
protection against bacterial pathogens and compensates for B-cell dysfunction.

In summary, CVID is a heterogeneous immunodeficiency syndrome that spans a broad clinical spectrum. A small subset of patients with a presumptive diagnosis of CVID is spared recurrent infections despite hypogammaglobulinemia and impaired antibody responses to vaccines. Identification and registry of these patients will lead to broader awareness of CVID and will offer an opportunity to study potential innate mechanisms harnessed by the immune system to compensate for cognate defects.

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REFERENCES

1. Cunningham-Rundles C, Bodian C. Common variable immunodeficiency: clinical and immunological features of 248 patients. *Clin Immunol*. 1992;92:34–48.
2. Bonilla FA, Bernstein IL, Khan DA, et al. Practice parameter for the diagnosis and management of primary immunodeficiency. *Ann Allergy Asthma Immunol*. 2005;94(suppl 1):S1–S63.
3. Hermans PE, Diaz-Buxo JA, Stobo JD. Idiopathic late-onset immunoglobulin deficiency: clinical observations in 50 patients. *Am J Med*. 1976;61:221–237.
4. Carbone J, Sarmiento E, Rodriguez-Molina JJ, Fernandez-Cruz E. Atypical presentation of common variable immunodeficiency without infections [in Spanish]. *Allergol Immunopathol (Madr)*. 2004;32:218–222.
5. Abonia JP, Castells MC. Common variable immunodeficiency. *Allergy Asthma Proc*. 2002;23:53–57.
6. Morimoto Y, Routes JM. Granulomatous disease in common variable immunodeficiency. *Curr Allergy Asthma Rep*. 2005;5:370–375.
7. Gompels MM, Hodges E, Lock RJ, et al. Lymphoproliferative disease in antibody deficiency: a multi-centre study. *Clin Exp Immunol*. 2003;134:314–320.

THE PANCAKE SYNDROME (ORAL MITE ANAPHYLAXIS) BY INGESTION AND INHALATION IN A 52-YEAR-OLD WOMAN IN THE NORTHEASTERN UNITED STATES

Oral anaphylaxis to ingested mites is a severe and potentially life-threatening reaction that has been reported primarily in semitropical and tropical climates. We describe a patient from the northeastern United States who experienced anaphylaxis on 2 separate occasions after ingesting and inhaling

cooking vapors from a commercial pancake mix contaminated with the house dust mite *Dermatophagoides farinae*. The diagnosis was made by visual inspection of and skin testing with the pancake mix.

A 52-year-old female resident of Danvers, Massachusetts, experienced her first episode of eye swelling and lip tingling shortly after eating pancakes made from a buttermilk pancake mix (Krusteaz Pancake Mix; Continental Mills, Seattle, Washington) that had been stored in her kitchen cabinet for 2 years. This reaction was initially attributed to an unknown environmental allergen, because the patient had a history of allergy to dust mites, pollen, mold, and dander, but no history of food allergy. Three months later, while cooking pancakes from the same mix, she experienced wheezing, diffuse redness, and facial swelling that again required emergency department care. The patient was not aspirin sensitive, but was taking a β -blocker for hypertension.

On allergy evaluation, she had positive skin prick test results (15-mm wheal) to house dust mites (*D farinae* and *Dermatophagoides pteronyssinus*; ALK Labs, Round Rock, Texas) and a 9-mm wheal and a 25-mm flare to the pancake mix. Skin prick test results to wheat were negative, and the patient had no history of problems after eating wheat-based products, including pancakes. Although grossly normal in appearance, microscopic examination of the pancake mix revealed a thriving colony of live mites, easily seen under low power (a video is available at <http://www.annallergy.org>). Larry Arlian, PhD, at Wright State University, Dayton, Ohio, confirmed the species as *D farinae*, and a 2-site monoclonal antibody enzyme-linked immunosorbent assay (Indoor Biotechnologies Inc, Charlottesville, Virginia) measured a Der f1 concentration of 322 $\mu\text{g/g}$ of the pancake mix.

A review article¹ in this journal described relatively few published cases of anaphylaxis from oral mite ingestion. To our knowledge, Erben et al² reported the first case of anaphylaxis to mite-contaminated food in 1993, when they described a 48-year-old house dust mite-allergic man who developed anaphylaxis after eating beignets contaminated with *D farinae*. Blanco et al³ have described patients from Spain and Venezuela, some of whom were aspirin sensitive, who experienced severe allergic reactions, some repeated and life threatening, to foodstuffs that contained high quantities of mites. Because most of their patients reacted to mite-contaminated pancake flour, they labeled this condition the “pancake syndrome.” Other reports^{4–6} describe additional cases of anaphylaxis after the ingestion of grains contaminated with mites, often storage mites.

In April 2006, a “Dear Abby” letter⁷ described a 14-year-old boy who experienced dyspnea and cyanosis 10 minutes after eating an outdated pancake mix. This letter cited a 2001 publication⁸ by medical examiners from Charleston, South Carolina, that described the death of a 19-year-old asthmatic man after the ingestion of pancakes made from a packaged mix that had been opened and stored in a cabinet for approximately 2 years. Autopsy showed laryngeal edema and hyperinflated lungs with mu-

Disclosures: Dr Miller is owner and CEO of Mission:Allergy, Inc, a manufacturer and distributor of products for allergen avoidance.

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cous plugging consistent with an asthma-related death. The pancake mix was analyzed and found to contain 700 CFU of mold per gram of mix. The researchers presented this case as an unusual case of anaphylaxis to mold-contaminated pancake mix in a patient with mold allergy.

However, anaphylactic reactions in mold-allergic patients to molds in fermented foods, such as cheese, dried fruits, yogurt, and wine, are quite rare, if even existent. The term *farinae* (as in *D farinae*) means “wheat,” and wheat products can be used as culture media for mites. High humidity favors the growth not only of mold but also of mites. In our opinion, the “Dear Abby” case and the death of the 19-year-old man in Charleston were more likely because of mites rather than mold in the pancake mix.

The message from our case report and others cited in the literature is that anaphylaxis from oral ingestion of mites—or from inhalation of mite-contaminated grain mixtures—is a severe, potentially lethal, allergic condition that may be frequently undiagnosed. Although the present patient was not aspirin sensitive, her use of a β -blocker may have increased the severity of her reaction. Most previous cases have been reported in tropical and subtropical countries. Our case from the northeastern United States suggests that this problem may be more prevalent in temperate climates than previously recognized. A female dust mite can lay 60 eggs in its 80-day lifetime. With it taking only weeks for a dust mite to reach maturity, contamination of a wheat product with only a few mites can, in conditions of adequate humidity, lead to a thriving culture within months.

Flour-based products should be stored in a refrigerator or freezer after purchase. Allergy specialists should microscopically examine suspected foodstuffs in cases of unexplained or idiopathic anaphylaxis.

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REFERENCES

1. Sanchez-Borges M, Suarez-Chacon R, Capriles-Hulett A, Caballero-Fonseca F. An update on oral anaphylaxis from mite ingestion. *Ann Allergy Asthma Immunol.* 2005;94:216–221.
2. Erben AM, Rodriguez JL, McCullough J, Ownby DR. Anaphylaxis after ingestion of beignets contaminated with *Dermatophagoides farinae*. *J Allergy Clin Immunol.* 1993;92:846–849.
3. Blanco C, Quiralte J, Castillo R, et al. Anaphylaxis after ingestion of wheat flour contaminated with mites. *J Allergy Clin Immunol.* 1997;99:311–314.
4. Sánchez-Borges M, Capriles-Hulett A, Fernández-Caldas E, et al. Mite-contaminated foods as a cause of anaphylaxis. *J Allergy Clin Immunol.* 1997;99:738–743.
5. Wen DC, Shyur SD, Ho CM, et al. Systemic anaphylaxis after the ingestion of pancake contaminated with the storage mite *Blomia freemani*. *Ann Allergy Asthma Immunol.* 2005;95:612–614.
6. Sánchez-Borges M, Capriles-Hulett A, Caballero-Fonseca F. Oral mite anaphylaxis (pancake syndrome) also observed in children. *Ann Allergy Asthma Immunol.* 2006;96:755–756.
7. Dear Abby: out-of-date pancake mix can harbor mold. *OC Register.* April 14, 2006. Available at: http://www.ocregister.com/ocregister/life/columns/article_1100653.php.
8. Bennett AT, Collins KA. An unusual case of anaphylaxis: mold in mix. *Am J Forensic Med Pathol.* 2001;22:292–295.

ADOLESCENT PRESENTATION OF X-LINKED LYMPHOPROLIFERATIVE DISEASE

X-linked lymphoproliferative disease (XLP; OMIM 308240) is characterized as a rare immunodeficiency that presents with Epstein-Barr virus (EBV) infection and has an average age of onset of 2.5 years.¹ The molecular characterization of XLP has led to improved and rapid diagnosis and advanced our understanding in immune responses and development. Gene sequencing in patients with XLP has revealed *SAP/SH2D1A* gene (chromosome Xq25 encodes the SAP protein) mutations in 60% of patients.² Recently, X-linked inhibitor-of-apoptosis deficiency has been reported to cause the clinical phenotype of XLP in 3 families.³ Since SAP is expressed in CD8⁺ T cells, a rapid screening test was developed recently for detection of SAP protein by flow cytometry.⁴

The patient was a previously healthy 16-year-old boy who presented with a 2.5-week history of cough, sore throat, and fever. Tonsillar exudates, lymphadenopathy, and a positive Monospot test result were documented. Five days later, he was admitted to a local hospital for dehydration and was treated with intravenous fluids and corticosteroids. After 4 days, he was transferred to a tertiary care hospital because of hepatic failure. On examination, he was febrile and appeared ill. He displayed respiratory distress, ascites, hepatosplenomegaly, jaundice, and generalized lymphadenopathy. The patient developed respiratory distress that required assisted ventilation and coagulopathy. He was treated with antibiotics, ganciclovir, and intravenous immunoglobulin. The patient had no history of recurrent infections, autoimmune disease, or prior known EBV infection. The family history was unremarkable in first-degree relatives, but a maternal great-uncle died at the age of 9 years of uncharacterized meningoencephalitis and pneumonia (Fig 1A).

EBV titers indicated an acute infection (EBV early antigen, EBV viral capsid antigen IgG, and EBV viral capsid antigen IgM positive and EBV nuclear antigen IgG negative). Polymerase chain reaction for EBV demonstrated 529,500 copies/mL. Immunoglobulin levels were polyclonally elevated, and flow cytometry from the peripheral blood demonstrated elevated B-cell counts and an inverted CD4⁺/CD8⁺ T-cell ratio of 0.7.

On the third day of hospitalization, while awaiting liver transplantation, the patient developed pulmonary hemorrhage that failed to respond to aggressive intervention. The patient's course deteriorated rapidly and a cardiopulmonary arrest ensued. Resuscitative efforts were withheld according to the family's request and the patient died.

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The pancake syndrome (oral mite anaphylaxis) by ingestion and inhalation in a 52-year old woman in the northeastern United States. *Ann Allergy Asthma Immunol.* 2008;100:397-8. A 17-year-old boy and a 15-year old boy presented with a 10-month and 3-month history, respectively, of erythema, small wheals, and pruritus after contact with water. Symptoms developed after taking a shower or swimming (sea and swimming pool), regardless of the temperature and type of water. Lesions appeared predominantly on the trunk and usually disappeared spontaneously in minutes. Oral mite anaphylaxis (OMA) (aka pancake syndrome) is a disease in which a person gets symptoms after eating food contaminated with particular mites. The disease name comes from reports of people becoming ill after eating pancakes made from contaminated flour. Mites suspected as causing the condition are *Dermatophagoides pteronyssinus*, *Blomia tropicalis*, and *Suidasia pontifica* (*Suidasia medanensis*).^[1] The latter two are the most likely to have caused the harmful effects, and they live in tropical and subtropical environments.^[1] "The pancake syndrome (oral mite anaphylaxis) by ingestion and inhalation in a 52-YEAR-OLD woman in the northeastern United States". *Annals of Allergy, Asthma & Immunology*. Oral mite anaphylaxis (OMA), also known as pancake syndrome, is a disease in which a person gets symptoms after eating food contaminated with particular mites. The disease name comes from reports of people becoming ill after eating pancakes made from contaminated wheat or corn (maize) flour. Dust mites are microscopic insects that produce very powerful allergens. If dust mites get into the food, then anyone eating the food is eating the allergens and may have an allergic reaction against them. The...