CASE REPORT

Leptomeningeal Cyst of the Orbital Roof in an Adult: Case Report and Literature Review

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ABSTRACT

Objective: To present the case of a leptomeningeal cyst involving the orbital roof in an adult. **Design:** Case report and literature review. **Setting:** Tertiary referral center. **Summary:** A 47-year-old female with a remote history of a skull fracture at 3 years of age presented with increasing headaches and retro-orbital pain. A computed tomogram and magnetic resonance image revealed a leptomeningeal cyst of the orbital roof. **Results:** Only one previous leptomeningeal cyst of the orbital roof has been reported in an adult. Surgical excision of the lesion was performed and follow-up imaging 18 months after the operation revealed no evidence of recurrence. **Conclusion:** Although extremely rare, adult patients can develop growing skull fractures or leptomeningeal cysts of the orbital roof. Such lesions should be included in the differential diagnosis when a patient presents with orbital pain or exophthalmos and a history of head trauma as a child.

KEYWORDS: Leptomeningeal cyst, growing skull fracture, orbital roof

Leptomeningeal cysts, commonly referred to as growing skull fractures, are uncommon complications of head trauma. These lesions are rare in adults, with over 90% resulting from fractures occurring in children under the age of 3 years.¹ Frequently located in the frontal or parietal skull, involvement of the orbit is rare. Although numerous reports of orbital roof involvement exist, only one describes these findings in an adult.² We now

present the second case of an adult with a traumatic leptomeningeal cyst of the orbital roof.

CASE REPORT

Our patient is a 47-year-old female who presented with a history of migraine headaches, noted to be

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Figure 1 Intraoperative photograph prior to surgical incision reveals slight exophthalmos of the right orbit.

worsening over the past 4 months. She complained of pain behind her right eye, more severe in the morning, with bilateral maxillary and mandibular pain. The patient described the pressure as constant, and reported that the symptoms were exacerbated with dependent head positioning. She denied any watery rhinorrhea, symptoms of recurrent sinusitis, decreased hearing, vertigo, vision changes, or diplopia. Notable in the patient's history was a skull fracture at age 3. Physical examination did not reveal any ocular abnormalities or neurologic deficits with the exception of mild exophthalmos

Figure 2 Coronal computed tomography image depicts a soft tissue lesion (arrow) extending through the orbital roof, lateral to the frontal sinus. A bony defect of the skull base with extension into the frontal bone is noted extending from the mass into the anterior cranial cavity.

(Fig. 1). A computed tomography (CT) scan of the sinuses revealed a defect in the orbital roof, with an isodense lesion extending superior and lateral to the frontal sinus (Fig. 2). A small defect was present in the anterior skull base, and this was continuous with the soft tissue mass in the superior orbit. Magnetic resonance imaging (MRI) of the mass was consistent with a cystic lesion (Fig. 3).

Figure 3 Noncontrast T1 magnetic resonance images reveal a cystic mass in the superior orbit, consistent with a leptomeningeal cyst. Extension of the cystic lesion (arrow) from the superior orbital vault into the anterior cranial cavity is seen in both (A) coronal and (B) sagittal views.

Figure 4 Intraoperative photograph reveals the fracture line extending along the frontal bone superiorly from the right orbital roof. The leptomeningeal cyst (arrow) is seen extending into the orbit.

The patient was subsequently taken to the operating room for excision of the cyst. Surgical exposure of the orbital roof was obtained through a coronal incision and subperiosteal dissection down to the orbital roof (Fig. 4). During the dissection, a vertical fracture was noted in the frontal bone, extending inferiorly into the orbital roof. Cerebrospinal fluid (CSF) was noted leaking from the fracture. Bone wax and Duraseal (Confluent Surgical, Waltham, MA) were placed over this area and the CSF leak resolved. Dissection continued around the orbital rim and posteriorly along the orbital roof. At this point, the leptomeningeal cyst was identified. It had created an indentation with dehiscence of the right supraorbital roof. The cyst lining was removed and the contents were evacuated by suction. An excess bar of bone that had been formed by osteoblastic changes was drilled down using a cutting burr. Postoperative histological examination of the tissue sent to pathology revealed fragments of bone and fibrous connective tissue consistent with a leptomeningeal cyst. Follow-up imaging 18 months after the operation revealed no evidence of recurrent disease.

DISCUSSION

The incidence of leptomeningeal cysts ranges from 0.05 to 0.6% of all skull fractures.^{3,4} The majority of these lesions occur in children, and the parietal skull is the most common location.⁵ A dural tear underlying a skull fracture is widely believed to be the initiating event, as originally suggested by Taveras and Ransohoff in 1953.⁶ With injury to the dura, arachnoid tissue becomes trapped within the fracture line. Cerebrospinal fluid collects inside the trapped leptomeninges, forming a cyst. Mechanical stress from the cyst and pulsating brain causes bone erosion and widening at the fracture site.^{7,8} In addition to mechanical stress, gravitational force has been suggested as an additional causative factor in the expansion of growing skull fractures at the skull base and orbital roof.⁹

Varied clinical presentations have been described in the literature. Children usually present with a nontender, pulsatile scalp mass or incidental findings of an enlarging skull fracture on radiologic imaging, while adults present with a nontender, nonpulsatile swelling and a distant history of

trauma.⁷ When the orbital roof is involved, the most common findings include exophthalmos, diplopia, and restricted ocular movements.^{2,9-12} Although the majority of patients present with complaints 2 to 12 months after the traumatic event, delay in presentation for 8 to 20 years has been reported.^{2,13} In the authors' case, the patient presented 44 years after the initial injury. Because the clinical presentation can vary significantly, a classification scheme has been suggested to clarify the different pathologic processes.¹⁴ Using this classification scheme, three types of growing skull fractures are defined. A Type I growing skull fracture includes herniated arachnoid tissue only (leptomeningeal cyst). Type II is defined by brain matter involvement (encephalocele). Type III is characterized by an additional porencephalic cyst of the underlying brain. Using this classification scheme, the previously reported adult case of the orbital roof would be considered a Type III growing fracture, while the authors' case would be described as a Type I growing fracture.

In addition to the history and clinical examination, imaging studies are helpful in making the diagnosis. Although a leptomeningeal cyst is usually identifiable on serial plain radiographs, a CT scan is required to precisely determine the extent of bone involvement.¹⁵ For definitive treatment planning, MRI is beneficial to distinguish the contents of the cyst (CSF versus cerebrum) and rule out other possibilities included in the differential diagnosis.

Cases of spontaneous stabilization of growing skull fractures have been reported.^{4,14,16} However, the decision to observe these lesions is controversial. Typically, surgical excision is recommended to prevent complications. Kutlay et al described nine patients who were diagnosed with growing skull fractures 8 to 13 years after the head injury.¹⁷ From these authors' review, stable lesions may undergo expansion after a period of quiescence and cause delayed neurological injury. They propose that the delayed cyst enlargement may be caused by hemorrhage. In our case, the delay in symptoms for greater than 40 years is unusual, and the cause for this prolonged interval from injury to clinical

presentation is unknown. In general, surgical treatment of a leptomeningeal cyst requires resection of the lesion and occasionally the herniated brain, followed by closure of the dura.¹⁸ The dural defect is typically closed using pericranium, and autologous calvarial bone grafts are preferred when cranioplasty is required.¹⁹

SUMMARY

Although rare, traumatic leptomeningeal cysts may involve the orbital roof. While usually presenting in the pediatric population within 1 year after injury, delayed presentation can occur many years later in an adult. In most cases, surgical treatment rather than observation is recommended.

REFERENCES

1. Lende RA, Erickson TC. Growing skull fractures of childhood. *J Neurosurg* 1961;18:479-489
2. Giuffrida M, Cultrera F, Antonelli V, Campobassi A, Servadei F. Growing-fracture of the orbital roof with post-traumatic encephalocele in an adult patient. *J Neurosurg Sci* 2002;46(3-4):131-134
3. Arsen C, Ciurea AV. Clinicotherapeutic aspects in the growing skull fracture. A review of the literature. *Childs' Brain* 1981;8:161-172
4. Ramamurthi B, Kalyanaraman S. Rationale for surgery in growing fractures of the skull. *J Neurosurg* 1970;32:427-430
5. Britz GW, Kim DK, Mayberg MR. Traumatic leptomeningeal cyst in an adult: a case report and review of the literature. *Surg Neurol* 1998;50:465-469
6. Taveras JM, Ransohoff J. Leptomeningeal cysts of the brain following trauma with erosion of the skull. *J Neurosurg* 1953;10:233-241
7. Britz GW, Kim DK, Mayberg MR. Traumatic leptomeningeal cyst in an adult: a case report and review of the literature. *Surg Neurol* 1998;50:465-469
8. Goldstein F, Sakoda T, Kepes JJ, Davidson K, Brackett CE. Enlarging skull fractures: an experimental study. *J Neurosurg* 1967;27:541-550
9. Amirjamshidi A, Abbassioun K, Tary AS. Growing traumatic leptomeningeal cyst of the roof of the orbit presenting with unilateral exophthalmos. *Surg Neurol* 2000;54:178-182
10. Bayar MA, Iplikcioglu AC, Kokes F, Gokcek C. Growing skull fracture of the orbital roof. *Surg Neurol* 1994;41:80-82

11. Caffo M, Germano A, Caruso G, Meli F, Calisto A, Tomasello F. Growing skull fracture of the posterior cranial fossa and of the orbital roof. *Acta Neurochir (Wien)* 2003; 145:201–208
12. Mohindra S, Mukherjee KK, Chhabra R, Gupta R. Orbital roof growing fractures: a report of four cases and literature review. *Br J Neurosurg* 2006;20:420–423
13. Colak A, Akbasak A, Biliciler B, Erten SF, Kocak A. An unusual variant of a growing skull fracture in an adolescent. *Pediatr Neurosurg* 1998;29:36–39
14. Naim-ur-Rahman, Jamjoom Z, Jamjoom A, Murshid W. Growing skull fractures: classification and management. *Br J Neurosurg* 1994;8:667–79
15. Jamjoom ZA. Growing fracture of the orbital roof. *Surg Neurol* 1997;48:184–188
16. Rothman L, Rose S, Laster DW, Quencer R, Tenner M. The spectrum of growing skull fractures in children. *Pediatrics* 1976;57:26–31
17. Kutlay M, Demircan N, Akin ON, Basekim C. Untreated growing cranial fractures detected in late stage. *Neurosurgery* 1998;43:72–76
18. Gupta SK, Reddy NM, Khosla VK, et al. Growing skull fractures: a clinical study of 41 patients. *Acta Neurochir (Wien)* 1997;139:928–932
19. Muhonen MG, Piper JG, Menezes AH. Pathogenesis and treatment of growing skull fractures. *Surg Neurol* 1995;43: 367–373