Case Report

A Rare Case of Cranial Multicentric Dermoid Cyst: Clinical and MRI Evaluation of Spontaneous Rupture

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Summary

In a 33-year-old male patient, magnetic resonance imaging (MRI) demonstrated cranial multicentric epidermoid cyst and rupture of the cysts. Dermoid cysts are extremely rare inclusion cysts constituting less than 0.5 % of all primary intracranial tumors. Most of them are asymptomatic. Multicentric type of dermoid cysts have been reported very rarely in the literature. Most of these cysts are asymptomatic. Hormonal changes with age can increase the glandular secretions resulting in the rupture of rapidly enlarging cysts. Magnetic resonance imaging displays the typical findings of rupture. There has been only one case of multicentric dermoid cyst reported in the literature until now.

Key words: Dermoid cyst, multicentric dermoid cyst, rupture, MRI

INTRODUCTION

Dermoid cysts are extremely rare inclusion cysts constituting less than 0.5 % of all primary intracranial tumors¹². They originate from the intracranial ectodermal cells during closure of the neural tube between the third and fifth weeks of embryogenesis, and are benign slow-growing tumors¹³,²⁰. Most of them are asymptomatic⁷. Hormonal changes with age can increase the glandular secretions resulting in rupture of these rapidly enlarging cysts²¹. Rupture is rare and generally spontaneous. Rarely, cases can demonstrate acute symptoms following rupture or infection⁷. Reports on multicentric dermoid cysts are extremely rare in the literature. The rupture of the cysts yields typical findings on MRI. Due to the rarity of the condition, a case of multicentric dermoid cyst that had spontaneously ruptured has been presented in this article.

CASE PRESENTATION

A 33-year-old male patient presenting with tremor in his right hand presented to our
Outpatient Clinic. In his history, there was no trauma or intracranial surgery. On his neurological examination, there were no findings other than the resting tremor in the right hand. On MRI studies, hyperintense foci (Fig. 1A) in the left Sylvian fissure on T1-weighted sequences, and heterogeneous hyperintense foci (Fig. 1B) on T2-weighted sequence, and hyperintense foci in the subarachnoid spaces indicating the dispersion of liquid cholesterol into the subarachnoid space (Fig. 1C) on T1-weighted sequences demonstrated two dermoid cysts (Fig. 2). In Fig. 2, beside the hyperintense lesion of the two dermoid cysts, there were also hyperintense foci of liquid cholesterol in the background. The patient was treated symptomatically, and his complaints decreased by time.

**Figure 1A:** Axial T1-weighted MRI image (without gadolinium): In the Sylvian fissure a lesion of mixed-signal-intensity with wall rupture (arrow) is displayed to be extending towards the ICA. Furthermore, hyperintense foci (arrow head) of liquid cholesterol droplets in basal cisternae that are released through rupture are seen. 1B. Axial T1-weighted MRI image: In the Sylvian fissure, a heterogeneous hyperintensity (arrow) in the lesion wall caused by fat and skin appendages in the lesion content is observed extending towards the ICA. 1C. Axial T1-weighted MRI image (without gadolinium): Multiple hyperintense presentation of liquid cholesterol droplets in the subarachnoid space in both cerebral hemispheres showing rupture (arrow head).

**Figure 2:** Coronal FLAIR MRI: In left Sylvian fissure, as result of multiple ruptures, two uncompressed hyperintense dermoid cysts (arrow) and hyperintense liquid cholesterol droplets (arrow head) dispersed in the subarachnoid space in both cerebral hemispheres can be observed.
DISCUSSION

Intracranial dermoid tumors are benign cystic tumors of epidermal and dermal origin that are rarely encountered. These tumors are most frequently located in the suprasellar and parasellar cisternae and rarely in the spinal canal and the Syrinx cavity\(^1,13\). Dermoid cysts are lined with squamous epithelium and contain skin appendages such as hair follicles, sweat glands, nails and teeth\(^9\). Dermoid cysts grow slowly and show a tendency to attach to adjacent tissues\(^10\). With the increase in contained glandular secretion and epithelial desquamation, dermoid cysts enlarge and compress the neural structures that can lead to symptoms. The enlargement of the tumor mass rarely results in spontaneous rupture\(^9\). However, head traumas and iatrogenic surgical complications have been reported as causes of rupture\(^14\). The clinical findings depend on the site of the lesion and compression of the adjacent parenchyma, often presenting with headache, seizures and neurological deficits; when ruptured, aseptic meningitis or ventriculitis can occur upon passing of the cyst content into the subarachnoid space\(^1,18\). Moreover, cerebral vasospasm and infarcts following rupture of dermoid cysts have been reported in the literature\(^9,21\). By way of the tract, the dermoid cysts may have a relationship with the dermoid sinuses on the skin. Based on this evidence, at those cases; there can be seen skin symptoms without neurological deficits\(^8\).

On MRI studies, dermoid cysts are visualized as hyperintense on T1-weighted images without iv gadolinium due to their liquid cholesterol content, and are visualized as heterogeneously hyperintense on T2-weighted images due to various components in their content\(^12,18\). In the case of rupture, hyperintense fat globules in the subarachnoid space and the ventricular system, as well as fat-fluid levels on T1-weighted images are diagnostic\(^16,19\). If the fat content of the lesion is relatively low, the lesion will reveal a cerebrospinal fluid–like signal intensity. In such cases, fluid attenuation inversion recovery (FLAIR) is useful, in that the fat will appear hyperintense (bright) on a background of suppressed fluid signal (dark). On MRI, the fat constituents create a chemical shift artifact due to misregistration of the signal in the frequency-encoded direction. This can be particularly useful in the preoperative diagnosis of these lesions\(^18\). Following rupture, sequences with hyperdense foci with fat-like signals are observed on T1-weighted\(^12,18,19\). This image is the best diagnostic clue\(^12\). Dermoid cysts do not display signal enhancement following iv injection of gadolinium, but extensive pial and ependymal enhancement can be seen from chemical meningitis caused by ruptured cysts\(^5,18\). Formerly, rupture of the cysts was accepted as very fatal, but with the wide the use of MRI, many asymptomatic cases of ruptured cysts have been increasingly diagnosed\(^1\). In their study on 7 patients comparing the results of MRI and CT, Smith et al.\(^17\) stated that MRI was not superior to CT scanning in the diagnosis of ruptured cysts, but that preoperative MRI was superior in displaying the situation and spread of the cyst in relation to the adjacent tissues\(^17\).

Although of similar origin, multiple intracranial epidermoid tumors have been frequently reported in the literature, whereas only one case of multicentric dermoid cysts has been reported thus far\(^4,6,11\). Goyal et al.\(^4\) made mention of surgery in their 40-year-old male patient with epidermoid cyst in the right cerebellopontine angle. Our case had two dermoid cysts in the left Sylvian fissure and no history of surgery or trauma. Since our case had been diagnosed after rupture, we could not determine whether the second focus diagnosed was a result of intrathecal spread or not\(^11\). However, this could be
caused by two cell foci in the period between the third and the fifth weeks of embryogenesis(15).

Malignant transformation of dermoid cyst is rare(2). In the differential diagnosis, epidermoid, craniopharyngioma, lipoma and teratoma should be considered. The differences are that epidermoids show limited diffusion, the solid components of teratoma and craniopharyngioma are hyperintense and enhance strongly, and lipomas demonstrate homogenous fat attenuation and/or signal intensity(3).

CONCLUSION
Dermoid cysts can be multicentric. Dermoid cysts and ruptured dermoid cysts are now more frequently diagnosed with the wide use of MRI. Rupture can occur either spontaneously or during head trauma or in the operative-postoperative period. Ruptured dermoid cysts can be symptomatic most frequently causing headache or other serious symptoms. Epidermoid, craniopharyngioma, lipoma and teratoma should be considered in the differential diagnosis. Magnetic resonance imaging provides an almost definite diagnosis.

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