Incidence and Clinical Characteristics of Epiretinal Membranes in Children

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Objective: To describe the incidence and clinical characteristics of epiretinal membranes (ERMs) in children.

Methods: The medical records of all pediatric (aged < 19 years) patients diagnosed as having an ERM from January 1, 1976, through December 31, 2005, at Olmsted Medical Group and Mayo Clinic were retrospectively reviewed.

Results: Five of the 44 patients were residents of Olmsted County, Minnesota, at the time of their diagnosis, yielding an annual age- and sex-adjusted incidence of 0.54 per 100,000 patients, or 1 in 20,896 patients younger than 19 years. The mean age at diagnosis of the study patients was 12.4 years (range, 4 months to 18 years), with a preponderance of boys (31 [70%]). The presenting visual acuity in the affected eye was 20/60 or less in 22 patients (50%), and 10 patients (23%) displayed strabismus. Common causes of ERMs were trauma (17 patients [39%]), idiopathic conditions (13 patients [30%]), and uveitis (9 patients [20%]). Of the 44 patients, 8 (18%) underwent pars plana vitrectomy with membrane peel, with at least 5 of the 8 experiencing an improvement in their postoperative visual acuity.

Conclusions: Epiretinal membranes are rare in children and are most frequently associated with a traumatic, idiopathic, or uveitic cause. Patients treated surgically generally have a favorable outcome.

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Epiretinal membranes (ERMs) are characterized by wrinkling or distortion of the macular surface caused by retinal cell proliferation. Also known as macular pucker or cellophane maculopathy, ERMs most often occur in patients older than 50 years and are usually bilateral although often asymmetrical. In adults, ERMs generally occur in association with ocular disease, idiopathically, or after retinal reattachment surgery. Less commonly, ERMs can also occur in younger patients after trauma or from ocular disorders, such as uveitis, retinal vascular disease, or tumors. Although there are studies of ERMs in patients up to age 40 years, 1,2 there are no studies, to our knowledge, devoted exclusively to children. The purpose of this study is to describe the incidence and clinical characteristics of ERMs in patients younger than 19 years studied during a 30-year period.

Methods

Institutional review board approval at Mayo Clinic and Olmsted Medical Group was obtained for this study. The medical records of all patients younger than 19 years diagnosed at our institution as having an ERM from January 1, 1976, through December 31, 2005, were retrospectively reviewed. We also identified pediatric patients who were residents of Olmsted County, Minnesota, when diagnosed as having an ERM during the same period. Olmsted County cases were identified by using the resources of the Rochester Epidemiology Project, a medical records linkage system designed to capture data on any patient-physician encounter in this county. 3 The population of Olmsted County (106,470 in 1990) is relatively isolated from other urban areas, and virtually all medical care is provided to residents by Mayo Clinic or Olmsted Medical Group and their affiliated hospitals. We also reviewed the ophthalmic records of all pediatric patients who underwent fundus photography or optical coherence tomography for any diagnosis of macular abnormality during the same interval.

Of 153 potential patients generated by the diagnostic code search from these 2 institutions, 38 were eliminated based on their age at diagnosis. Of the remaining 95 patients, 31 were excluded owing to an incorrect diagnosis, including 2 with persistent fetal vasculature. The remaining 44 patients were included in this study. Historical characteristics concerning age, race, sex, trauma, date of onset of symptoms, and date of diagnosis were collected. The ophthalmic record was carefully reviewed for visual acuity, ocular misalignment, anterior segment findings, and refractive error. Funduscopy examination included indirect ophthalmoscopy of the pos-
Forty-four new cases of ERMs in children were diagnosed during the 30-year study. There were 13 girls (30%) and 5 (11%) of the study patients exhibited bilateral involvement. The mean age at diagnosis of the 44 study patients was 12.4 years (range, 4 months to 18 years). Epiretinal membranes tended to be more common in boys than in girls in the second decade of life (Figure 1). The presenting visual acuity in the affected (or worse) eye was 20/60 or less in 22 patients (50%), and 10 of the 44 children (23%) displayed strabismus. Seventeen of the ERMs (39%) were associated with trauma, 13 (30%) were idiopathic, 9 (20%) occurred in the setting of uveitis, and the remaining 5 (11%) developed in association with other causes (Table 1).

Of the 44 patients, 5 (11%) resided in Olmsted County at the time of their diagnosis, yielding an annual age- and sex-adjusted incidence of 0.54 per 100,000 persons, or 1 in 20,896 patients younger than 19 years. Four of the 5 patients were boys. The mean age at diagnosis of the 5 patients was 13.2 years (range, 5-18 years). All 5 patients had unilateral involvement, and 1 patient displayed strabismus. The initial median visual acuity of the affected eye was 20/40, with a final median visual acuity of 20/125 during a mean follow-up of 6.1 years. Three of these ERMs were secondary to trauma, and all 5 were managed with observation alone.

Fundus imaging was available in 27 of the 44 patients (61%), and the images were reviewed by a retinal specialist (C.A.M.) who made these determinations without previewing the patients’ medical records or diagnoses. One of these patients had bilateral ERMs, yielding 28 eyes with ERMs. Epiretinal membranes of the PMF type were the most frequently diagnosed form, found in 21 of 28 eyes with ERMs (75%), and ERMs of the CMR type were the most frequently diagnosed form, found in 14 of 28 eyes (25%). A representative fundus photograph and corresponding optical coherence tomography image of the 3 primary forms of ERM (traumatic, idiopathic, and uveitic) are shown in Figure 2.
The median visual acuity from first to last examination (mean duration, 3.1 years) improved from 20/65 to 20/40 (from 0.52 to 0.3 logMAR) for the 44 study patients. Of the 44 patients, 24 (55%) with ERMs were managed with observation alone, 8 (18%) were managed with pars plana vitrectomy, and 1 (2%) was managed with corticosteroids. The remaining 11 patients (25%) were followed up elsewhere, and treatment information was unavailable. Figure 3 illustrates the change in visual acuity in patients managed with surgery vs those who were observed (ie, 26 of the 44 patients with sufficient data followed up at Mayo Clinic). Table 2 outlines surgical outcomes for the 8 patients who were managed with pars plana vitrectomy and membrane peel. The final postoperative visual acuity for these patients demonstrated a mean improvement of 3.8 lines during mean follow-up of 33 months. The vision in all 4 of the uveitic patients and at least 1 of the idiopathic patients improved postoperatively. One of the 8 patients had bilateral ERMs secondary to uveitis and underwent a surgical procedure in each eye. Postoperative complications included cataract formation (1 patient) and an ERM recurrence requiring a second membrane peel (1 patient).
This study describes a population-based incidence rate and the clinical characteristics of ERMs from a large cohort of pediatric patients. Membrane formation occurred in approximately 1 in 20,896 individuals younger than 19 years in Olmsted County. There was a preponderance of boys, with most membranes occurring in the second decade of life. The initial visual acuity of the affected eye was 20/60 or less in 50% of the patients, and 23% presented with strabismus. Surgical intervention, although relatively uncommon in this study, resulted in improved visual acuity for most patients.

This study also reports the common causes of ERMs in children. Trauma to the eye or orbit occurred in 17 of 44 patients (39%), and boys composed 16 of these 17 patients. Boys, especially in the second decade of life, are more likely than similarly aged girls to engage in risk-taking behaviors that may expose them to ocular trauma and subsequent membrane formation. Isolated anterior segment injury was the most common form of trauma leading to ERM formation in this cohort. Membranes associated with uveitis occurred in 1 of 5 children in this study and were most often a result of chronic inflammation with cystoid macular edema. Idiopathic membranes were found in approximately 30% of patients, with clinical features similar to the other forms of ERMs. Banach and associates examined ERMs in 20 eyes in patients younger than 40 years, and all had the idiopathic cause. Benhamou and associates, reporting on 20 patients aged 7 to 26 years, found an idiopathic cause in 11 (55%) of their patients.

The present study also reports the relative prevalence of the 2 main varieties of ERMs in children. The cellophane macular reflex type, often considered the most common and benign form of ERMs, was relatively uncommon in this population, found in only 25% of all patients. This difference may be attributed to the relatively benign symptoms of the CMR-type ERMs, which may make them less likely to come to attention. The more severe PMF type of ERMs was diagnosed in 3 of 4 chil-

<table>
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<th>Age at Diagnosis, y</th>
<th>Cause</th>
<th>Type</th>
<th>VA in Affected Eye (LogMAR)</th>
<th>Change in VA, LogMAR Lines</th>
<th>Follow-up, mo</th>
<th>Complications</th>
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<tr>
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Abbreviations: ERM, epiretinal membrane; NA, not available; PMF, premacular fibrosis; PPV, pars plana vitrectomy; VA, visual acuity.
dren in this study. Although Banach and associates\textsuperscript{1} did not classify the type of ERM in their patients, Benhamou and associates\textsuperscript{2} reported findings similar to this study. They described 13 of 20 eyes (65\%) as having a “thick, white, and contractile” membrane (similar to the PMF designation) and the remaining 7 eyes as having a “thin transparent membrane” (similar to the CMR designation).\textsuperscript{2} The predominance of the PMF type of ERM in this study may be related to the high prevalence of the traumatic and uveitic causes, both of which may be more prone to significant injury and inflammation. Such a classification system seems to be useful in assessing clinical characteristics and surgical outcomes.

Published studies on the management of ERMs in young patients have demonstrated that membrane peeling can be safely performed, with an improvement in visual acuity.\textsuperscript{1,2} Spontaneous peeling of the ERM in young patients has been reported,\textsuperscript{6,7} and observation alone may be warranted. Banach and associates\textsuperscript{1} compared 2 groups of patients younger than 40 years with idiopathic ERMs. One group, with a visual acuity of 20/50 or better, was merely observed, and the second group, with a visual acuity of 20/60 or worse, underwent surgery. Of 13 patients, 10 (77\%) who underwent surgical peeling experienced an improvement in vision, and 50\% of the patients managed with observation alone had stable or improved vision. Benhamou and colleagues\textsuperscript{2} found ERM peeling to be safe in 20 young patients who had an improvement in postoperative visual acuity.

The 8 patients who underwent pars plana vitrectomy in this study had a mean visual improvement of 3.8 lines, which is comparable to the 4.25 lines of improvement reported in a previous study of young patients.\textsuperscript{2} One of the patients in this study subsequently developed a nuclear sclerotic cataract, and another developed a recurrent ERM sufficient to require a second peeling. Subsequent operation for an ERM recurrence in young patients has been reported to be as high as 5 of 20 patients (25\%).\textsuperscript{1}

Pars plana vitrectomy with membrane peeling is a viable treatment option for pediatric ERMs and seems to be as safe in children as it is in adults. Surgical intervention may be more successful for ERMs secondary to chronic uveitis compared with idiopathic membranes, and a subsequent procedure may be required. Only 1 of the 8 patients in this study who underwent pars plana vitrectomy had a recurrent ERM requiring a second procedure. This relatively low rate may be owing to variations in surgical technique and equipment at different institutions.

There are several limitations to the findings in this study. The reported incidence rate may be less than the true value because this condition is often discovered on routine eye examination, and some patients with an ERM may have gone unnoticed by the caretaker, thereby avoiding an evaluation by the study ophthalmologists. Moreover, some residents may have sought care outside of Olmsted County. This study is also limited by its retrospective nature. The diagnostic criteria and methods of examination have evolved during the past 30 years; however, we retrospectively reviewed the available imaging for 27 of the 44 study patients (61\%) for confirmation and consistency of diagnosis.

We found that ERMs occur in approximately 1 in 21,000 children. The most common causes were trauma, uveitis, and idiopathic, with a preponderance of boys, especially in the second decade of life. For patients who underwent pars plana vitrectomy with membrane peel, most experienced an improvement in vision.

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REFERENCES