. The presence of hair is common within the tumor [5]. Pus-like fluid due to sebaceous secretions and desquamation, to a more yellowish-brown mucoid fluid, has also been described. Teeth are rarely seen. Some investigators have reported more than one cyst in the same location [10]. Dermoid tumors grow faster than epidermoids [9]. This is due to secretions as well as desquamation process. This may also explain their earlier presentation in life.

The presence of dermal sinus in the occipital region is an important clue to the underlying dermoid cyst and considered by some a diagnostic feature [10]. The importance of the dermal sinus lies in the fact that the open connection between the skin surface and the cyst is a direct route for bacterial infection and thus the source for recurrent meningitis or abscess formation. Most tumors are associated with a complete or incomplete dermal sinus; however, the IV ventricle tumors may not have an associated sinus [2, 18]. In McComb's experience, the lesions involving the region of the IV ventricle have all been dermoids and all been associated with dermal sinus [9]. In another study, 89 % of dermal sinuses are associated with an inclusion tumor and were mostly dermoid tumors, with 18 % only found extradurally, and the rest were intradural [25]. The congenital dermal sinuses associated with dermoid tumor have both dermal and epidermal elements, and it can sometimes regress to a

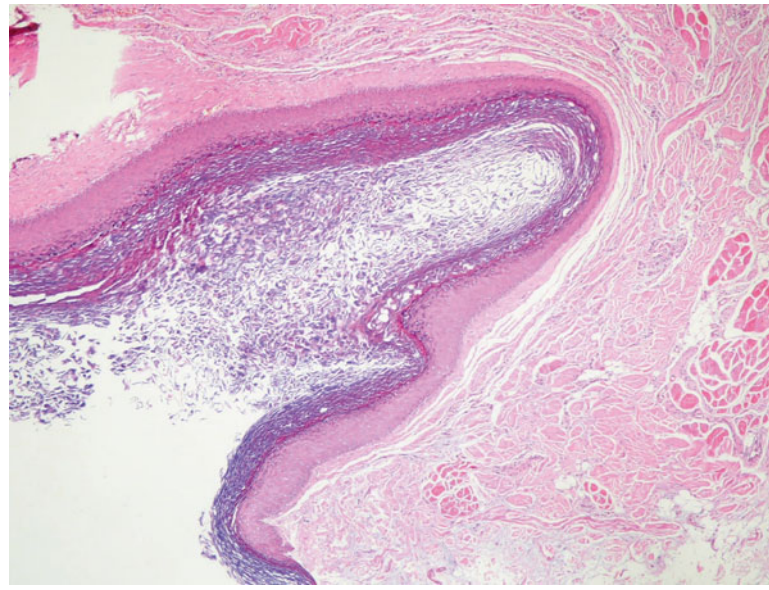

Fig. 52.2 Dermoid cyst lined by stratified squamous epithelium (HE, ×40)

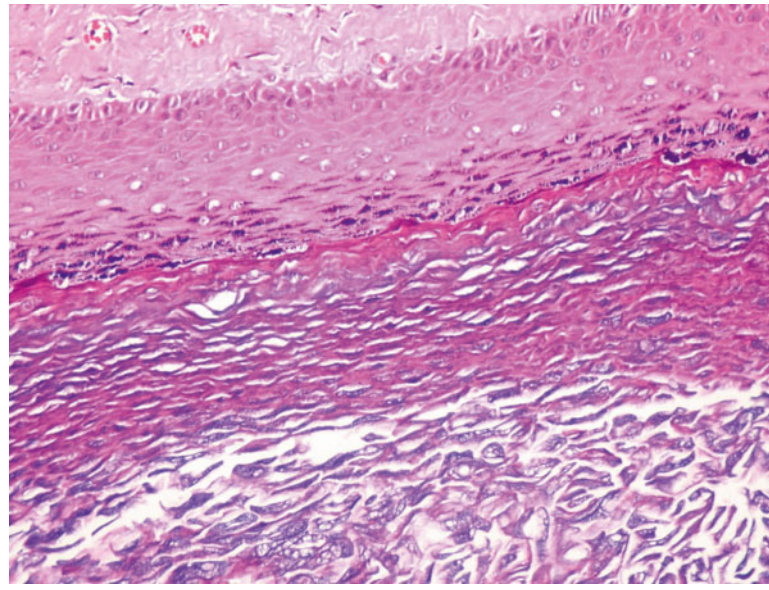

Fig. 52.3 Dermoid cyst lined by cornified epithelium has a distinct granular layer and contains anucleate squames (HE, ×200)

connective tissue band or a nodule. There may be enlargements along the tract forming other dermoid cysts [9]. The opening of the tract is usually very small and difficult to see without shaving the hair on occipital region [18].

Microscopically, the wall of the dermoid cyst (or the capsule of the tumor) is formed by stratified squamous epithelium as seen in the skin and includes hair follicles and sebaceous and sweat glands [1] (Figs. 52.2 and 52.3). Much of the lining of the cyst may be simple squamous epithelium supported by collagen, similar to epidermoid tumors, while in the thicker parts it is supported by dermis and its appendages [10, 17]. It is important to emphasize that dermoid cysts do not contain *fat cells* per se as this is of mesodermal origin, and the lipid metabolites seen are the result of breakdown products of hair and glandular secretions [26].

52.5 Clinical Manifestations

Symptoms from dermoid tumors generally result from mass effect, infection, inflammation, or occasionally hydrocephalus as presenting symptom [9, 15, 27–29]. Recurrent hyperpyrexia without meningism was the presenting symptom in 4 out of 8 pediatric posterior fossa dermoids, while only one presented with aseptic meningitis [2]. Another common finding in pediatric age is localized swelling secondary to inflammation and infection which may or may not be accompanied by discharge from the dermal sinus. These symptoms are often reported for one or more episodes before patient is brought to attention [2, 9].

Recurrent bacterial meningitis is thought to result from bacterial entry via the dermal tract as the port of entry into the subarachnoid space [9]. Recurrent septic meningitis should alert the physician to the possibility of dermal tract existence [10, 15, 18, 30]. The usual infecting organism is *Staphylococcus epidermidis*, but other organisms such as *Klebsiella*, *Proteus* species, pneumococcus, and even *E. coli* have been reported [15, 18, 30, 31].

Cerebellar abscess is a rare and serious sequela of posterior fossa dermoid cyst with dermal sinus which may or may not be associated with bacterial meningitis [9, 29] (Fig. 52.4a–c). This will result in significant inflammation and more rapid rise in size of the lesion and consequently more significant mass effect, exacerbating the symptoms of posterior fossa mass. Akhddar et al. [29] in their review of literature between 1943 and 2001 identified 14 such cases.

Hydrocephalus may be obstructive due to mass effect within the IV ventricle or communicating secondary to arachnoiditis caused by repeated bouts of meningitis [9, 15, 18, 30]. Although hydrocephalus due to mass effect is a late-onset phenomenon, these lesions are slow growing and the surrounding brain tissue accommodates them until they reach a large size [9].

The rare intra-axial brainstem location is reported in association with intermittent cranial nerve palsy probably secondary to transient ischemic attacks, which is thought to be due to spillage of cyst contents within the CSF spaces [32].

Spontaneous or traumatic rupture of the cyst has been reported, which can present with mild to severe neck pain and headache, confusion, and decreased level of consciousness [1]. Histologically, rupture can cause severe granulomatous meningitis with formation of foreign body giant cell reaction to the contents of the cyst and hydrocephalus [3, 10, 33]. This is well demonstrated with the presence of fat droplets within the ventricles on MRI [3, 33]. The cause of spontaneous rupture is not clear. However, El-Bahy and his colleagues [4] reported two cases

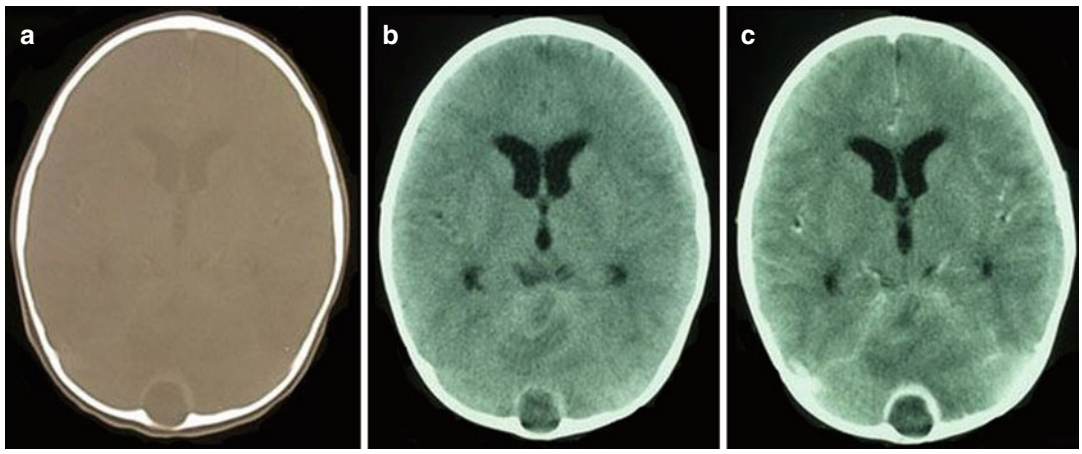

Fig. 52.4 CT images of an occipital dermoid. (**a**) Bone window study demonstrates scalloping of the occipital bone, (**b**) non-contrast CT, (**c**) contrast enhancement of the cyst wall due to inflammation

(16- and 18-year-olds) of spontaneous rupture and reviewed 49 reported cases of ruptured dermoid cyst in the literature. They supported Stendel's theory [34] that the spontaneous rupture can be explained by age-dependent hormonal changes which leads to increase secretion within the cyst and hence faster enlargement and subsequent leakage. The presence of fat within the ventricles and subarachnoid space causes hydrocephalus, meningitis, vasospasm, and cerebral ischemia [4].

52.6 Diagnosis

52.6.1 History and Examination

History of recurrent meningitis, in particular aseptic meningitis, or discharge from a pimple-like lesion on the head which is not healing should raise suspicion. Furthermore, the symptoms of intracranial hypertension, posterior fossa mass (e.g., nausea, vomiting, unsteady gait, nystagmus), as well as cranial nerve palsies should be sought [1, 6, 12]. The natural history of these tumors consists of slow growth which eventually will lead to hydrocephalus, seizures, dysarthria, and dementia [1].

Protracted history is reported in most series with the average length to presentation of 6.8 years in one series, with only one of eight patients presenting in less than 1 year [12].

The slow presentation of these lesions is because of a linear slow rate of growth as opposed to neoplastic lesions that have exponential growth [1]. The duration of symptoms before the era of modern imaging as reported often extended for several years. However, symptoms can also rarely present more acutely as a result of development of hydrocephalus or meningitis. Rupture of a cyst and release of cholesterol-rich contents can cause severe inflammatory reaction, leading to chemical meningitis, acute brain swelling, and vascular compromise.

On the other hand, in pediatric series the duration of symptoms was commonly short, with average of only 1.5 months in one series [13]. In this series, most common presenting symptoms were due to raised ICP followed by bacterial meningitis [13].

Inspection of scalp may reveal the opening of the dermal sinus that is often very small and may be associated with other cutaneous markers such as protruding hair from the opening, capillary telangiectasis, abnormal pigmentation, or increased subcutaneous fat tissue. The presence of abnormal hair pattern is more indicative of rudimentary encephalocele rather than a dermal sinus. There may also be evidence of infection such as skin inflammation or discharge [9]. Gentle pressure around the dermal sinus can help detection of discharging material; however, probing the sinus or injecting the tract (e.g., with contrast material) must be avoided as it can introduce bacteria [9].

52.6.2 Imaging

Plain skull films may reveal an occipital bony defect with densely sclerosing margins and a hypodense tract, although a small tract going obliquely in posterior fossa can be difficult to detect [9, 28]. The films are normal in the absence of dermal sinus. Furthermore, if any evidence of a lesion is seen, further examination with CT or preferably MRI is required. Therefore, the use of plain x-rays is no longer appropriate when other methods are available.

Dermoid tumors usually have low density on CT in keeping with their fatty content (Fig. 52.5b). The contents however more typically have density between that of fat and CSF [18, 35], although rarely isodense or hyperdense lesions have been reported [12, 15, 35, 36]. Enhancement of the wall is uncommon and is thought to be due to the presence of inflammation [15, 35] (Fig. 52.5c). Vasogenic edema is almost never seen in an uncomplicated cyst [36]. In some instances, the contents can have CSF density; hence, the cyst can look similar to normal CSF spaces or an arachnoid cyst if large enough [9]. The presence of calcification within the wall of the cyst is also commonly demonstrated on CT scans [35]. CT scans can also demonstrate scalloping of the cranial bone and the presence of hydrocephalus [9] (Fig. 52.5a), and in cases with ruptured cyst hypodense lipid droplets within the subarachnoid space or ventricles have been demonstrated [33].

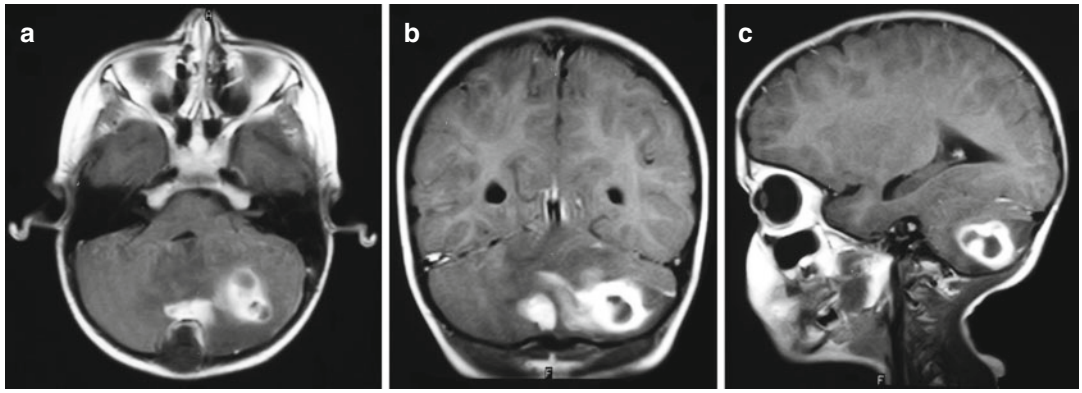

Fig. 52.5 MRI images showing a complicated occipital dermoid at the torcula with associated cerebellar abscess and significant amount of edema involving the cerebellar hemispheres

Although CT scans can demonstrate the lesion, it cannot help in distinguishing it from other lesions, and therefore the next step would be MR imaging. Thus, in cases where history and/or examination suggests this pathology, MRI scan with various sequences can assist in making the diagnosis and therefore is the investigation of choice as first step [2].

Uncomplicated dermoid tumors appear as well-circumscribed lesions with no vasogenic edema, hyperintense on T1W, and hypo- to hyperintense or heterogeneous on T2W images [2, 35, 36] (Fig. 52.6a–e). Fourth ventricular lesions have been reported with mixed hypointensity on T1W [28].

Fat suppression sequence is very useful for detection of lipid, after rupture or within the lesion for establishing a diagnosis [4, 35]. The fat droplets can be detected by hyperintense signal within the cyst [35]. Another useful MR sequence is the diffusion-weighted images (DWI). With this technique, epidermoid tumors show "restricted diffusion" (high signal), while dermoids do not.

Angiography is not proven useful in investigation of these lesions as they are avascular, and it will only confirm the presence of space-occupying lesion.

The differential diagnosis for dermoid cysts includes atretic cephalocele, arachnoid cyst, and low-grade gliomas [2, 23, 28]. Atretic cephalocele lesions which may look similar to subgaleal dermoids enhance with contrast, while dermoids usually do not, and MRI sequences as described above should distinguish these lesions from a glioma or an arachnoid cyst.

52.7 Management

Once the diagnosis is established, the surgical intervention with complete removal is the definitive treatment. These tumors are not radiosensitive and have risk of recurrence if partially removed [2, 9, 12, 18]. Although it is not an urgent issue in an uncomplicated dermoid tumor (e.g., no hydrocephalus, severe compression, or abscess), no significant delay should intervene, in particular when associated dermal sinus is present, due to risk of infection [9].

Complicated lesions presenting with infection, abscess, or ruptured cyst require urgent surgery. Patients with meningitis must be treated with broad-spectrum antibiotics covering for gram-positive cocci with optimal CSF penetration, and if abscess is present, anaerobic cover must be added [16]. In cases of ruptured cyst, most authors suggest prompt surgery and washout of subarachnoid and ventricular spaces (some with hydrocortisone solution) and insertion of

ventricular drain [4]. In the presence of abscess and mass effect, urgent surgical intervention with appropriate antibiotic cover is recommended.

In relation to dermal sinuses, it is important to note that when a dermal sinus is detected, the presence of the tract or bony defect can still be missed on CT or MR imaging, and hence the sinus must be explored with preparation for posterior fossa exploration. In these cases, if no bony defect is detected, this excludes intracranial extension [9].

Repeated infections can cause increased adhesions and make the removal even more difficult. Hence, prompt surgery is advisable upon discovery of the sinus to prevent further infection morbidity and adhesions from surgical point of view. One should plan for a major posterior fossa exploration once dermal sinus is discovered [18].

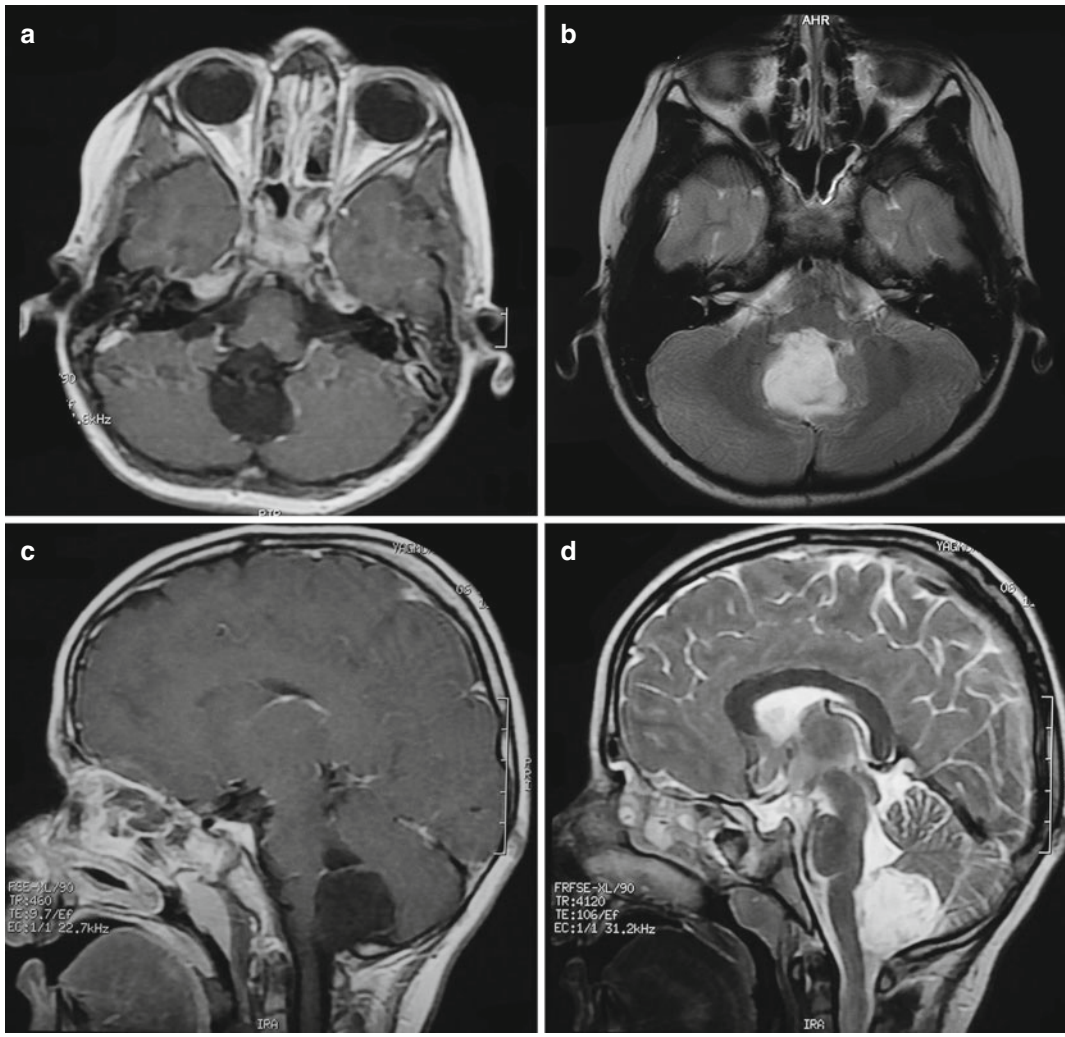

Fig. 52.6 (**a**–**d**) Axial and sagittal MRI images (T2 and T1) demonstrating a well-circumscribed posterior fossa lesion with CSF intensity and no surrounding edema but significant mass effect at midline, (**e**) dermoid tumor with hair content in the cisterna magna

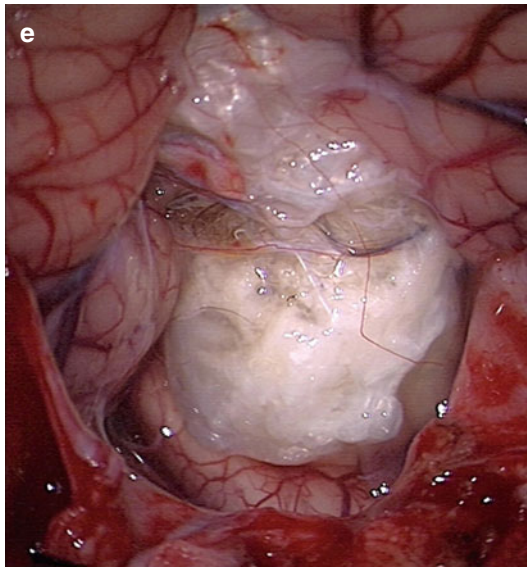

Fig. 52.6 (continued)

52.8 Preoperative Considerations

An important surgical anatomy to consider is the direction of occipital dermal sinuses which is typically directed inferiorly, and when extending intracranially, they enter below the torcula and can be adherent to dural sinus [6, 9, 15, 28]. Another well-described consideration is to avoid spillage of content within subarachnoid space. Leakage of contents may lead to meningitis as well as deposition epithelial cells within the surgical field and increase risk of recurrence [15]. To minimize the spillage of contents into the subarachnoid space, if the cyst is punctured, the contents must be aspirated promptly, and avoid irrigating the area until contents cleared. However, if spillage does occur, after aspirating the content, the operative site must be thoroughly irrigated, and corticosteroid wash has been recommended by some authors [12, 26]. On the other hand, if the cyst is large, deliberate drainage of the contents through a small opening may be necessary to allow working space and decrease manipulation of surrounding already compressed tissues [2, 12, 15].

If obstructive hydrocephalus is present, the removal of the tumor should obviate the need for the treatment of hydrocephalus as in none of the posterior fossa lesions in Caldarelli's pediatric series [8] required shunt insertion.

52.9 Technical Consideration

With the use of microscope and microsurgical techniques, it is often possible to achieve complete removal. Starting with the removal of dermal sinus, an elliptical skin incision is made around the dermal sinus opening in order to remove the tract completely (Fig. 52.1b). The extent of the craniotomy can be limited superiorly as the tract travels obliquely and inferiorly [9]. To accommodate resection of the dermoid tumor within the fourth ventricle or cisterna magna, a standard suboccipital exposure and craniotomy are necessary. Care must be taken when lifting the bone flap as the tract can be attached to the dura and venous sinuses [15]. Where the sinus is entering the dura, a small elliptical dural incision should be made to make certain the entire tract is removed. Again one should be prepared to deal with venous sinus bleeding in particular when close to the torcula. Once the tract is separated from the dura, the durotomy is extended via a vertical linear incision to gain access to intradural structures and the dermoid cyst. Maximal care must be taken to keep the capsule intact, which can be adherent to the surrounding tissues, including the brain and vascular tissue. Adhesions can be more significant if an abscess is present, which also can make the wall more friable.

Extradural location of dermoid tumors is very rare as reported by Martinez-Lage [6], and as these lesions occur close to the torcula, the high risk of significant hemorrhage must be kept in mind when performing the craniotomy.

52.10 Postoperative Considerations

During early post-op period, the main concerns are with infection, aseptic meningitis, hydrocephalus or pseudomeningocele, and CSF leak [9]. Aseptic meningitis is reported in many series, which is transient and self-limiting, with routine

administration of corticosteroids for few days postoperatively [9, 12].

Follow-up imaging in the form of MRI should be obtained within first 24 h, to look for possible residual and give a baseline for comparison for future studies should the need arise. In our unit, we prefer CT scan immediately post-op to detect any significant post-op hematoma and perform an MRI on the following day as for all tumor surgery procedures. Provided hydrocephalus is not a concern, follow-up imaging can be performed after 3 months and then at 1 year. If no evidence of tumor or recurrence is found, no further imaging is necessary.

Long-term follow-up, up to 17 years, has been reported in the literature, and several studies have reported very rare incidence of recurrence after partial removal [1, 10, 13]. Caldarelli et al. reported residual in three patients out of eight posterior fossa dermoids and one recurrence during an 8-year follow-up which occurred 2 years after initial operation and required surgery 5 years later [2]. Although incomplete removal rather than risk of significant morbidity with aggressive surgery is warranted [1], some authors recommend consideration for re-exploration if a significant residual exists [9]. Malignant changes have been reported but are extremely rare [4, 10].

In summary, dermoid tumors are rare. Clinically, recurrent meningitis or presences of dermal sinus are important clues in diagnosis of intracranial dermoid tumors. Surgical key points include attention to possible involvement of dural sinuses by the dermal sinus and avoiding spillage of the contents. Dermoids generally have less complicated postoperative period compared with epidermoids, and complete removal is curative.

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