Intracranial epidermoid cyst: Characteristics, appearance, diagnosis, treatment and prognosis

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Abstract
Intracranial epidermoid cysts are uncommon congenital inclusion cysts. Cerebellopontine angle is the most common location that harbors the intracranial epidermoid cyst; however, they are also found in other parts of the brain. These have a characteristic hallmarks radiological feature and well defined hypoattenuated mass resembling cerebro-spinal fluid on computed tomography. T1 weighted image shows the lesion as low signal, and T2 weighted image shows high signal whereas diffusion weighted image shows restricted diffusion leading to hyperintensity of the lesion. We present a unique review compiling the data from the literature that includes the history, characteristics of benign and malignant epidermoid cyst, typical and atypical appearances, very near differential diagnosis, followed by treatment and prognosis at one place.

Key words: Diagnosis, hypoattenuated mass, intracranial epidermoid cyst, treatment and prognosis.

Introduction
An intracranial epidermoid cyst is a rare congenital anomaly developed because of the dyssembryoplasia during the third to fifth week of gestation. Epidermoid tumors are also called as inclusion tumors because they arise from remnants of epithelial tissue during the neural tube closure at the third and fifth week of developmental stages of a fetus. These account for 0.2% to 1.8% of all intracranial tumors [1]. These typical epidermoid cysts usually appear with signs and symptoms, but may also appear incidentally in the early stages of life with less signs and symptoms. Commonest location is cerebellopontine angle, but can also be seen in other parts of the brain. The classical and typical neuroimaging of epidermoid cysts are hypodense lesions resembling CSF that does not enhance on post contrast, low signal intensity on T1WI, high signal on T2WI sequences and on diffusion weighted images they show up with high intensity [2, 3].

Epidermoid cysts were first discovered by an artist in a French medical school in 1807 [4]. Cruveilhier described the epidermoids as “pearly tumors” and Dandy as “most beautiful tumors of the body”. Among epidermoids, the highest incidence of housing of these lesions was at cerebellopontine angle which were 51.8% followed by lateral ventricle with 4.9%, fourth ventricle with 3.2%, third ventricle with 0.7% and other sites includes 39.4% [5]. The epidermoid cysts of the brainstem are very rare, configuring of about almost six cases of precisely intrinsic epidermoid cyst of the brain stem in the literature [6, 7]. The procedure for initial diagnosis of these tumors which benefit us with useful information mostly regarding the enhancement pattern is computed tomography [8].

The MRI stands separately and in the foremost position from other imaging modalities because of the fact that it allows us to differentiate the tumor from the CSF filled arachnoid cyst [9, 10] and cisterns containing CSF that are present adjacent to tumor [11, 12]. The high contrast enhancement of these cystic lesions is unusual and rarely reported [13]. The keratin rich content of the epidermoid cyst differentiates it from the thin walled cholesterol rich content Craniopharyngioma and mucoid colloidal filled fluid in Rathkes cleft cyst [14]. Epidermoid cyst should be considered as a differential diagnosis for tumors occurring at velum interpositum [15-17] such as pilocytic astrocytoma, atypical rhabdoid/teratoma, meningioma [18-20]. Misdiagnosis can sometimes occur clinically as the patients undergo remission of neurological symptoms and signs frequently. The hypothesis that explains the remissions of signs and symptoms are quite different; among which one explains that the decompression of cyst subsequently by spillage of cyst contents, and the other one goes this way that the slow growth rate and affinity to spread into the subarachnoid space causing no adjacent neural compression and thus allowing the flow of CSF in interstices normally [21, 22]. It has been concluded in the recently published studies that these epidermoids may present with localized, well encapsulated collections rather than being classic pearly white solid lesions that insinuates in the pathways of CSF [23]. Although being
relatively avascular tumors, these cause hemorrhages due to the neovascularisation caused by outpouring of the cystic contents and furtherly causing granulation into surrounding that lead to neovascularisation and thereby causing hemorrhage. Hemorrhage as a typical epidermoid cyst is also correlated with the presence of cholesterol crystals and granulation. It has also been stated that the growth rate of these epidermoid cysts which is known to be slow, causes the slow displacement of adjacent structures that do not lead to hydrocephalus in some of the cases [24]. The squamous cell carcinoma of central nervous system usually arises as a result of direct spread of tumor from head/neck or metastasis from the tumor located primarily in other parts of the body [25]. Intracranial squamous cell carcinoma primarily occurring in the brain is rare and if done, is usually from the intracranial epidermoid or intracranial dermoid cyst transforming into malignant from benign variety [26]. Fourth to fifth decade is the usual timing of epidermoid to occur [27]. This usual timing helps with the appearance of epidermoid mostly at the commonest locations such as cerebellopontine angles. Sequestration taking place in the third week of embryogenesis is believed to be the cause for occurrence of epidermoid cyst at intracerebral/superficial cerebral, or intraventricular, whereas if the same sequestration procedure taking place during the fifth week of embryogenesis is thought to be the reason for the occurrence of epidermoid cyst at orbit middle ear and cerebellopontine angle [28]. Spinal canal, diploe of cranium, middle cranial fossa, intraparenchymal cerebrum, brain stem and supratentorial are the rarest sites of epidermoid cyst to occur [29]. Epidermoid cyst occurring entirely intraparenchymally are rare and may be less than 1%, but still there are few case reports of frontal lobe intraparenchymal epidermoid cyst [30]. The differential diagnosis includes the dermoid cyst, cystic neoplasm like ganglioma, arachnoid cyst, neuroepithelial cyst, hydatid cyst and even brain abscess. Dermoid cysts are usually fat resembling not cerebro-spinal fluid, but epidermoid resembles cerebro-spinal fluid. Abscess often enhances with presence of surrounding edema and do not resemble cerebro-spinal fluid completely. The hallmark indicators for hydatid cyst are circular, on-enhanced, thin walled, singular, CSF-like lesion in the parietal regions of brain without perilesional edema while neuroepithelial cysts resemble CSF, which are sharp and regularly border lined.

**Typical appearance**

The classical and typical appearance of epidermoid cyst on MRI shows low signal intensity on T1WI and high signal intensity on T2WI [2, 3]. Diffusion weighted image is hyperintense and has a high value on apparent diffusion coefficient [31]. Capsular enhancement of the cyst is not often seen; the high contrast enhancement of epidermoid capsule is rare [13].

**Atypical appearance**

Epidermoid cyst undergoing hemorrhage may show different signal intensities on MRI depending on the duration of bleeding [32]. A case of epidermoid cyst with repetitive hemorrhage in the supracallosal region had atypical findings radiologically that included hyperdense multinodular appearance on CT, heterogeneous hyperintensity on T1WI and homogenous hypointensity on T2WI and was diagnosed as cavernous malformation but histological examination revealed as stratified squamous epithelium without hair follicles suggestive of epidermoid cyst, hence it comes to a conclusion that epidermoid cyst must be taken into consideration as a differential diagnosis if cavernous malformation at any location is suspicious [33]. The atypical features like hyperintensity on both T1WI and T2WI may be due to the increase in protein concentration and semi liquid content [34]. They also even showed hyperintensity on T1WI and hypointensity on T2WI due to the combo of increased viscosity and protein content [35]. The unusual muddy fluid like contents in the cystic lesions has been reported in a particular case report [23]. The other rare variety known as “white epidermoid” may appear hyperattenuated on CT scan due to the increased protein content. The cystic contents can sometimes be very clear mimicking the arachnoid cyst [36]. Lipid and cholesterol components of cyst attribute to the hypodensity on CT [37]. The saponification of these lipids within the capsule is also one of the attributions to hyperdensity of epidermoid cyst on CT [38]. It is to be noted that not all epidermoid cyst contains cholesterol [39-41]. Occasionally the appearance of epidermoid cyst on CT is hyperdense [42, 44, 45], that can be difficult for interpretation [46, 47, 48]. The hyperdensity may be due to keratinized debris calcification [38], and increased polymorphonuclear leukocytes [49]. The correlation between protein concentration of fluid and CT has been reported [50]. Spontaneous intracystic micro-bleeding or trauma.
also fits in the causative factors of hyperdensity, and
the yellow color fluid (xanthochromic fluid) collection in the cyst is also attributed by
microbleeding [37, 43, 51]. The reason for
hyperdensity of epidermoid cyst at cerebellopontine
angle may be due to increased concentration of
protein [34]. Even though the pathophysiology still
remains unclear, some of the factors have been
contributed suggesting to it. Pearly showing up with
waxy material is seen in hypodense epidermoids
considering the hyperdense epidermoids that contain
fluids of different colors and is cystic in nature. The
increased protein concentration may be due to
generative and exudative reaction to minor leaks
occurring recurrently because of the lipid material in
the capsule. The signals of MRI confide on the
configuration of keratin and cholesterol contents of
cyst relatively. Typically epidermoids appear
hypointense on T1WI due to solidity of cholesterol
[10]. Short T1 values have increased lipid content
and show non-enhancing mass on CT, whereas long
T1 values have decreased lipid content and show
water density mass on CT [52]. White epidermoid,
the another variant has the atypical appearance of
hyperdensity on CT, hyperintense on T1WI and
variable signal intensity on T2WI; the breakdown
products of hemoglobin cause the paramagnetic
effect that results in hyperintensity on both T1WI and
T2WI [2]. Neuroenteric cyst can be similar to a rare
white epidermoid cyst in which both of them have the
hyperintensity on T1WI but are difficult to
differentiate it from epidermoid cyst if present in the
midline [53, 54]. The variable signal intensities on
T2WI are fundamentally dependant on the proportion
of free water and protein content. According to
Ahmadi et al. [55], the increase in signal intensity of
intracystic fluid on T1WI is because of the protein
level of more than 100 ml or 9.0g. There is a gradual
increase and decrease in T1WI signal intensities as
the protein concentration of lysozomal solution
increases [55], whereas the signal intensity on T2WI
decreases with increasing protein concentration [56].
The other feature that differentiates the arachnoid
cyst and epidermoid cyst is that the former displaces
the adjacent structures, whereas the latter insinuates
between them. The presence of lipids and cholesterol
within the capsule is the cause for low density on CT.
The decreased apparent diffusion co-efficient is due
to the interference of oily contents of the cyst with
the Brownian motion of water molecules within it
that leads to restriction of the molecules and causes
hyperintensity on DWI which is also a differentiation
factor for arachnoid cyst that shows absence of
restriction on DWI and shows hypointense on DWI
[57]. MRI of arachnoid cyst shows a smoothly
marginated mass that is isointense to cerebro-spinal
fluid on all imaging sequences.

**Differential diagnosis**

1. **Arachnoid cyst** most commonly appears as
hypointense on both FLAIR and DWI located mostly
at middle cranial fossa, whereas epidermoid appears
as hyperintense due to the restricted Brownian
motion of the molecules within the cyst.

2. **Dermoid cyst** can be differentiated from
epidermoid cyst in terms of location as it has the
tendency to occur in the mid line 5-9 times fold than
epidermoid cyst. Dermoid cysts usually contain
dermal appendages like hair follicles, teeth, lipids. In
terms of appearance, it is hypodense on CT,
hyperintense on T1WI and iso-hyperintense on T2WI
and in terms of onset dermoid cyst appears in 2nd to
3rd decade of life whereas epidermoids appears in
later decades of life and grows slowly, but the
dermoids grow fast and rapid leading to the
occurrence of clinical features. Lining of dermoid
cyst is simple stratified squamous epithelium
supported by collagen.

3. **Colloid cyst** occurs most commonly at the
foramen of Monroe (third ventricle) which is very
helpful in the differential diagnosis, whereas the
epidermoids are very rarely seen in the third
ventricle.

4. **Rathkes cleft cyst** arises from remnants of
Rathkes pouch at 3-4 weeks of embryogenesis. It
shows low signal intensity to isointensity on T1WI,
iso to high signal intensity on T2WI, non-enhancing,
usually containing mucoid colloid fluid in the cyst.
Lining of Rathkes cleft cyst is single cuboidal or
columnar epithelium, with ciliated, goblet cells.

5. **Cranioopharyngioma** is a true neoplasm that
appears to be iso to hypointense to brain parenchyma
on T1WI, variable/mixed intensities on T2WI, vivid
enhancement, cyst is usually thin walled containing
cholesterol.

6. **Hydatid cyst** in the brain is mostly located in the
interhemispheric parenchyma especially in the
perfusion area of middle cerebral artery. These cysts
are single, large, spherical masses and can be
differentiated by the presence of their daughter cysts
and have signal intensity similar to CSF on CT and
MRI.

7. **Neurocysticercosis** unlike epidermoid cyst shows
different images finding at different stages of cyst
development, especially the cyst with a particular
“dot” like appearance in the cyst which represents the scolex, a particular feature of vesicular stage.

8. **Neuroenteric cyst** almost appears same as the epidermoid with the very near imaging features like iso to mild hypointense on T1WI and indeed definitive bright hyperintense on T2WI, but the point which is the best diagnostic evidence of neuroenteric cyst is the location in which it appears that is infront of the brainstem/pontomedullary junction with its lobulated or mass lesion.

**Signs and symptoms**

The symptoms produced by epidermoid are due to the mass effect of the tumor locally on the brain parenchyma, vascular elements, and cranial nerves present alongside. The symptom of epidermoid tumor is difficulty in hearing that account for 37.6%, trigeminal neuralgia 29.7%, facial palsy 19.4%, headache 17.9% and diplopia 6.7% [58]. Other uncommon symptoms include seizures, changes in mental status [59], glossopharyngeal neuralgia [60] and neuralgiform attacks of headache lasting for a short span of time [61]. Tumors causing trigeminal neuralgia arise mostly from the posterior fossa like epidermoid cyst, acoustic neuromas, and some even from central forces such as schwannomas, meningiomas and adenomas of the pituitary gland [62]. Among them epidermoid cyst contributes to 1.0-1.4% of cases [58, 62]. The posterior fossa epidermoid tumor presents with initial signs of bilateral gaze nystagmus that may be due to the follicular compression by enlarged CPA cyst [63, 64]. Nonetheless, these can also sometimes cause abrupt neurologic deficits, such as pituitary apoplexy if tumor present at intrasellar as reported by Hakan et al. [65] leading to clinical features such as nausea, ophthalmoplegia, acute onset of headache, vomiting, visual acuity loss, meningismus and endocrine dysfunc-

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**Table 1** The cases of remnant epidermoid cyst transforming into malignancy after the initial excision.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Site</th>
<th>Time to form SquCC</th>
<th>Treatment</th>
<th>Postoperative prognosis</th>
<th>Reported year</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>52</td>
<td>Male</td>
<td>Temporal lobe</td>
<td>6 months</td>
<td>S + R therapy</td>
<td>Good follow-up for 3 months</td>
<td>2011</td>
<td>Lakhdar et al. [85]</td>
</tr>
<tr>
<td>2</td>
<td>74</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>20 yrs</td>
<td>S + R therapy</td>
<td>Alive for 17 months</td>
<td>2010</td>
<td>Nakao et al. [79]</td>
</tr>
<tr>
<td>3</td>
<td>50</td>
<td>Male</td>
<td>Temporal lobe</td>
<td>6 yrs</td>
<td>S</td>
<td>Not described</td>
<td>2009</td>
<td>Ge et al. [86]</td>
</tr>
<tr>
<td>4</td>
<td>56</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>8 yrs</td>
<td>S + Gamma knife</td>
<td>Alive for 13 months</td>
<td>2006</td>
<td>Tamura et al. [26]</td>
</tr>
<tr>
<td>5</td>
<td>45</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>11 yrs</td>
<td>S + R therapy</td>
<td>Alive for 12 months</td>
<td>2004</td>
<td>Guan et al. [87]</td>
</tr>
<tr>
<td>6</td>
<td>54</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>3 months</td>
<td>Ch</td>
<td>Death after 13 months</td>
<td>2003</td>
<td>Hamlat et al. [88]</td>
</tr>
<tr>
<td>7</td>
<td>46</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>-</td>
<td>S</td>
<td>Not described</td>
<td>2001</td>
<td>Nawashiro et al. [82]</td>
</tr>
<tr>
<td>8</td>
<td>55</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>13 yrs</td>
<td>S</td>
<td>Death after 3 months</td>
<td>2001</td>
<td>Asahi et al. [89]</td>
</tr>
<tr>
<td>9</td>
<td>50</td>
<td>Female</td>
<td>Temporal lobe</td>
<td>10 yrs</td>
<td>S + Ch + R therapy</td>
<td>Alive for 60 months</td>
<td>1999</td>
<td>Murase et al. [90]</td>
</tr>
<tr>
<td>10</td>
<td>67</td>
<td>Female</td>
<td>Frontotemporal</td>
<td>31 yrs</td>
<td>S</td>
<td>Death after 1 month</td>
<td>1991</td>
<td>Tognetti et al. [91]</td>
</tr>
<tr>
<td>11</td>
<td>36</td>
<td>Male</td>
<td>Temporal lobe</td>
<td>2 yrs</td>
<td>S</td>
<td>Not described</td>
<td>1989</td>
<td>Abramson et al. [77]</td>
</tr>
<tr>
<td>12</td>
<td>59</td>
<td>Female</td>
<td>Intraventricular</td>
<td>33 yrs</td>
<td>S + R therapy</td>
<td>Alive for 36 months</td>
<td>1987</td>
<td>Goldman and gandy [92]</td>
</tr>
<tr>
<td>13</td>
<td>43</td>
<td>Male</td>
<td>Temporal Lobe</td>
<td>7 yrs</td>
<td>S</td>
<td>Death after 1 month</td>
<td>1965</td>
<td>Fox and south [93]</td>
</tr>
<tr>
<td>14</td>
<td>54</td>
<td>Male</td>
<td>Base of brain</td>
<td>1 yr</td>
<td>S</td>
<td>Death after 6 weeks</td>
<td>1965</td>
<td>Toglia et al. [94]</td>
</tr>
</tbody>
</table>

S = surgery, R = radiation, Ch = chemotherapy. All patients have one operative history except case no. 13 and 14 who have 2 and 3 operative history, respectively.

Epidermoid cyst transforming into malignancy after the initial excision. Epidermoid cyst rupture can sometimes lead to chemical meningitis, persistent headaches. It has also been stated that the growth rate of these epidermoid cysts is known to be slow, hence causing the slower displacement of adjacent structures which do not lead to abrupt hydrocephalus [24]. Mutism has also been reported in the literature as a clinical presentation of epidermoid cyst in the quadrigeminal cistern by Kawal et al. [66]. Epidermoid cyst manifesting in the pineal region presents with diverse syndromes such as Sylvian aqueduct syndrome causing paralysis of downward gaze with palsy of horizontal gaze; Parinaud’s syndrome resulting due to compression of superior colliculi causing convergence, paralysis of upward gaze, pupillary dissociation with near light; Colliers syndrome which results due to compression of dorsal brainstem leading to ptosis and retraction of eyelid [67]. Sometimes the vascular compression at the cerebellopontine angle by the tumor can result in hemifacial spasm [68]. Hyperprolactinemia was reported in a case of intrasellar epidermoid by Hakan et al. [65], the reason for hyperprolactinemia was thus examined as the pituitary stalk compression resulted in the disruption of transport of prolactin inhibiting factor [69]. 15% of all intracranial epidermoid tumors manifest in quadrigeminal cistern [70, 72, 73]. Median location of epidermoid tumor occurs with separation of neuroectoderm from the cutaneous counterpart. Epidermoid cyst at other intracranial locations has the unusual findings of hydrocephalus [73, 74]. Epidermoids in the pineal region also contrarily appear with raised intracranial pressure and hydrocephalus very early rather than the usual; this may be due to the presence and obstruction of cerebral aqueduct Sylvius and the posterior part of third ventricle adjacent [70, 71, 75]. Epidermoid cysts have been reported to be
present at velum interpositum due to the epithelial tissue displacement within the velum interpositum [76]. It can be here argued that they can even arise in quadrigeminal cistern present posteriorly, and migrate to cavaum velum interpositum, in such cases Desai et al. reported a series of epidermoid tumors in quadrigeminal cisterns that even though being large in size the cyst has displaced the third ventricle, but did not enter the velum interpositum or the ventricles [70]. MRI here is useful in portraying the precise anatomy of the velum interpositum region by clearly demonstrating the displaced neurovascular structures [20, 71]. Displacement of the internal cerebral veins ventrally, corpus callosum and posterior peri callosal arteries dorsally assure the presence of a mass in the velum interpositum [71], whereas the third ventricular tumors displaces the internal cerebral veins dorsally [16].

Malignant changes

The transformation of the benign condition of cyst into malignant is not common, and if occurs is more aggressive. The precise mechanism of benign epidermoid cyst transforming to malignancy is still yet unclear, but causes like subtotal excision of the cystic wall, and recurrent cystic rupture leading to severe inflammatory response may lead to malignant transformation [77-79] or the inflammation due to foreign body reaction or with carcinoma in situ. Symptoms and signs that have the sudden onset and develop speedily is the significant indication clinically, of epidermoid cyst transforming into malignancy [80], the typical imaging pattern includes the features such as growth of the focally enhanced mass rapidly, on MRI and CT imaging [81]. The appearance of malignant part of epidermoid cyst on DWI is hypointense [82], whereas the classical epidermoid cyst on DWI appears as hyperintense that can be useful in diagnosing and follow up post operatively [83]. Immunohistochemistry of the malignant epidermoid cyst reported in the literature showed the positivity of tumor cells with P53 protein. The abnormal or the rampant growth illustrates malignant transformation rather than the linear growth [84]. The cases of remnant epidermoid cyst after initial excision transforming into malignancy are remarkably rare and are reported here till the recent (Table 1).

In the above mentioned cases (Table 2), all the patients at least had one operative history before the recurrence or malignant transformation, which can be one of the causes of transformation into malignancy. Among all the above cases, case no: 1, 2, 4, 5, 9, 12, who have been treated with radiotherapy after the excision of malignant epidermoid cyst have shown better prognosis, which states that postoperative radiotherapy leads to a better prognosis of patients and is highly recommended. There are even some cases (Table 2) that showed malignant epidermoid tumor without any previous operative history.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Site</th>
<th>Treatment</th>
<th>Postoperative prognosis</th>
<th>Reported year</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>52</td>
<td>Male</td>
<td>Cerebellopontine angle</td>
<td>-</td>
<td>Autopsy</td>
<td>1912</td>
<td>Ernst</td>
</tr>
<tr>
<td>2</td>
<td>46</td>
<td>Male</td>
<td>Frontal lobe</td>
<td>S</td>
<td>Alive</td>
<td>1960</td>
<td>Davidson and small [95]</td>
</tr>
<tr>
<td>3</td>
<td>73</td>
<td>Male</td>
<td>Frontopolar</td>
<td>-</td>
<td>Autopsy</td>
<td>1960</td>
<td>Landers and Danielski [96]</td>
</tr>
<tr>
<td>4</td>
<td>53</td>
<td>Male</td>
<td>4th ventricle</td>
<td>S + R therapy</td>
<td>Death after 2 month</td>
<td>1981</td>
<td>Dubois et al. [97]</td>
</tr>
<tr>
<td>5</td>
<td>53</td>
<td>Female</td>
<td>Parasellar</td>
<td>S</td>
<td>Death after several weeks</td>
<td>1983</td>
<td>Lewis et al. [98]</td>
</tr>
<tr>
<td>6</td>
<td>62</td>
<td>Male</td>
<td>Parasellar</td>
<td>S</td>
<td>Death after 1 week</td>
<td>1993</td>
<td>Acciari et al. [99]</td>
</tr>
<tr>
<td>7</td>
<td>58</td>
<td>Male</td>
<td>Cerebellopontine angle</td>
<td>S + R therapy</td>
<td>Alive 2.5 yrs</td>
<td>1995</td>
<td>Nishio et al. [100]</td>
</tr>
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<td>8</td>
<td>45</td>
<td>Male</td>
<td>Preoptic</td>
<td>S + R therapy</td>
<td>Death after 12 months</td>
<td>2005</td>
<td>Michael et al. [101]</td>
</tr>
<tr>
<td>9</td>
<td>65</td>
<td>Female</td>
<td>Pineal region</td>
<td>S</td>
<td>Vegetative state</td>
<td>2007</td>
<td>Pagni et al. [102]</td>
</tr>
<tr>
<td>10</td>
<td>67</td>
<td>Male</td>
<td>Cerebellopontine angle</td>
<td>R therapy</td>
<td>Death after 11 months</td>
<td>2007</td>
<td>Kodum et al. [103]</td>
</tr>
<tr>
<td>11</td>
<td>45</td>
<td>Male</td>
<td>Posterior fossa</td>
<td>S</td>
<td>No follow up</td>
<td>2007</td>
<td>Agarwal et al. [104]</td>
</tr>
<tr>
<td>12</td>
<td>72</td>
<td>Female</td>
<td>Cerebellopontine angle</td>
<td>S + R therapy</td>
<td>Alive 12 months</td>
<td>2008</td>
<td>Kim et al. [105]</td>
</tr>
<tr>
<td>13</td>
<td>63</td>
<td>Female</td>
<td>Pre pontine</td>
<td>S</td>
<td>Death after 36 days</td>
<td>2008</td>
<td>Shuyu et al. [106]</td>
</tr>
</tbody>
</table>

S = surgery. R = radiation. All patients have no operative history and time taken to form SqCC was 6 years only for case no. 13.

The treatment is generally carrying very good prognosis for epidermoid cyst is total surgical resection grossly [79]. Whereas the decompression of cyst can just be a temporary relief [107-109], while subtotal resection can result in recurrence of tumor which is estimated to appear at the time of an interval equal to age of patient with the addition of nine months to the initial presentation of symptom [110]. A routinely suggested approach for suprasellar epidermoid cyst is transcranial, peculiarly if the epidermoid is adhered to the adjacent vessels and nerves. The adjuvant therapy coupled up with subtotal/total resection depending upon the
adherence is promising management strategy if the malignant transformation has occurred. The benefits of the radiosurgery have been well documented for the control of malignant epidermoid tumors [90, 111]. Postoperatively stereotactic radiation therapy has been effective, that showed the control of local tumor growth for two and half years, and with disease free survival rate for more than five to eight years in some reported cases [78, 112]. In case of adjuvant therapy, gamma knife radiosurgery has been reported as a useful treatment [26]. The epidermoid tumors, in which the complete removal of them is difficult, have to be followed up for the recognition of their malignant transformation. The immediate surgical procedure should be carried out for epidermoid cyst located in the sellar region as in the delayed cases may cause a sudden neurological deficit due to the proximity of abundant neurovascular structures.

Surgical complications

The main post-operative complications are dysfunction of cranial nerves and aseptic meningitis [113, 114]. The cautious imaging study preoperatively especially regarding to the region of the internal cerebral veins (velum interpositum); will forbid the damage to these structures intraoperatively while undergoing for masses in these areas. Aseptic meningitis stands between 2-50% incidences occurring postoperatively according to literature data; patients mostly whose cystic capsule has been excised incompletely are prone to these complications [115, 116]. Chemical meningitis results in the rupture of the cyst. Removal of epidermoid cyst leads to complications such as hydrocephalus, which may be obstructive or communicating [117]. The surgical resection of epidermoid in the posterior fossa may sometimes lead to hypertrophic olivary degeneration. The clinical picture of this includes palatal myoclonus, which is an involuntary movement of oropharynx, uvula and soft palate, ocular myoclonus and dentatorubral tremor [118].

Prognosis

As the growth rate of these epidermoid tumors resembles the same as the human skin any cell retained can lead to risk of recurrence or regeneration. According to the recently published studies, subtotal excision of posterior fossa epidermoid cyst has the recurrence rate between 0-54% [120-122]. Children after initial resection of epidermoid cysts in the pineal region should have a close follow up, as a potential to develop a metachronous neoplasm (germinoma) is possible [123]. Whereas even the subtotal removal of the fourth ventricular epidermoid cyst carries a good prognosis [124], even if the tumor is subtotally resected long term survival can be expected in atypical epidermoid cystic patients. Even if the tumor is excised subtotally in neurologically preserved patients the prognosis remains quite good which is confirmed by the experience [125, 126]. The location of the tumor and the time taken from onset to diagnosis are the important factors for the prognosis than the size of tumor.

Conclusions

It is also considered necessary to revive that these epidermoids may be provincial, well enveloped with collections on behalf of the classical solid pearly white tumor insinusating between the adjacent structures as described usually. One of the major tools for diagnosing and post-operative follow-up is diffusion weighted Mirth subtotal or partial resection of the tumor can cause the regeneration of the tumor from the remnants leading to recurrence or malignancy as the leftover cells of the cystic wall/tumor are nothing but the epidermal cells that has the regenerating capacity as such of epidermis of skin. The adjuvant treatment with radiotherapy after surgical resection is important and has good results.

References


