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# Malignant transformation of a cerebral dermoid cyst into a squamous cell carcinoma with malignant intraperitoneal spreading along a ventriculoperitoneal shunt: illustrative case

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**BACKGROUND** Malignant progression of intracranial dermoid cysts into squamous cell carcinoma is extremely rare with only three reports published so far. Intracranial dermoid cysts are uncommon benign tumors lined by stratified squamous epithelium of embryonic ectodermal origin.

**OBSERVATIONS** Here, the authors present the case of a 64-year-old female with a recurrent temporal dermoid cyst. After surgery for the recurrent dermoid cyst, once in the early 1990s and another 16 years later, the patient presented with headache and nausea due to hydrocephalus. After implantation of a ventriculoperitoneal shunt, she deteriorated rapidly and died only 60 days after admission. Autopsy revealed malignant transformation of the epithelial lining of the dermoid cyst into a squamous cell carcinoma resulting in neoplastic meningiosis and intraperitoneal tumor spread along a previously implanted ventriculoperitoneal shunt.

**LESSONS** Malignant transformation should be considered in patients with dermoid cyst who show new leptomeningeal contrast enhancement. In the case of hydrocephalus, alternatives to peritoneal shunting should be considered.

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**KEYWORDS** dermoid cyst; malignant transformation; intracranial tumor; squamous cell carcinoma; peritoneal spreading; ventriculoperitoneal shunt

Dermoid cysts (DCs) comprise 0.5%–1% of intracranial tumors.[1] Histologically, they contain keratin, cellular debris and cholesterol and arise from primitive pluripotent ectodermal cells trapped within the neural tube. Usual sites of occurrence are the midline, Sylvian fissure and the perisellar area.[2] Common presentations include headache, seizures and focal neurological deficits. Rupture of a cyst with dissemination of its contents causes an aseptic meningitis and is a rare but devastating event.[3] Following cyst rupture, dispersion of its fatty contents into the subarachnoid space causing obstructive hydrocephalus is possible.[4] Moreover, vasospasm may occur in patients with involvement of the meninges or subarachnoid space.[5] Radical resection of the tumor especially around critical neurovascular structures is not always feasible. Transformation into squamous cell carcinoma (SCC) is an extremely uncommon event that has been much more often been reported in epidermoid cysts.[6–11] Only few cases of malignant transformation are known in DCs.[12–15] Here, we present a patient with malignant transformation of a temporal DC into a SCC with meningeosis carcinomatosa, abdominal spreading via a ventriculoperitoneal (VP) shunt and a rapid clinical deterioration with fatal outcome.

## Illustrative Case

A 64-year-old female was admitted to our institution due to headache and nausea. She had undergone previous resections, one in the early 1990s and another 16 years later, of a right temporomesial DC, with only subtotal removal of the DC due to its firm adhesions in both surgeries. The computed tomography (CT) scan on admission revealed a hydrocephalus (Fig. 1A) treated by the insertion of a VP

**ABBREVIATIONS** CSF = cerebrospinal fluid; CT = computed tomography; DC = dermoid cyst; DWI = diffusion weighted imaging; ETV = endoscopic third ventriculostomy; ICU = intensive care unit; MRI = magnetic resonance imaging; SCC = squamous cell carcinoma; VP = ventriculoperitoneal.

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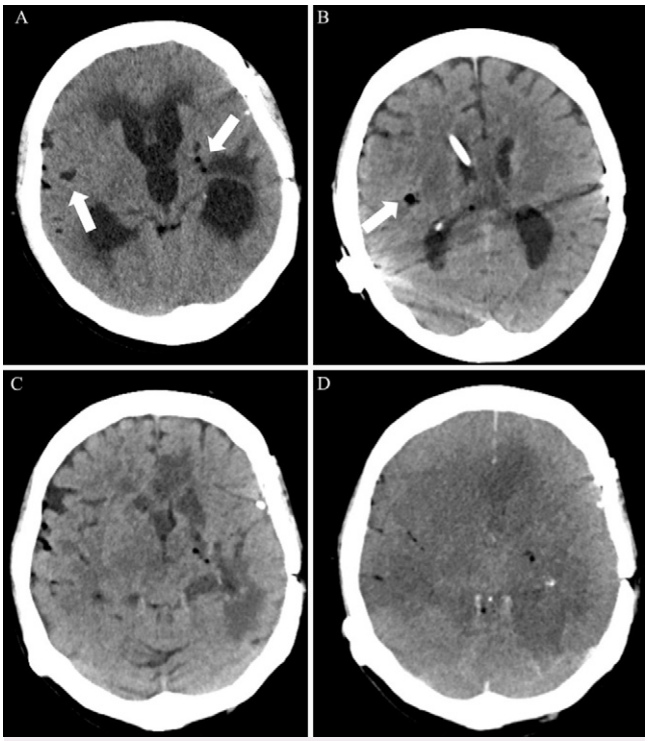


**FIG. 1. A:** An axial CT scan on admission showed hydrocephalus and lipid-equivalent hypodense lesions (*white arrows*, −100 HU). **B:** Emergency axial CT scan obtained 7 days after admission displays decrease of hydrocephalus after insertion of a VP shunt and lipid-equivalent hypodense lesions (*white arrow*). Multiple infarctions are shown in the axial CT scan 10 days after admission (**C**). Global brain edema on the axial CT scan 17 days after admission (**D**).

shunt. The thorough investigation of the cerebrospinal fluid (CSF) showed no evidence of tumor cells or signs of aseptic or bacterial meningitis. Seven days after surgery, the patient presented with a facial palsy and dysphagia. Only 6 hours after the onset of symptoms, the patient deteriorated into coma and had to be intubated and

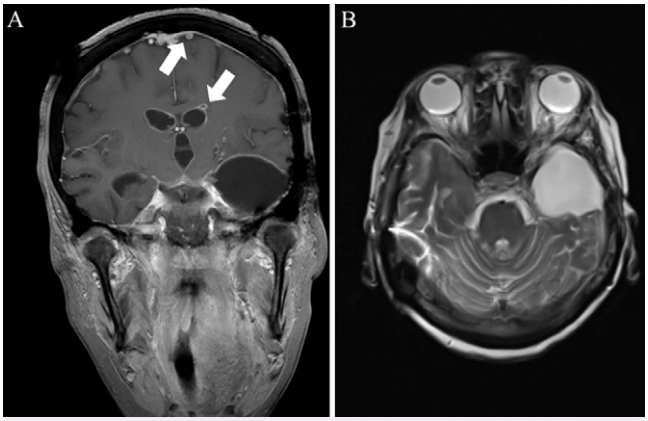


**FIG. 2.** Hyperintense T1 intensity of the DC on coronal contrast-enhanced MRI (**A**) and iso- to slightly hypointense T2 intensity of the DC n axial MRI 51 days after admission (**B**). Leptomeningeal enhancement is indicated by *white arrows*.

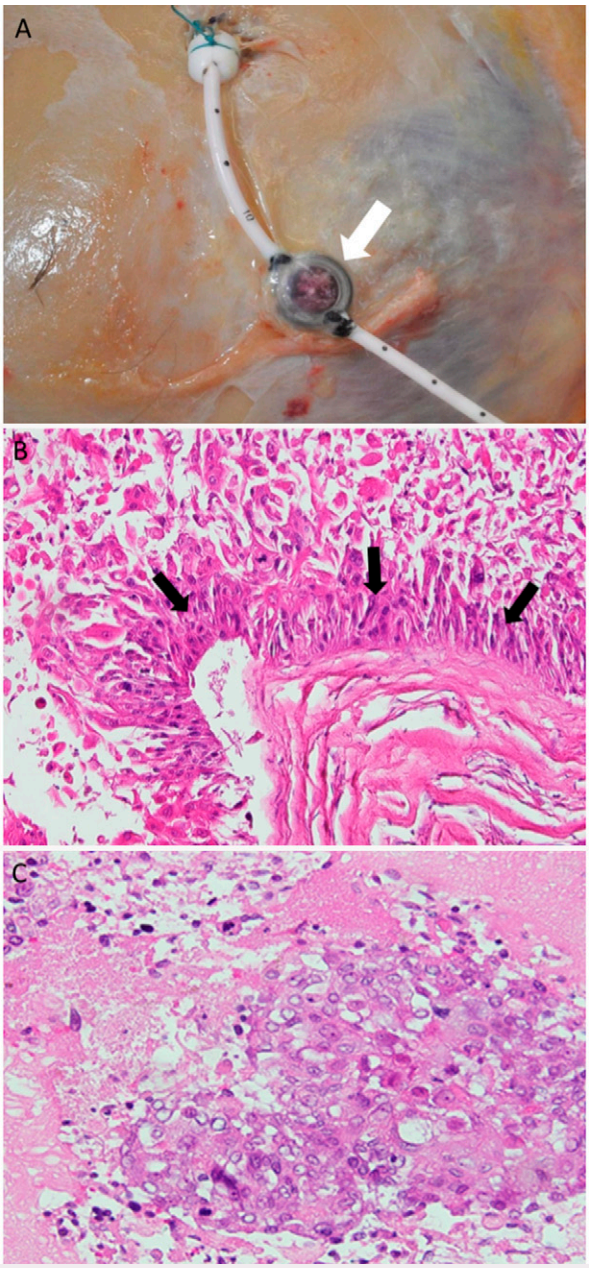


**FIG. 3.** Postmortem inline shunt reservoir (*white arrow*, **A**) showing tumor cell invasion in the chamber proximal to the valve. Light microscopy image of the cells of a squamous cell carcinoma (*black arrows*, **B**) within the shunt reservoir. Malignant infiltration of the temporo-basal dura and trigeminal ganglion (**C**). Hematoxylin and eosin, original magnification ×400 (**B and C**).

mechanically ventilated. The emergency axial nonenhanced CT scan displayed lipid-equivalent hypodense lesions in the right temporomesial region with residual marginal calcifications compatible with a dissemination of the DC, which retrospectively was already obvious on the CT scan at admission (Fig. 1A and B). Additionally, the decrease in ventricular size indicated a proper function of the VP shunt (Fig. 1B). Cranial magnetic resonance imaging (MRI) revealed dural and ependymal enhancement with an increased diffusion-weighted

**TABLE 1. Summary of case reports of malignant transformation**

| Authors & Year | Age (yrs) | Sex | Transformation (yrs) | Survival (days) | Location |
|---|---|---|---|---|---|
| Gluszcz, 1962[12] | 52 | M | 0 | 5 | Frontal lobe |
| Kobayashi et al., 1993[13] | 59 | F | n/a | 180 | Frontal lobe |
| Tsugu et al., 2001[15] | 47 | M | 0 | Alive | Cerebellum |
| Nickl et al., 2021 | 64 | F | 11 | 60 | Temporal lobe |

n/a = not available; survival = overall survival from admission to hospital to death in days; transformation = time in years from first diagnosis of DC until malignant transformation to SCC.

imaging (DWI) signal in the basal ganglia on both sides and the right cerebellar vermis indicating ischemia. On cranial MRI the DC showed a hyperintense T1 signal intensity without contrast enhancement, an iso- to slightly hypointense T2 signal and a negative DWI restriction (Fig. 2). Fatty droplets were located in the subarachnoid space and ventricles with chemical shift artifacts in the DWI. Furthermore, profuse leptomeningeal enhancement as a correlate of carcinomatous meningitis, progressive hydrocephalus and transependymal diapedesis of cerebrospinal fluid were encountered. Suspecting chemical meningitis as a complication of the DC, steroid medication was commenced. Follow-up CT scan after 3 days revealed multifocal infarctions (Fig. 1C). Unfortunately, the comatose patient developed fixed and dilated pupils after 10 days on the intensive care unit (ICU), and follow-up CT scan revealed global brain edema, which led to cerebral circulatory arrest (Fig. 1D). The patient died 60 days after initial admission for the hydrocephalus. At autopsy, cellular tumor invasion within the abdomen and the VP shunt (Fig. 3A and B) was found. Histopathological examination of the temporal tumor showed an infiltration of the trigeminal ganglion and perineural invasion by atypical squamous epithelium with pleomorphic cellularity and increased mitotic activity (Fig. 3C). Immunoreactivity for cytokeratin 5/6, CD56, p16, and Ki-67 was strong. Small spreadings of tumor cells were found in the CSF and in the Douglas pouch. Considering the clinical history and after exclusion of another primary SCC by postmortem examination, the final diagnosis was malignant transformation of a benign DC into a SCC with tumor spreading along the VP shunt.

## Discussion

### Observations

Dermoid tumors are usually benign developmental tumors that arise from aberrant ectodermal embryonic tissue in the neural groove at 4 to 5 weeks after gestation. DC usually consist of squamous cell epithelium, hair follicles, and keratine-like material. Malignant transformation of a DC is extremely rare and only four cases of intracranial transformation to SCC have been reported so far including the present case (Table 1). In this case, the squamous cell component of the DC has transformed into SCC as previously reported by Tsugu et al.[15] Typical radiological features of DC include high-intensity signal on T1-weighted images due to its fatty content and a variable signal on T2-weighted images. On DWI, DC typically appears hyperintense due to a decrease of free water diffusion with no associated contrast enhancement.[2,16] In our case, the initial DC presented with the above-mentioned MRI features. When malignant transformation occurs, edema, invasion of adjacent structures and contrast enhancement indicates cellular proliferation.[7,15] However, the mechanisms of malignant transformation of a DC remain unclear. Chronic inflammatory processes due to cyst rupture after subtotal resection are discussed as a cause.[10,17] The prognosis of malignant transformation of a DC or epidermoid cyst into an SCC is poor with a mortality of three of the four reported patients within 180 days (Table 1).[12] Surgical resection following radiotherapy is considered a treatment option.[15]

Peritoneal spreading of central nervous tumors is a rare event with reports previously for solitary brain tumors and meningiomatosis.[18,19] Several risk factors for shunt-related extraneural metastases of primary central nervous tumors have been reported: type of tumor including glioblastoma and medulloblastoma, the presence of leptomeningeal metastases and atrial shunt placement.[20] Although the prevalence of shunt related metastases is still unknown especially in the case of obstructive hydrocephalus, the use of an endoscopic third ventriculostomy (ETV) is an alternative treatment option.[21] To the best of our knowledge, this is the first case of an extracerebral spread of a malignant DC, which occurred through a previously implanted VP shunt due to hydrocephalus after presumed cyst rupture.

### Lessons

We emphasize to take malignant transformation as a differential diagnosis into account, if leptomeningeal contrast-enhancement is present in case of an intracranial DC. Whenever possible, alternative treatment options for tumor-associated hydrocephalus like ETV should be taken into consideration in such a case.

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## Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

## Author Contributions

Conception and design: RC Nickl, Monoranu, Ernestus, Löhr. Acquisition of data: RC Nickl, Monoranu, Löhr. Analysis and interpretation of data: RC Nickl, Monoranu, Löhr. Drafting the article: RC Nickl, V Nickl, Schindehütte, Löhr. Critically revising the article: all authors. Reviewed submitted version of manuscript: RC Nickl, Monoranu, Löhr. Approved the final version of the manuscript on behalf of all authors: RC Nickl. Statistical analysis: RC Nickl. Administrative/ technical/material support: RC Nickl. Study supervision: RC Nickl, Ernestus.

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