CASE REPORT

Giant supratentorial dermoid cyst in an infant: a case report

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ABSTRACT

Background: Dermoid cysts are rare, benign cystic lesions. The giant supratentorial variant is an exceptional presentation, both in terms of location and size.
Case description: We report the case of a 6-month-old female infant from northeastern Peru who presented with seizures, progressive cranial enlargement since 4 months of age, and a preliminary diagnosis of a cerebral cyst based on computed tomography (CT). She was referred to a national pediatric referral center in Lima, where contrast-enhanced brain CT revealed a large cystic lesion in the right hemisphere with posterior wall enhancement after contrast administration, thinning of the surrounding brain parenchyma, and signs of hydrocephalus. Laboratory tests showed leukocytosis, and pathological examination confirmed a dermoid cyst with a marked inflammatory component, numerous foamy macrophages, entrapped hair follicles, calcifications, and osseous tissue within the fibrous cyst wall.
Conclusion: This case of a dermoid cyst in an infant represents an extremely rare presentation, due to its supratentorial location, the size of the lesion, and the patient's young age. Imaging studies play a key role in diagnosis, surgical planning, and follow-up of this condition.

Keywords: Dermoid Cyst; Brain Cysts; Computed Tomography; Case Reports (Source: MeSH)

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Quiste dermoide supratentorial gigante en una lactante: reporte de caso

RESUMEN

Antecedentes: Los quistes dermoides son lesiones quísticas benignas y poco frecuentes. La variante supratentorial gigante constituye una presentación excepcional, tanto por su localización como por su tamaño.
Descripción del caso: Se describe el caso de una lactante de sexo femenino de 6 meses de edad, procedente del nororiente del Perú, que presentó convulsiones, incremento progresivo del tamaño craneal desde los 4 meses y un diagnóstico preliminar de quiste cerebral por tomografía. Fue referida a un centro pediátrico de referencia nacional en Lima, donde la tomografía cerebral contrastada evidenció una extensa lesión quística en el hemisferio derecho con realce de la pared posterior a la administración del contraste, adelgazamiento del parénquima cerebral y signos de hidrocefalia. Los análisis de laboratorio evidenciaron leucocitosis y los estudios de anatomía patológica confirmaron un quiste dermoide con abundante componente inflamatorio, presencia de macrófagos espumosos, folículos pilosos atrapados, calcificaciones y tejido óseo en la pared fibrosa del quiste.
Conclusión: Este caso de quiste dermoide en una lactante representa una presentación extremadamente rara, tanto por la localización supratentorial y el tamaño de la lesión, como por la corta edad de la paciente. Los estudios de imágenes desempeñan un papel clave en el diagnóstico, la planificación quirúrgica y el seguimiento de la lesión.

Palabras clave: Quiste Dermoide; Encéfalo; Quistes; Tomografía Computarizada; Informe de Caso (Fuente: DeCS)

INTRODUCTION

Intracranial dermoid cysts are rare, benign cystic lesions that contain variable amounts of ectodermal and mesodermal tissue remnants (1).

They occur most frequently during the first three decades of life and are more common in women (2,3). Their intracranial location is most often reported in the midline of the suprasellar, parasellar, and frontal regions, as well as in the posterior fossa.

The clinical presentation depends on their location: they are usually asymptomatic when small, but as they enlarge, they may cause symptoms such as headache, seizures, or aseptic meningitis due to their expansive nature. In some reports, cyst rupture has been described (3,4), as well as unusual presentations such as intracranial hemorrhage (5).

We present the case of a female infant referred from a province in Peru to a national pediatric referral center in Lima, with a presumptive diagnosis of a giant cerebral cyst.

CASE DESCRIPTION

A 6-month-old female infant from the province of Bagua, Amazonas, was born by eutocic delivery, with no apparent alterations in neonatal anthropometry. At birth, her head circumference measured 35 cm. According to the mother, progressive head enlargement was noted beginning at 4 months of age; however, she sought medical attention only after the patient experienced generalized tonic-clonic seizures. On physical examination, there was marked widening of the fontanelles and a head circumference of 46 cm (98th percentile). A non-contrast brain CT revealed an extensive extra-axial cystic lesion in the right frontotemporal region and left ventricular dilation. Due to the complexity of the case, the patient was referred to a national pediatric referral center in Lima.

In the emergency department, the patient appeared awake, active, responsive to stimuli, and without evident neurological deficit. Initial hematologic studies revealed a hemoglobin level of 11.8 g/dL, a hematocrit of 36.2%, a total leukocyte count of 14,770/µL, a platelet count of 447,000/µL, and percentages of neutrophils, band cells, eosinophils, and lymphocytes of 23%, 0%, 7%, and 9.6%, respectively. Biochemical tests revealed glucose levels of 141 mg/dL, urea levels of 3.9 mg/dL, and creatinine levels of 0.2 mg/dL. Immunologic tests for HIV, hepatitis B, hepatitis C, and VDRL were negative, as was the urinalysis. During her stay in the emergency unit, she experienced another generalized tonic-clonic seizure, prompting a contrast-enhanced brain CT (Figure 1). The CT scan showed a large cystic lesion occupying almost the entire right cerebral hemisphere, measuring approximately 75 mm × 117 mm × 72 mm (craniocaudal × anteroposterior × transverse). The lesion had a thin wall with post-contrast enhancement, homogeneous fluid-density content, and two additional adjacent cystic lesions of the exact nature located in the posterior parietal region, measuring 11 mm × 10 mm and 8 mm × 7 mm, respectively (figura 2). The central lesion caused marked thinning of the right cerebral parenchyma, collapse of the ipsilateral lateral ventricle, approximately 5 mm midline shift, significant dilatation of the left lateral ventricle, and mild transependymal edema.

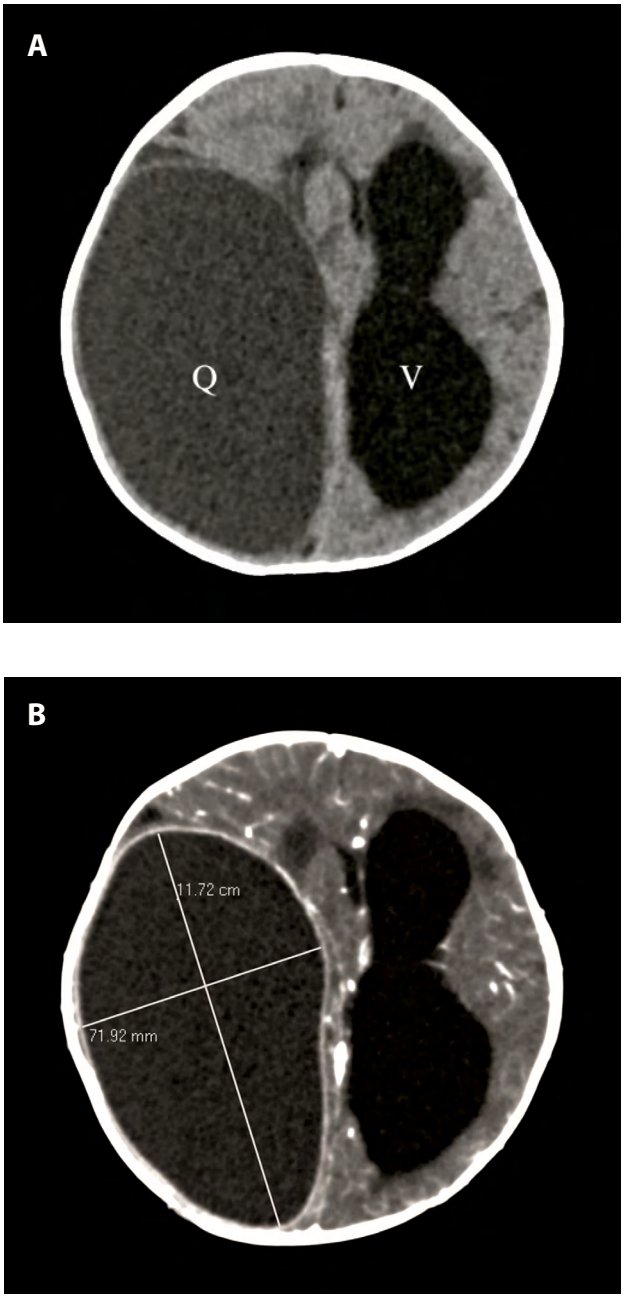

Figure 1. Axial brain CT showing an extensive cystic lesion (Q) in the right cerebral hemisphere. Panel A shows collapse of the right ventricle (V), dilation of the left ventricle, and midline shift. Panel B shows the cyst dimensions and posterior wall enhancement after contrast administration (CTDI: 60 mGy).

Given the emergent situation and to prevent clinical deterioration, a decompressive craniectomy was performed. Intraoperatively, an extensive cystic lesion with a significant mass effect on the cerebral parenchyma was confirmed and completely resected. Subsequent histopathological examination (Figure 3) revealed a cystic lesion with a wall composed of fibroconnective tissue, some entrapped hair shafts, calcifications, and loose areas with abundant inflammatory infiltrates consisting of foamy macrophages.

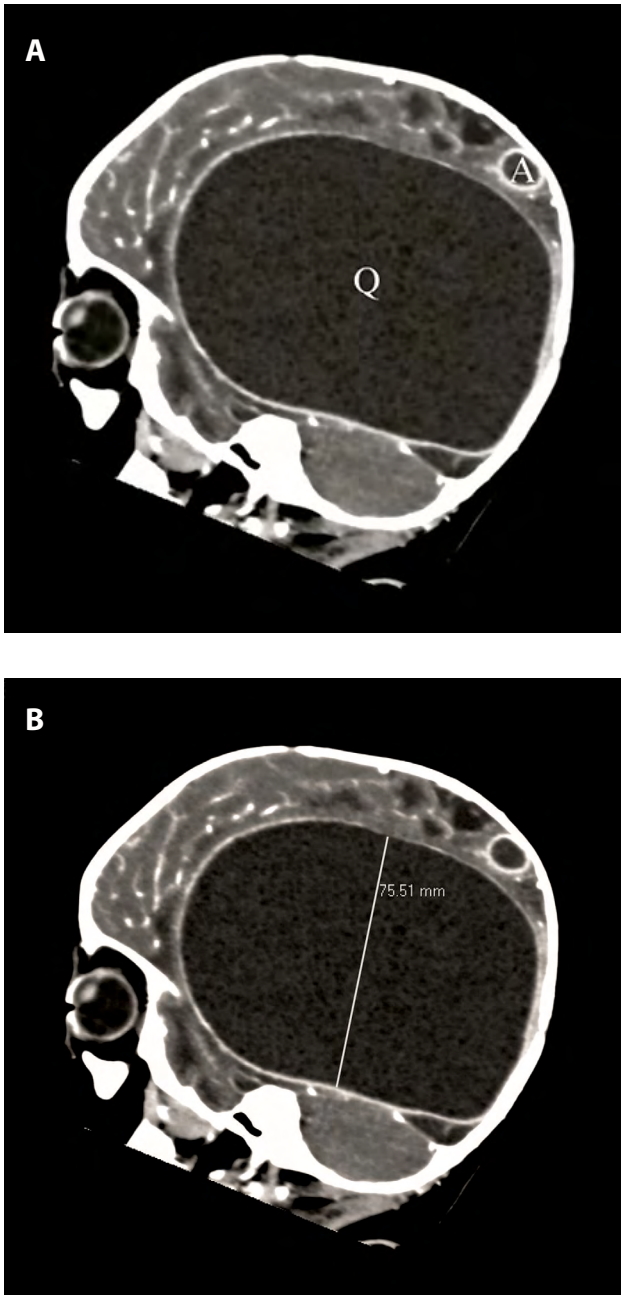

Figure 2. Sagittal brain CT showing an extensive cystic lesion (Q) and a smaller adjacent cystic lesion (A) in the posterior parietal region. Panel A shows the cystic lesions, and panel B shows the craniocaudal measurement of the primary cyst (75.51 mm).

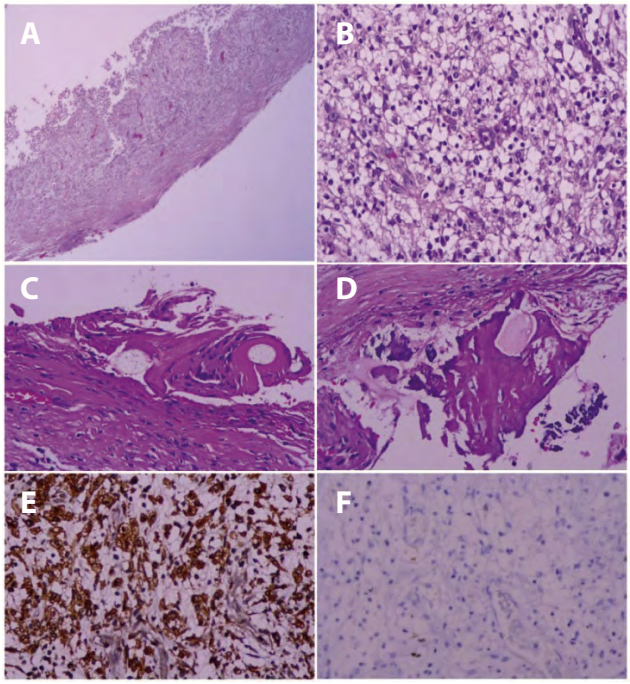

Figure 3. Histological sections of the dermoid cyst. Panel A shows the cyst wall composed of vascularized fibrous connective tissue, lacking lining epithelium, and with an abundant inflammatory component. Panel B shows an inflammatory infiltrate consisting of numerous foamy macrophages, lymphocytes, and some neutrophils and eosinophils. Panel C shows the fibrous cyst wall with trapped follicle and hair shafts. Panel D shows calcifications and bone tissue in the fibrous cyst wall. Panel E shows positive immunostaining for CD-68 in the cytoplasm of macrophages. Panel F shows negative immunostaining for S-100 in macrophages/histiocytes.

The adjacent brain tissue showed no alterations, and no infectious agents were identified. Immunohistochemistry demonstrated the presence of CD45+ lymphocytes and CD68+ macrophages. Ck (cocktail), EMA, and GFAP were negative. The Ki-67 proliferative index was 2–5%. Synaptophysin, S-100 protein, and Olig-2 were also negative.

During the postoperative period, one week after emergency surgery, an external ventricular drain was placed. Five days later, the patient developed a respiratory complication due to pneumonia caused by *Klebsiella spp.* The infection was treated with broad-spectrum antibiotics (vancomycin and meropenem) for 16 days. After completing treatment, she was discharged and continued outpatient follow-up without neurological complications. One month later, she was readmitted for ventriculoperitoneal shunt placement. Follow-up imaging demonstrated progressive reduction of hydrocephalus, along with right hemispheric cerebral atrophy as a sequela of the dermoid cyst's location.

DISCUSSION

Only a few cases of giant intracranial dermoid cysts have been reported in the international literature (13–15), each described in different locations. The present report constitutes an exceptional case, documenting a giant dermoid cyst in a 6-month-old infant, associated with seizure episodes, significant head growth, and a possible abscess as a complication.

Dermoid and epidermoid cysts are considered congenital inclusion cysts; however, epidermoid cysts are 4 to 9 times more common than dermoid cysts (1). Both entities originate from defective closure of the neural tube, which occurs between the third and fifth weeks of gestation (6). The main difference lies in the fact that dermoid cysts contain cutaneous appendages, such as hair follicles and sebaceous and sweat glands, which appear as hyperintense signal areas on T1-weighted MRI images.

The usual location of dermoid cysts is in the midline, particularly in the sellar, parasellar, and posterior fossa regions. In our case, the cyst was located in the right cerebral hemisphere, a site more typically associated with epidermoid cysts. Nevertheless, dermoid cysts in atypical locations have also been reported (7–9). It has been postulated that this non-midline intraparenchymal location results from lateral displacement of primitive ectodermal and mesodermal cells induced by developing cerebral vasculature through the Virchow–Robin spaces (10,11). In 1991, Lunardi and Missori proposed that intradural intracranial dermoid cysts tend to be located near large venous structures: the cavernous sinuses for supratentorial dermoid cysts, and the torcular Herophili for infratentorial dermoid cysts (11). In the present case, the cyst likely originated from ectodermal and mesodermal cells displaced along the venous channels of the right cavernous sinus, given its proximity to this structure in the inferior segment, with subsequent cephalic growth.

The typical content of a dermoid cyst is mainly composed of fat molecules, which appear on CT as areas of low attenuation (–20 to –140 HU) and as hyperintense signals on T1-weighted MRI sequences (12). In our case, the cyst content showed hypodensity with an average fluid density of 17 HU, along with wall enhancement after contrast administration, suggesting an increase in high-density inflammatory molecules. These findings correlate with the leukocytosis observed in laboratory tests and the abundant inflammatory response seen in the histological study of the cyst wall, characterized by foamy macrophages, supporting the hypothesis of a possible abscess as a complication. Other complications reported in the literature and observed in this case include compression of vascular structures and adjacent brain parenchyma, hydrocephalus, and intracranial hypertension (10). Differential diagnoses such as arachnoid, pineal, or neoplastic cysts were ruled out based on the cyst's location and the absence of other associated congenital malformations.

The current treatment of intracranial dermoid cysts primarily depends on the location of the cyst. Since they are often located in the midline, surgical access can be complex; however, complete resection of the capsule and its contents is usually relatively straightforward, as these lesions do not exhibit infiltrative projections. In the present case, total excision of the lesion was achieved through careful separation from the adjacent brain tissue.

The main limitations of this case include the inability to generalize the findings, given that it represents a single report; the limited availability of variables for evaluation, as the data were obtained from a retrospective review of the clinical record; the inability to perform MRI due to the critical clinical condition requiring emergency decompressive craniectomy, which prevented assessing the utility of this imaging technique for diagnosis, surgical planning, and prognostic follow-up; and the short follow-up period, which limited the evaluation of late complications and the medium- to long-term effectiveness of surgical treatment.

This case report of a giant supratentorial dermoid cyst represents an infrequent presentation, due to its location, the lesion's size, and the patient's young age. The neurological symptoms can be explained by the cyst's expansive process and the possibility of rupture. Imaging studies, including CT and MRI, are fundamental for diagnosis, surgical planning, and prognostic follow-up.

This case report was prepared with the express authorization of the patient's parents, obtained through signed informed consent, for the publication of clinical and laboratory information. Ethical approval was also obtained from the Institutional Research Ethics Committee of the national referral center, in accordance with the ethical principles of the Declaration of Helsinki.

Author contributions

Boris Borja-Zapata: Conceptualization, Investigation, Methodology, Project administration, Supervision, Validation, Visualization, and Writing – review & editing.

Ramón Huamán-Olarte: Investigation, Validation, Visualization, and Writing – review & editing.

Carlos Ugas-Charcape: Investigation, Supervision, and Methodology.

Carla Cruzado-Villanueva: Resources.

Conflicts of interest

The authors declare no relevant financial or non-financial conflicts of interest.

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Ethical aspects

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