gest that the term paraneoplastic melanocytic proliferation rather than BDUMP syndrome be used to describe this entity because it more accurately reflects the clinical spectrum and pathogenesis of this unique paraneoplastic syndrome.

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**Limited Wegener Granulomatosis With 40 Years of Follow-up**

Wegener granulomatosis (WG) classically consists of necrotizing granulomatous inflammation of the upper and/or lower respiratory tract, necrotizing granulomatous vasculitis, usually affecting small vessels; and focal segmental glomerulonephritis.1 A limited form occurs, however, in which there is no renal involvement.2,3 We report a case of limited WG with apparent orbital involvement and nearly 40 years of follow-up.

**Report of a Case.** A 14-year-old white girl had painless swelling of her right upper eyelid and diplopia 1 week after a bout of tonsillitis in 1962. The eyelid swelling originally appeared transiently 2 to 3 months prior to the initial visit and then disappeared completely. The tonsillitis was treated with antibiotics, steroids, and tonsillectomy. Examination showed visual acuity of 20/20 OU, marked ptosis of the right upper eyelid, and a firm, nontender, immobile mass below the supraorbital rim, extending posteriorly.

A general physical examination revealed normal vital signs, lungs, heart, abdomen, and integument. Urinalysis findings were normal. The mass effect and ptosis partially improved during the next 4 months with oral corticosteroids and antibiotics. Proptosis of 2 mm OD remained. Orbital pseudotumor was tentatively diagnosed, and an anterior orbitotomy through the right upper eyelid was performed to allow histopathologic diagnosis. A white, fibrous, nonencapsulated mass was partially resected. The diplopia improved postoperatively but the ptosis and proptosis persisted.

Histopathologic examination revealed dense fibrous tissue containing scattered lymphoid follicles and many eosinophils (Figure 1). Neither granuloma formation nor vasculitis was seen, and a diagnosis of chronic nonspecific dacryoadenitis was made. Oral steroids, nonsteroidal anti-inflammatory medications, and chemotherapeutic agents were not used postoperatively. The patient’s right upper eyelid ptosis was corrected by levator resection in 1964.

The patient exhibited dyspnea and dysphonia in 1973 and 1979. Examination by an otolaryngologist revealed subglottic stenosis on both occasions, and the patient was treated with laser fulguration. In 1979, she additionally required a tracheostomy, which was removed uneventfully a year later. Histopathologic ex-

Figure 1. Histopathologic findings from the orbitotomy in 1962 show dense fibrous tissue and numerous eosinophils without evidence of granulomas or vasculitis.
amination revealed nonspecific chronic inflammation without vasculitis or granulomas (Figure 2). In 1973, a saddle nose deformity was noted (Figure 3).

In 1985, the patient had worsening proptosis secondary to slow growth of the right orbital mass. Her best-corrected visual acuity was 20/70 OD and 20/20 OS. The right eye showed hypotropia, exotropia, decreased elevation and adduction, and an afferent pupillary defect. Exophthalmometry measured 23 mm OD and 14 mm OS. Intraocular pressure measured 19 mm Hg OD and 15 mm Hg OS. Lagophthalmos of 3 mm was present on the right.

Hematologic testing revealed an erythrocyte sedimentation rate of 32 mm/h. Antinuclear, mitochondrial, and smooth muscle antibody tests were negative. Findings from urinalysis and chest radiographs were normal. Orbital computed tomography without contrast revealed right maxillary sinus disease and marked enlargement of the right lateral and medial rectus muscles (Figure 4). A contiguous soft tissue mass extended into the superior aspect of the right orbit.

The patient underwent another anterior orbitotomy with biopsy and debulking of the orbital mass. Histopathologic examination revealed necrotizing granulomatous inflammation and vasculitis with accumulations of lymphocytes and histiocytes (Figure 5). Staining for CD68 was performed subsequently, owing to its specificity for macrophages, and revealed a strongly positive vasculitis (Figure 5). Staining for acid-fast organisms and fungi was negative. The patient’s pressure sensation and ocular motility improved, and proptosis was reduced by 5 mm postoperatively, but the right afferent pupillary defect persisted. Again, oral steroids, nonsteroidal anti-inflammatory agents, and chemotherapeutic agents were not used postoperatively.

In 1993, the patient exhibited stridor, dyspnea, and epiphora of the right eye. Ophthalmic evaluation revealed nasolacrimal duct obstruction. Computed tomography revealed subglottic stenosis. Laser fulguration of the patient’s subglottic stenosis and right external dacryocystorhinostomy were performed uneventfully.

In 2000, the patient had epiphora and fullness over the nasolacrimal sac on the left. Nasolacrimal duct obstruction with dacrocystocele was diagnosed, and left external dacryocystorhinostomy was performed. A lacrimal sac biopsy revealed chronic granulomatous inflammation. Vasculitis, granulomas, and multinucleated giant cells were not observed.

Results of urinalysis and renal function tests have been normal. Proteinase 3, c-antineutrophil cytoplasmic antibody and p-antineutrophil cytoplasmic antibody tests were performed in 2001 and were negative. Results of rapid plasma reagent for syphilis have been normal. Physical examination and chest radiographs have never revealed signs of lung disease. The patient’s visual acuity has been stable at 20/50 OD and 20/20 OS. The most recent Hertel exophthalmometry showed 3 mm of proptosis on the right. The right eyelid ptosis has improved but persists. The epiphora in both eyes resolved following bilateral external dacrystocystorhinostomy. She has not had dyspnea since her subglottic stenosis in 1993.

Comment. Prior to 1985, this patient did not exhibit necrotizing granulomatous inflammation, so a tentative diagnosis of idiopathic midline destructive disease, a subgroup of the midline granuloma syndrome, was made because of the nonspecific histopathologic specimens and lack of systemic disease. The diagnostic difficulties were exacerbated by the histopathologic findings prior to 1985, revealing nonspecific inflammation, and the biopsy in 1985, whose findings were consistent with WG but not pathognomonic. Fifty percent of orbital biopsies for suspected WG are nonconclusive. In addition, there was a paucity of...
tissue from the biopsies of the subglottic stenosis in 1973 and 1979, and c-antineutrophil cytoplasmic antibody is often negative in cases of limited WG, with false-negative rates as high as 68%. Furthermore, this test was performed late in the course of the patient’s disease, when few symptoms were apparent. Earlier or repeated testing may have yielded a positive result. This patient lacked renal involvement, which is classically associated with WG. Also, pulmonary nodules, another common finding, were not present.

The patient was ultimately diagnosed as having limited WG. Her saddle nose deformity, clinical signs and symptoms, and the specimen from 1985, which was strongly CD68 positive and contained necrotizing granulomatous vasculitis, were consistent with WG. In addition, specimens from the orbitotomy in 1962, the area of subglottic stenosis in 1973, and the orbitotomy in 1985 were sent to the National Institutes of Health (Bethesda, Md) in 1986. Necrotizing vasculitis was seen in the specimens from 1962 and 1985, and numerous granulomas were seen in the specimen from 1985. No granulomas were seen in the walls of vessels. Classic WG is usually rapidly fatal without treatment with agents such as cyclophosphamide and corticosteroids. However, neither agent was used in this patient given the slowly progressive course and the risks associated with long-term treatment. In addition, proptosis and subglottic stenosis are often refractory to treatment with cyclophosphamide and corticosteroids. This case shows the value of long-term follow-up, the occasional need for multiple biopsies, and the necessity to combine the clinical and pathologic findings of disease to achieve a diagnosis of limited WG.

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