

Case report

## Sylvian fissure dermoid cyst with intratumoral hemorrhage: case report

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### Abstract

It is rare for a dermoid cyst to develop intratumoral hemorrhage. A 61-year-old woman had a sudden-onset left hemiparesis and slow response to verbal requests for one week when unenhanced computed tomography scanning revealed a mixed iso- and hypo-dense heterogeneous lesion in the right fronto-temporal area. T1-weighted magnetic resonance imaging (MRI) of the brain showed a mixed hyper- and hypo-intense tumor in the right fronto-temporal area. The tumor became hyperintense on T2-weighted MRI and was faintly enhanced at tumor periphery on T1-weighted MRI. The tumor was excised and pathological examination revealed a dermoid cyst with intratumoral hemorrhage. The post-operative course was complicated by hemorrhage in the tumor bed, which was evacuated immediately. The patient improved and could walk without support two weeks after the second operation. After 1 year of follow-up, she was well and without neurological deficits. To the best of our knowledge after a literature review, only two previous cases of dermoid cyst have featured intratumoral hemorrhage.

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### 1. Introduction

The intracranial dermoid cyst is a rare congenital tumor with a frequency of less than 1% of all intracranial neoplasms [1–9]. Dermoid cysts may be intact or ruptured on presentation [2,5,10–15]. Unruptured dermoid cysts usually present with focal mass effect, hydrocephalus, or elevated intracranial pressure [2,5,10–15]. By contrast, ruptured dermoid cysts may cause seizures, aseptic meningitis, acute or chronic obstructive hydrocephalus, or cerebral ischemia [2,5,10–15]. However, it is rare for a dermoid cyst to have intratumoral hemorrhage; only two such cases have been reported in the literature [1,16]. This report presents a patient with left sylvian fissure dermoid cyst harboring intratumoral hemorrhage.

### 2. Case report

A 61-year-old woman had a sudden-onset left limb weakness, poor memory, and slow response to verbal requests for one week prior to admission. There was no history of trauma. On admission, she had mildly impaired orientation to person and place. No sensory deficits were noted. Her right limbs had no motor deficits. However, she had left central type facial palsy and the muscle power of her left limbs was grade 4/5. Deep tendon reflexes were increased on the left side and the left-sided plantar response was extension. The complete blood count, serum biochemistry, bleeding time, prothrombin time and partial thromboplastin time were all within normal limits. Unenhanced computed tomography (CT) scanning revealed a mixed iso- and hypo-dense heterogeneous lesion in the right fronto-temporal area, with mass effect and a midline shift to the left (Fig. 1A). Calcification was noted in the outer part of the lesion (Fig. 1A). T1-weighted magnetic resonance imaging (MRI) showed a mixed hyperin-

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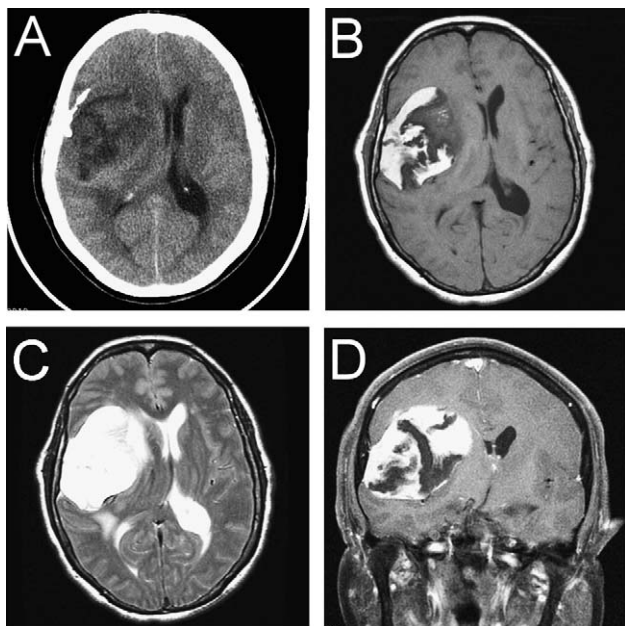


Fig. 1. CT and MRI visualization of the intracranial dermoid cyst. (A) Axial section of unenhanced CT scan showing mixed iso- and hypo-dense heterogeneous lesion in the right fronto-temporal area. Note mass effect and midline shift to the left. Calcification was noted in the outer part of the lesion. (B) Axial section of T1-weighted MRI showing a mixed hyper- and hypo-intense tumor in the right fronto-temporal area. The tumor became hyperintense on T2-weighted MRI (C, axial section) and was faintly enhanced at the periphery of the tumor on gadolinium-enhanced T1-weighted MRI (D, coronal section).

tense and hypointense tumor in the right fronto-temporal area (Fig. 1B). The tumor became hyperintense on T2-weighted MRI (Fig. 1C) and was faintly enhanced at the periphery on gadolinium-enhanced T1-weighted MRI (Fig. 1D). Craniotomy was done and a well-encapsulated tumor, about 7 cm × 5 cm × 4 cm in size, was found in the right sylvian fissure. The outer part of the tumor capsule had areas of calcification. The tumor tightly adhered to the dura, and dark-reddish liquefied hematoma gushed out upon opening of the dura. In addition to the hematoma, the cyst contained hair and a pearly white, oily substance (Fig. 2A). The tumor was totally excised. Pathological examination revealed a dermoid cyst with intratumoral hemorrhage (Fig. 2). The capsule of the dermoid cyst was lined with granular squamous epithelium (Fig. 2B). The cyst contained keratinous material, hairy substance and sebaceous glands (Fig. 2B and C). Old hemorrhage, sheets or nests of hemosiderin-containing histiocytes and chronic inflammatory cell infiltrate were also noted (Fig. 2D). The patient came through the operation well and regained consciousness soon after surgery. However, her level of consciousness deteriorated roughly 12 h after surgery and follow-up CT revealed hemorrhage in the tumor bed. An emergency evacuation of the hematoma was done. After the second operation, she regained consciousness and her motor weakness improved. She could walk without support two weeks after the second procedure. After 1

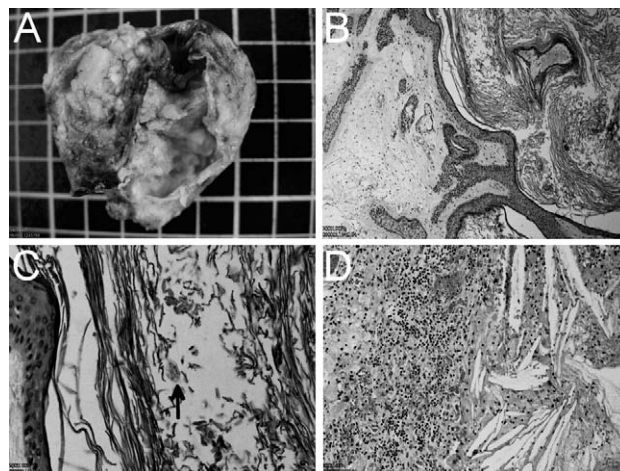


Fig. 2. Gross pathology and histopathology of the dermoid cyst. (A) Photograph of the gross pathology of the dermoid cyst. (B–D) Histopathology of the dermoid cyst. The capsule of the dermoid cyst was lined with granular squamous epithelium (B). The cyst contained keratinous material, hairy substance, and sebaceous glands (B and C). Old hemorrhage, sheets or nests of hemosiderin-containing histiocytes, and chronic inflammatory cell infiltrate were noted (D). (Hematoxylin and Eosin, (B) 40×; (C) 200×; (D) 100×).

year of follow-up, she was well and without neurological deficits.

### 3. Discussion

The intracranial dermoid cyst is a rare congenital tumor that is often located in the posterior fossa and the base of the brain; however, it may occur anywhere within the cranial vault [6]. The symptoms of patients with intracranial dermoid cysts depend upon the location of the tumor, its depth and size, and whether it has ruptured or not; no specific symptomatology predominates [2,5,6,8,10,17]. Usually intact dermoid cysts grow insidiously by spreading in the subarachnoid space, enveloping critical neurovascular structures long before they become clinically evident [14]. Common clinical presentations for patients with intact intracranial dermoid cysts include headache, visual deficits, and seizures [6,8,17]; ruptured dermoid cysts may present with meningitis, acute hemiparesis, cheiro-oral syndrome, arachnoiditis, ventriculitis, hydrocephalus, and cerebral ischemia [2,5,10–15]. Our patient's tumor manifested as sudden-onset motor weakness, and she was initially thought to have cerebrovascular disease, before she eventually was found to have an intracranial dermoid cyst with intratumoral hemorrhage.

The clinical diagnosis of intracranial dermoid cyst is difficult and depends on imaging studies such as CT or MRI [6,8,17]. Most dermoid cysts are hypodense on CT scan [5,10,18], however, there may appear hyperdensity, which mimics intratumoral hemorrhage, although they actually have no bleeding [19]. The dermoid cysts are usually hyperintense on both T1- and T2-weighted MRI because of their fat contents; however, not all dermoid cysts display homogeneous

hyperintensity on MRI because the fat can be absent or other components such as hair or other debris are predominant [5,10,18]. Our patient had hemorrhage within intracranial dermoid cysts, which is considered rare; we identified only two such cases in the literature [1,16]. One patient was an 80-year-old man who presented with acute confusion and right hemiparesis and was found to have a left frontal dermoid cyst with intratumoral subacute and old hemorrhage [16]. The other patient was a 63-year-old woman who presented with sensory impairment over the territory of the left trigeminal nerve and was found to have a parasellar dermoid cyst with old hemorrhage [1]. The CT and MRI features of the dermoid cysts with bleeding were related to the amount and the age of the hematomas, as well the predominant contents of the dermoid cysts [1,16]; thus, it may be difficult to diagnose the intratumoral hemorrhage and assess the age of the hematomas by imaging findings [1]. In our patient, the CT scan revealed a mixed iso- and hypo-dense heterogeneous lesion, which did not indicate whether there was intratumoral hemorrhage or not. However, the medial part of the tumor showed hypointensity on T1-weighted MRI and hyperintensity on T2-weighted MRI, which suggested there was subacute or old hemorrhage. Although these three patients have subacute or old hemorrhage, however, their clinical manifestations might or might not be related to the bleeding episode. For the patient with trigeminal nerve deficit [1], the symptoms were considered not related to the bleeding episode; by contrast, the acute onset of symptoms in another previously reported patient [16] might be related to the intratumoral hemorrhage. In our patient, the one-week time window between the onset of symptoms and surgery might explain the histological findings and inflammatory reaction of the dermoid cyst with hemorrhage. Thus, her clinical manifestations might be related to the bleeding episode, unlikely related to expansion of tumor size during resolution of hematoma or due to tumor growth because dermoid cysts are slow-growing tumors [2,5,6,8,10,17].

The mechanisms of bleeding in the dermoid cyst are unclear. Several factors, such as hypervascularity of tumor, tumor associated with neoplastic aneurysm, and trauma, have been considered as general contributors to intratumoral bleeding [1,20]. However, these factors were unlikely to contribute to the intratumoral bleeding seen in our patient because dermoid cysts are generally poorly vascularized [21]; in addition, pathological examination of the dermoid cyst in our patient did not reveal any aneurysm or vessel wall degeneration, and the patient had no history of trauma or bleeding tendency. On the other hand, although dermoid cysts are hypovascular tumors [21], the tendency of these tumors to occur near or adherent to large venous structures such as the cavernous sinus and the torcular herophili suggests that angiogenesis may still happen at some portion of the dermoid cysts [5] and contribute to later development of intratumoral hemorrhage. The dermoid cyst in our patient was also located in an area rich in venous structures, the sylvian fissure, however, there was no evidence of any significant tumor angiogenesis

in our patient to support such speculation. Another probable contributing factor is tumor vessel degeneration [1,20]. The two previously reported patients (80 and 63 years old, respectively) [1,16] and our patient (61 years old) were relatively older than most dermoid cyst patients because dermoid cysts are diagnosed more frequently in children than in adults [1,4]. The advanced age of clinical presentation suggested that the regressive alterations of vessel walls such as hyalinization and degeneration in the tumors may play a role in the pathogenesis of the intratumoral hemorrhage.

Complete surgical resection is the treatment of choice for dermoid cysts [5,10,15,22]. However, most of these lesions are incompletely excised and recurrence is common due to their widespread nature and firm adherence to vasculature, meninges, and normal cerebral tissue [5,6,8,15,22]. Although the dermoid cyst in our patient seemed to be totally excised, it was tight adherent to the dura. Therefore, any small amount of residual epithelial lining of the cyst wall might continue the process of keratinization and desquamation. Frequent follow-up of this patient to monitor for possible tumor recurrence is mandatory.

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