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Pediatric Periocular Dermoid Cysts: Incidence, Clinical Characteristics, and Surgical Outcomes

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Abstract

PURPOSE: To report the incidence, clinical findings, and surgical outcomes of periocular dermoid cysts diagnosed among children over a 20-year period.

METHODS: All patients 5 years of age, who were diagnosed with a periocular dermoid cyst in Olmsted County, Minnesota from 1986 to 2005 were reviewed to determine the population incidence, clinical presentation, and management outcomes.

RESULTS: A total of 54 children were diagnosed with a periocular dermoid cyst during the 20-year period, yielding a birth incidence of 1 in 638 live births. The mean age at diagnosis was 12 months (range, 1 month to 60 months) and 29 (53.7%) were female. Forty-four cysts (81.5%) occurred in the superotemporal orbital rim, 6 (11.1%) in the superonasal orbital rim, 3 (5.6%) in other periocular areas, and one (1.9%) within the orbit. Thirty-four (63%) children had an ophthalmic exam, all without amblyopia or other ocular sequelae. Forty-eight (88.9%) patients underwent surgical excision with 5 (10.4%) having documented intra-operative cyst rupture, none of whom had post-operative complications. Two (4.2%) other patients were, however, noted to have lesion recurrence following surgical intervention.

CONCLUSIONS: Periocular dermoid cysts occur in approximately 1 in 650 live births and most commonly occur in the superotemporal region of children aged 1 year or less. Ocular sequelae are rare and surgical excision often yields excellent results.

Dermoid cysts are benign congenital choristomas formed by the sequestration of ectoderm along the lines of embryonic fusion.¹ While found throughout the body, a majority are diagnosed in the head and neck, with a predilection for the periocular region.^{1–4} Cyst growth may lead to focal mass effects causing cosmetic deformity, mechanical ptosis, and proptosis. Cyst rupture can cause inflammatory sequelae including pain, edema, and redness. Histologically, dermoid cysts are filled with keratin and surrounded by keratinized stratified squamous epithelium with dermal appendages such as hair follicles, sebaceous glands, and

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sweat glands.^{1,5} Complete surgical excision is the definitive treatment.^{6,7} Previous studies have described the clinical characteristics and management of periocular dermoid cysts.^{2,4,6–10} However, to the best of our knowledge, there are no known population-based studies of periocular dermoid cysts. The purpose of this study is to determine the incidence and clinical characteristics of periocular dermoid cysts diagnosed among a population-based cohort of children over a 20-year period.

METHODS

Institutional review board approval was obtained prospectively from the Mayo Clinic and Olmsted Medical Group, which adhere to the Health Insurance Portability and Accountability Act. The resources of the Rochester Epidemiology Project (EP) were utilized to identify cases of periocular dermoid cysts in patients 5 years of age or younger who were diagnosed while residing in Olmsted County, Minnesota, between January 1st, 1986 and December 31st, 2005. We choose 2005 as the final year of the study in order to allow for at least 10 years of follow-up for cyst recurrence. We also chose to study children aged 5 years or younger, as most cysts are diagnosed in children in the first year or two of life. The REP is a medical records linkage system designed to capture data on any patient-physician encounter in Olmsted County, Minnesota.¹¹ Thus, this study is population-based.

The medical records of 333 patients with the following diagnoses were reviewed: dermoid cyst, cyst eyelid, cyst eye, cyst orbit, swelling periorbital, swelling or mass of eye, neoplasm benign eyelid skin, neoplasm benign eyelid, neoplasm benign orbit, and cyst dermoid skin. The periocular region was defined as involving the eyelids, orbit, eyebrow, glabella, and nasal bridge. Patients having a cyst in the orbital and periocular area and a pathology report confirming the diagnosis of a dermoid or epidermoid cyst were included. In the absence of a surgical pathology report for patients who did not have surgery (6 patients), a clinical diagnosis of dermoid cyst by the examining physician was used as the study inclusion criterion. Patients with any other type of periocular lesion or dermoid cysts located outside the periorbital region were excluded. The records of patients with orbital and periorbital dermoid cysts were reviewed for demographic information, birth and medical history, imaging studies, ophthalmologic reports, surgical reports, and pathology reports.

The incidence of periocular dermoid cysts was estimated using gender specific population figures for births in Olmsted County. Yearly incidence rates for each gender group was determined by dividing the number of cases within that group by the estimated total Olmsted County resident population of the group for that given year. Population figures come from the US census data every 10 years and population figures for intercensus years were estimated by using a linear interpolation.

RESULTS

Fifty-four cases of periocular dermoid cysts were diagnosed during the 20-year study period, yielding a birth incidence of 1 in 638 live births [Confidence Interval (CI) 1 in 491 to 1 in 850]. The demographics and clinical characteristics of the 54 patients are summarized in the Table 1. The mean age at diagnosis was 15 months (range 1–60 months), with 50% of

patients diagnosed before the first year of life. Twenty-nine (53.7%) patients were female. All patients presented with a periocular mass, without inflammation or erythema. The majority of patients (81.5%) had a dermoid cyst located in the superotemporal periocular region. Five (9.3%) of the 54 patients had other non-ocular congenital anomalies. The majority (n=34, 64%) of patients had a full ophthalmologic exam, and none had any ocular or periocular sequelae including ptosis or amblyopia. Patients who did not undergo an ophthalmic examination were managed by non-ophthalmologists such as plastic surgeons, craniofacial surgeons, and otolaryngology surgeons. One patient was diagnosed with amblyopia secondary to exotropia, and another patient with accommodative esotropia without amblyopia. A minority (n=14, 26%) of patients had imaging: 9 patients had a computed tomography (CT) scan [4/6 (67%) patients with a superonasal cyst, 4/44 (9%) patients with a superotemporal cyst and 1/1 (100%) patient with an orbital cyst], 4 patients had an X-ray (3 superotemporal, 1 central) and 1 patient had an ultrasound (central).

Surgical excision was performed for the majority (n=48, 89%) of patients. Five (10.4%) had documented unintentional intra-operative rupture of the cyst; however, none of these developed post-operative complications (including inflammation) or cyst recurrence. Five (10.4%) other patients had post-operative complications including hematoma (n=3, 5.6%), transient frontalis muscle weakness (n=1, 1.9%), and prominent scar (n=1, 1.9%). Two (4.2%) patients had cyst recurrence after surgical intervention; one recurred 8 months following surgery, while the second recurred one year after surgery. The patient with the 8-month recurrence had a superotemporal cyst that was excised by a direct incision over the lesion. This patient was noted to have a prominent scar as a post-operative complication, however no intra-operative complications were noted. The patient with the 1-year recurrence had a superonasal cyst that was excised through an upper eyelid crease incision (medial half) without intra or post-operative complications noted. Neither patient's family elected for additional surgery.

Histopathologic analysis demonstrated that 32 (66.7%) specimens were dermoid-type cysts, while the remaining 16 (33.3%) specimens were epidermoid-type cysts that lack dermal adnexal elements in the cyst wall. The mean volume of the excised cysts was 0.70 cm³ (SD 0.52 cm³). Only two (4.2%) cysts were found to have evidence of granulomatous inflammation.

DISCUSSION

In this population-based cohort from the Midwestern United States, a dermoid cyst occurred in approximately 1 in 638 live births. Most children presented with a cyst in the superotemporal region before the first year of life, without ocular findings, and responded well to surgical excision.

There are no other known population-based reports with which to compare these findings. Among previous studies, dermoid cysts comprised 19% to 46% of orbital masses excised from children, however, neither were from a defined population.^{12,13} This report reveals a greater birth prevalence of pediatric periocular dermoid cysts compared to population-based studies on some common childhood periocular disorders, including periocular infantile

hemangiomas (1 in 1586 live births) and congenital dacryocystoceles (1 in 3884 live births), and a similar birth prevalence to simple congenital eyelid ptosis (1 in 842 live births).^{14–16}

Consistent with previous reports, most periorbital dermoid cysts were found in the superotemporal region, corresponding to the frontozygomatic suture line.^{6–8} A small percentage (11%) presented with a superonasal lesion and only one lesion occurred in the orbit. Sherman et al. reported that children with superficial dermoids most often present in infancy, while the average age of children presenting with deep orbital dermoids was 18 years.¹⁰ The single orbital cyst identified in this cohort is likely due to our exclusion of children diagnosed beyond five years of age.

The majority (63%) of patients had a complete ophthalmologic exam, although none had any ocular or periocular sequelae including ptosis or amblyopia. However, any ocular sequelae are unknown in remaining one third of patients who were managed by non-ophthalmologists. Because previous reports have occasionally noted ptosis, proptosis or globe displacement secondary to the mass effect of dermoid cysts, it is reasonable to perform a complete ocular examination for these patients.^{4,7,8,10} However, the role of imaging remains somewhat controversial. While not required in routine superotemporal-based lesions, cysts located in the superonasal region or orbit are concerning for intracranial extension and may warrant further neuroimaging. Olmsted County physicians tended to follow this practice, as 4 out of 6 patients with a superonasal dermoid were imaged, as well as the single patient with an orbital dermoid cyst. A previously published treatment algorithm for the management of orbitofacial dermoids recommends imaging dermoids with indistinct margins, nasoglabellar masses, and sinuses or puncti caudad to the nasofrontal suture.⁷

Surgery was performed on a majority (88.9%) of the patients in this study, and while five (10.4%) were noted to have intra-operative rupture of the cyst, none had a recurrence or any post-operative complications. Although few in number, this series suggest that intraoperative cyst rupture may not have negative sequelae and could be intentionally performed to allow for easier cyst excision, particularly for those located within the orbit. There were, however, two (4.2%) patients without rupture who had later recurrence, neither of whom had additional surgery. For these patients, one was noted to have a prominent scar as a post-operative side-effect, suggesting surgical trauma. The other had a superonasal lesion that was approached through a medial half upper eyelid crease incision, which may have prevented proper cyst exposure and excision. It is possible that cyst recurrence may be due to remaining nests of cells trapped deep within bony suture lines. Nonetheless, the rate of surgical success in this cohort was favorable compared to previous literature.^{6–8}

Only two (4.2%) surgically excised dermoid cysts had evidence of granulomatous inflammation on histopathology, which differs significantly from two previous larger non-population-based reports.^{17,18} Abou-Rayyah et al. reported on 124 patients of all ages with periocular dermoid cysts that had undergone imaging, of which 29 of 42 (69%) patients 5 years of age or younger, had histopathological evidence of varying degrees of inflammation.¹⁷ Colombo et al. described 59 patients 5 years of age or less, of which 25% of the histopathological specimens revealed evidence of granulomatous inflammation.¹⁸ These

differences in inflammation rates, compared to the present study, may be due to the referral nature of the previous study populations which were conducted at large European referral hospitals.^{17,18}

There are several limitations to the findings of this study. Although most patients in Olmsted County are solely treated at two medical centers, there may have been patients who sought care elsewhere, thus underestimating the incidence of these cysts.¹¹ Furthermore, although most periocular dermoid cysts present before age 6, deeper orbital cysts typically present later in life, and thus may be underrepresented in this study that is limited to children 5 years of age or younger.^{6,10,19} Lastly, the ability to generalize these findings is limited by the demographics of Olmsted County, a relatively homogeneous semi-urban white population.¹¹

The principal findings of this study are that periocular dermoid cysts occur in approximately 1 in 650 live births and are most common in the superotemporal region of children aged one or less. Ocular sequelae are rare and surgical excision generally yields excellent results.

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REFERENCES

- 1). New GB, Erich JB. Dermoid cysts of the head and neck. *Surg Gynecol Obstet* 1937;65:48–55.
- 2). Pollard ZF, Harley RD, Calhoun J. Dermoid Cysts in Children. *Pediatrics* 1976;57(3):379–382. [PubMed: 1256948]
- 3). Orozco-Covarrubias L, Lara-Carpio R, Saez-De-Ocariz M, Duran-McKinster C, Palacios-Lopez C, Ruiz-Maldonado R. Dermoid cysts: a report of 75 pediatric patients. *Pediatr Dermatol* 2013;30(6):706–711. [PubMed: 23488469]
- 4). Pryor SG, Lewis JE, Weaver AL, Orvidas LJ. Pediatric dermoid cysts of the head and neck. *Otolaryngol Head Neck Surg* 2005;132(6):938–942. [PubMed: 15944568]
- 5). Reissis D, Pfaff MJ, Patel A, Steinbacher DM. Craniofacial dermoid cysts: histological analysis and inter-site comparison. *Yale J Biol Med* 2014;87(3):349–357. [PubMed: 25191150]
- 6). Shields JA, Kaden IH, Eagle RC, Shields CL. Orbital dermoid cysts: clinicopathologic correlations, classification and management. The 1997 Josephine E. Schueler Lecture. *Ophthalmic Plast Reconstr Surg* 1997;13(4):265–276. [PubMed: 9430304]
- 7). Bartlett SP, Lin KY, Grossman R, Katowitz J. The surgical management of orbitofacial dermoids in the pediatric patient. *Plast Reconstr Surg* 1993;91(7):1208–1215. [PubMed: 8497520]
- 8). Cavazza S, Laffi GL, Lodi L, Gasparrini E, Tassinari G. Orbital dermoid cyst of childhood: clinical pathologic findings, classification and management. *Int Ophthalmol* 2011;31(2):93–97. [PubMed: 21267629]
- 9). McAvoy JM, Zuckerbraun L. Dermoid cysts of the head and neck in children. *Arch Otolaryngol* 1976;102(9):529–531. [PubMed: 962697]
- 10). Sherman RP, Rootman J, Lapointe JS. Orbital dermoids: clinical presentation and management. *Br J Ophthalmol* 1984;68(9):642–652. [PubMed: 6466593]
- 11). Melton LJ. History of the Rochester Epidemiology Project. *Mayo Clin Proc* 1996;71(3):266–274. [PubMed: 8594285]
- 12). Kodsi SR, Shetlar DJ, Campbell J, Garrity JA, Bartley GB. A review of 340 orbital tumors in children during a 60-year period. *Am J Ophthalmol* 1994; 117(2):177–182. [PubMed: 8116746]

- 13). Shields JA, Bakewell B, Augsburger JJ, et al.: Space-occupying orbital masses in children: a review of 250 consecutive biopsies. *Ophthalmology* 1986;93(3):379–384. [PubMed: 3703507]
- 14). Griepentrog G, Diehl N, Mohny BG. Incidence and demographics of childhood ptosis. *Ophthalmology* 2011;152(2):314–319.
- 15). Alniemi ST, Griepentrog GJ, Diehl N, Mohny BG. Incidence and clinical characteristics of periocular infantile hemangiomas. *Arch Ophthalmol* 2012;130(7):889–894. [PubMed: 22776927]
- 16). Shekunov J, Griepentrog GJ, Diehl N, Mohny BG. Prevalence and clinical characteristics of congenital dacryocystocele. *J AAPOS* 2010;14(5):417–420. [PubMed: 21035068]
- 17). Abou-Rayyah Y, Rose GE, Konrad H, Chawla SJ, Moseley IF. Clinical, radiological and pathological examination of periocular dermoid cysts: evidence of inflammation from an early age. *Eye* 2002;16(5):507–512. [PubMed: 12194059]
- 18). Colombo F, Holbach LM, Naumann GOH. Chronic inflammation in dermoid cysts: a clinicopathologic study of 115 patients. *Orbit* 2000;19(2):97–107. [PubMed: 12045954]
- 19). Ahuja R, Azar NF. Orbital dermoids in children. *Semin Ophthalmol* 2006;21(3):207–211. [PubMed: 16912019]

Table 1.

Demographics and Clinical Characteristics of Children Diagnosed with Periorbital Dermoid Cysts from 1986 to 2005 in Olmstead County, Minnesota

Gender		Number (%)
Female		29 (53.7)
Male		25 (46.3)
Ethnicity		
Caucasian		38 (70.4)
Asian		4 (7.4)
African American		2 (3.7)
Hispanic		2 (3.7)
Unknown		8 (14.8)
Age at Diagnosis		
< 1 year old		27 (50.0)
1 year old		14 (25.9)
2 years old		7 (13.0)
3 years old		2 (3.7)
4 years old		1 (1.9)
5 years old		3 (5.6)
Laterality		
Left		32 (59.3) (p = 0.17)
Right		21 (38.9)
Central		1 (1.9)
Location		
Superotemporal		44 (81.5)
Superonasal		6 (11.1)
Orbital		1 (1.9)
Other		3 (5.6)
Upper eyelid		
Central eyebrow		
Supraorbital ridge		
Congenital Anomalies		5 (9.3)
Syndactyly or Polydactyly		2 (3.7)
Sagittal and coronal synostosis		1 (1.9)
Atrial septal defect		1 (1.9)
Torticollis		1 (1.9)
Epidermoid cyst of penis		1 (1.9)