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RUPTURED INTRACRANIAL DERMOID- A CASE REPORT

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ABSTRACT

Intracranial dermoids rarely rupture. They are associated with significant risk of morbidity and mortality when they rupture. The rupture within the ventricle is further rarer. Case report: We present a case of intraventricular rupture of a dermoid cystic tumor along with dissemination of subarachnoid fat. The diagnosis was made on CT scan and MRI imaging. The patient died in few days from chemical meningitis. Intracranial dermoid cystic tumors account for <1% of all intracranial masses. Dermoids are nonneoplastic, congenital ectodermal inclusion cysts that contain varying amounts of ectoderm derivatives to include apocrine, sweat, and sebaceous cysts as well as hair follicles, squamous epithelium, and possibly teeth. They are separate from an epidermoid cyst or Teratomas. Teratomas are true neoplasms that contain tissue from all three embryonic germ cell layers (1).

Key words: Intracranial dermoid, Intraventricular rupture, Chemical meningitis.

INTRODUCTION

Dermoid cystic tumors arise from the inclusion of ectodermally committed cells at the time of neural tube closure during the third to fifth week of embryogenesis. These lesions are slow growing due to the active production of hair and oils from the internal dermal elements (2). The presentation of dermoid tumors is quite variable. Occasionally they are incidental findings discovered on imaging otherwise unrelated clinical complaints, or they are discovered during imaging investigation of unexplained headaches, seizures, and rarely olfactory delusions (3, 4). When dermoid cystic tumors rupture and spread their contents into the ventricles and subarachnoid and/or subdural spaces, the most common clinical presentation is that of headache and seizures.

We here describe rare intracranial rupture of dermoid within the ventricle and subarachnoid space.

CASE REPORT

A twenty five old female presented with constant headaches since one month. She complained of severe left trigeminal nerve-type pain. The patient had no associated complaints of nausea, vomiting, altered mental status, or seizures in last month. On presentation, vital signs were normal. Laboratory evaluation was normal. No focal neurologic deficits were identified. CT scan of the brain without contrast administration demonstrated a 7 cm diameter fat-containing mass at left tempero-parietal region. The lesion showed clumps of fat density at its margins. (Figure 1 and 2). Scattered fat-containing droplets were disseminated throughout the subarachnoid space. Small fat density was appreciated within the occipital horn of left lateral ventricle.

The patient developed fever, chills and seizures in next 24 hours. A diagnosis of chemical meningitis was made secondary to ruptured dermoid. The patient died the next day.

DISCUSSION

Intracranial dermoid cysts are extremely rare, constituting less than 0.5% of primary intracranial tumors. They tend to occur in the midline sellar, parasellar, or frontonasal regions (10,11). Other dermoid cysts are midline in the posterior fossa, where they occur either as vermian lesions or within the fourth ventricle (9,10,11). These cysts increase in size by means of glandular secretion and epithelial desquamation. Growth can lead to rupture of the cyst contents, causing chemical meningitis that may lead to vasospasm, infarction, and even death (12). Malignant transformation into squamous cell carcinoma has also been described (11). Symptoms typically are ascribed to mass effect created on adjacent intracranial structures. If rupture occurs, aseptic chemical meningitis may ensue with profound irritative effects from the disseminated cholesterol debris. Chemical meningitis is a relatively rare development and is reported in approximately 7% of cases of dermoid tumor rupture (1, 5). Chemical meningitis may elicit transient cerebral ischemia secondary to vasospasm with complicating infarction that may result in the death of the patient. Morbidity may also be related to chemical arachnoiditis (7).

Dermoid cystic tumor rupture usually occurs spontaneously; however, cases of rupture secondary to closed head trauma or iatrogenic surgical complications have been reported (5). Supratentorial dermoids often present in the second or third decades of life, while posterior fossa dermoids typically present in the first decade of life as a consequence of mass effect exerted on the fourth ventricle with resulting hydrocephalus (3).

On CT scan intracranial dermoid tumors are fat containing lesions. Density values greater than fat may be encountered depending on the nature of an individual tumor's contents. Calcification is appreciated typically at the wall of the dermoid. The wall may show enhancement on contrast administration. On MRI scans, dermoids will be hyperintense on T1-weighted imaging and heterogenous on T2-weighted images. When a dermoid tumor ruptures, fat droplets may be seen scattered within the ventricular system and/or subarachnoid space. Chemical meningitis would show intense pial and ventricular ependymal enhancement after the administration of contrast.

A differential diagnosis, such as epidermoids, teratomas, lipomas, craniopharyngiomas, and occasionally arachnoid cysts needs to be considered in case of fat containing lesion within the cranium. CT or MRI helps in getting the right diagnosis and differentiating dermoid from other lesions.

Surgery aims on complete microsurgical resection of the mass and wall. Great care is made to avoid spilling the contents in if the dermoid is not ruptured. Recurrence is rare but is more common if there are retained portions of the tumor wall. Rare reports describe the development of squamous cell carcinoma in retained remnants of a dermoid cystic tumor wall (3, 8).

CONCLUSION

Ruptured intracranial Dermoid is a rare disease to occur. It can be easily diagnosed on a CT scan examination. Intraventricular rupture of the Dermoid is further rarer. This case is reported because of its rarity. Because of the concern of rupture of dermoid and associated mortality the surgical treatment is a prudent course for this condition.

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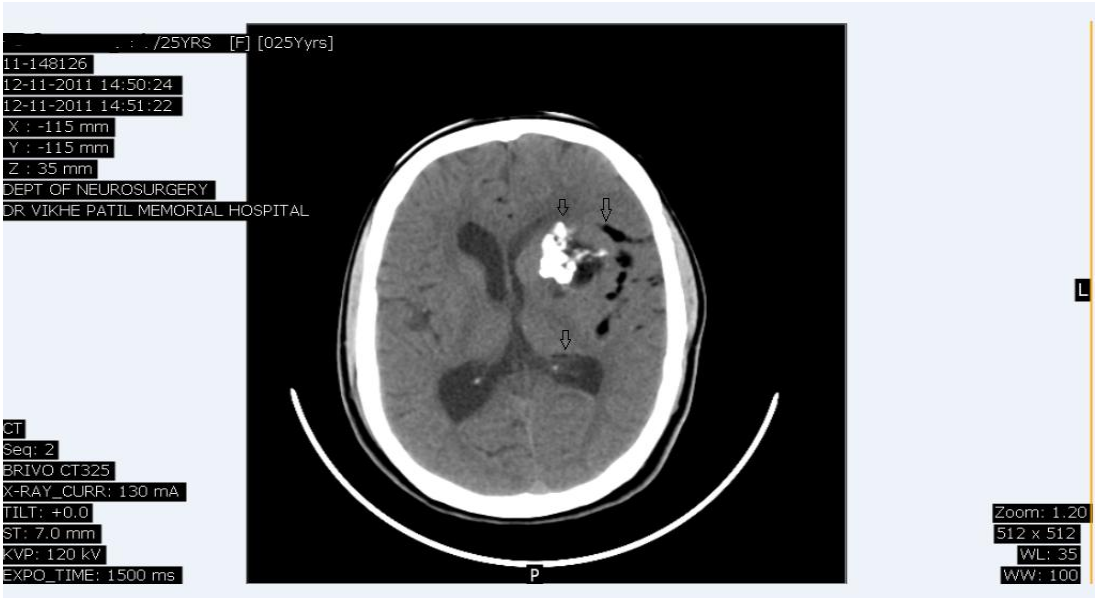

Figure 1: Unenhanced CT scan of brain shows fat density droplets in subarachnoid space and also within the left lateral ventricle. Lobulated fat component is noted within the lesion

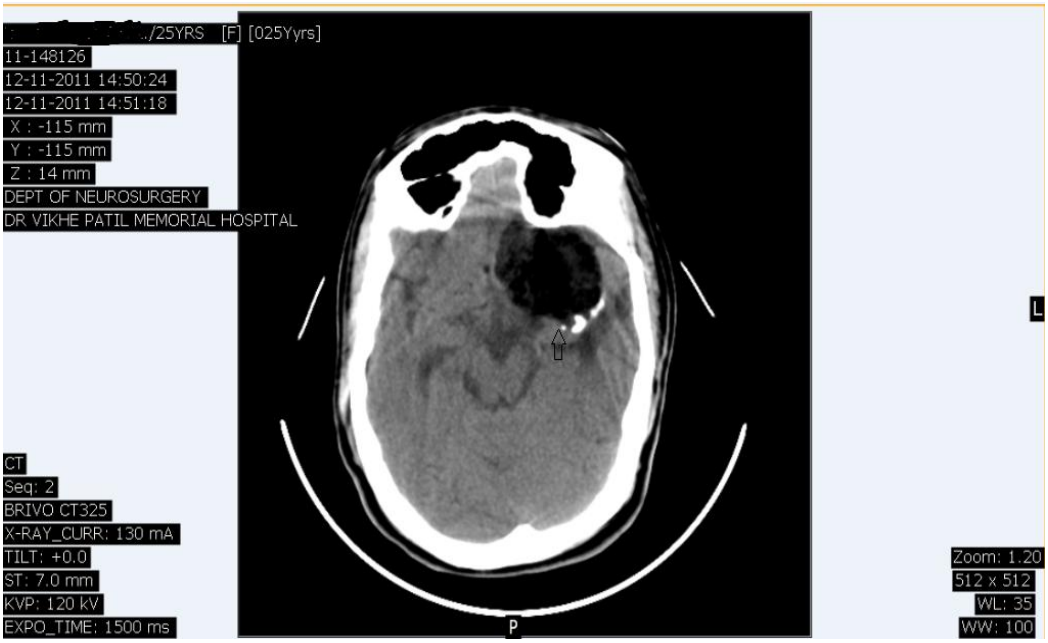

Figure 2: Unenhanced CT scan of brain shows fat density lesion at left temperoparietal region. Partial calcification of the wall of the lesion is seen

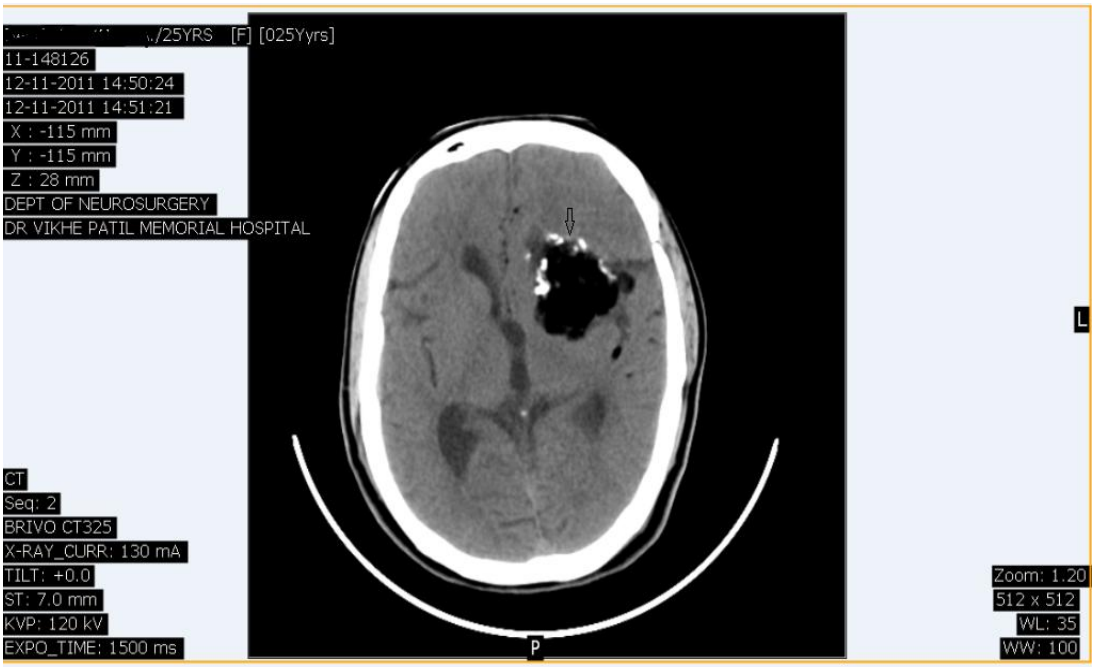

Figure 3: Unenhanced CT scan of brain shows fat density lesion at left temperoparietal region. Partial calcification of the wall of the lesion is seen.