

DERM D X

A 71-year-old woman presented with expanding necrolytic erythema on her tongue and perineum. Her history was remarkable for eczema, anemia, and painful glossitis during the previous year. She had experienced nausea and vomiting for 6 months and "striking" 20 kg weight loss during the preceding 18 months. She'd had a recent attack of deep vein thrombosis and transient ischemia. What's your diagnosis?



COURTESY DR. PAUL R. VANDERSTEEN

KEY BISCAYNE, FLA. — The 71-year-old woman's laboratory findings included a hemoglobin of 9.8 g/dL, an erythrocyte sedimentation rate of 94 mm/hr, and a fasting blood sugar level of 163 mg/dL. Her total serum protein was 4.2 g/dL (normal range, 6-8 g/dL), and her plasma glucagon was 405 pg/mL (normal range, 50-200 pg/mL).

Histology showed significant changes in upper skin strata, spongiosis, minor blister formations, and significant parakeratosis.

Abdominal CT scan and abdominal sonogram were negative. "I almost had to beg one of my surgeons to do a laparotomy," Paul R. Vandersteen, M.D., said at the annual meeting of the Noah Worcester Dermatological Society. Endoscopy showed a large encapsulated tumor, 5 by 4 by 3 cm, at the head of the pancreas. There were no metastases.

Her diagnosis was glucagonoma syndrome, a rare α -cell tumor of the pancreas; there are only about 400 reported cases. The syndrome typically arises in a person's fifth decade. "What is striking in the [two] patients I've seen ... is the significant pain, similar to a burn," said Dr. Vandersteen, a dermatologist at MeritCare Medical Group in Fargo, N.D.

Necrolytic migratory erythema (NME) is the hallmark sign and the overall incidence of NME is 57% and up to 77% at initial presentation. It typically appears periorally or perinasally, as well as on the lower abdomen, perineum, genitals, and lower extremities. Generally, the erythematous patch follows a 7- to 14-day cycle during which it blisters centrally, erodes and crusts over, and heals with hyperpigmentation.

Glucagonoma is also known as the 4D syndrome, for dermatitis, diabetes, depression, and deep venous thrombosis. In addition to NME and diabetes mellitus, the most common presenting symptoms are weight loss, cheilosis, stomatitis, diarrhea, weakness, and mental changes. Laboratory signs include hypoaaminoacidemia and hypoproteinemia, said Dr. Vandersteen, who is also a clinical professor of medicine at the University of North Dakota, Grand Forks.

The differential diagnosis includes acrodermatitis enteropathica, essential fatty acid deficiency, annular chronic lupus erythematosus, adverse drug reactions, contact dermatitis, and various vitamin deficiencies.

Imaging is helpful for diagnosis, particularly endoscopic ultrasonography and abdominal CT scans, "but remember, ours was negative," Dr. Vandersteen said. Other imaging modalities are less useful, such as angiography, transabdominal ultrasound, or somatostatin receptor scintigraphy.

Treatment was a partial pancreatectomy. Postoperatively, the patient developed another deep venous thrombosis. Her glucagon level decreased significantly to 126 pg/mL at 4 days. Her NME gradually resolved over the following month.

Seven years after initial presentation the patient had pancreatic surgery. Mean survival is 3-7 years, but some live up to 20 years. Another 7 years later, she was hospitalized and a large mass was detected. Dr. Vandersteen said, "They decided to do nothing; she went home and succumbed 2 days later."

—Damian McNamara

Identify, Eliminate Triggers To Reduce Hair Shedding

BY DIANA MAHONEY
New England Bureau

STOWE, VT. — Getting to the root of diffuse hair shedding requires uncovering and eliminating or treating the possible triggers, said Wilma F. Bergfeld, M.D.

"The key to optimal management of shedding, or telogen effluvium, is detective work," Dr. Bergfeld said at a dermatology conference sponsored by the University of Vermont. "You can't make most diagnoses from the door. You have to get up close and really look at the scalp."

Taking adequate time for a thorough history, determining whether the hair is coming out at the root or breaking off, acquiring hair and scalp samples for biopsy and culture in the absence of a well-defined trigger, and assessing hair density and loss patterns using hair-loss measurement scales can provide important clues, as can the results of laboratory tests, including complete blood count, comprehensive metabolic panel, mineral levels, and an androgen screen, said Dr. Bergfeld, head of the clinical research and dermatopathology sections at the Cleveland Clinic.

"Ask the patient to describe

how much hair they think they shed and when they notice it most," Dr. Bergfeld suggested. "And if a patient comes in with a bag of hair that she's lost, remember to ask her how long she's been collecting it."

Ask patients when the hair loss began, if it has been consistent, and whether there's a family history, Dr. Bergfeld continued. Also note all medications the patient takes, routine hair care habits, and whether the patient has had recent medical problems, surgeries, or childbirth.

Sometimes, the hair-loss trigger may be something as easy to identify as a deficiency of dietary zinc or vitamin A. In such cases, "when the nutrients are replaced, the shedding disappears," Dr. Bergfeld said. In other cases, however, multiple triggers may be responsible. The range of possibilities includes illness or infection, childbirth, drug reactions, metabolic events (such as iron-storage anemia), hormonal abnormalities, endocrine disorders, surgery, psychological stress, scalp disease, systemic disorders, or genetic conditions such as androgenic alopecia.

"Noticeable shedding usually begins within 2-4 weeks after the insult, or trigger, although

it can be months in some cases," Dr. Bergfeld said. "For example, significant shedding is common in young females post partum as a consequence of the trauma and hormonal changes associated with birthing, but breast-feeding can delay the shed by months."

Patients should be reminded that some shedding of hair is normal. In most people, up to 20% of the scalp hair is in telogen at a given time. Telogen effluvium is triggered when a physiologic or other event causes a greater number of hairs to enter telogen at one time. Acute telogen effluvium is usually self-limiting as the trigger event doesn't recur, Dr. Bergfeld said. When this type of nonscarring hair loss is persistent, however, either the trigger has not been eliminated or there is a more pervasive underlying medical condition that warrants further evaluation, she noted.

Management should include education about the hair growth cycle, treatment of underlying nutritional deficiencies, and possibly a prescription for minoxidil to promote hair growth, she said. "Often what is most important is reassurance that the shedding will not lead to baldness and that hair is being replaced." ■

Treatment of Lice Requires Specificity

BY DIANA MAHONEY
New England Bureau

STOWE, VT. — Location, location, location. Where lice live on the body and how they got there are important considerations for optimal diagnosis and therapy, according to Dirk M. Elston, M.D.

While much attention is given to the identification and treatment of head lice because they are hyperendemic in many areas of the world, body lice and pubic lice are unique entities with specific treatment requirements, Dr. Elston said at a dermatology conference sponsored by the University of Vermont.

Similar in appearance to head lice, body lice (*Pediculus humanus corporis*) live in clothes and bedding rather than head hair. Scratch marks, hives, and small raised red bumps on the shoulders, torso, or buttocks are possible signs of body lice infestation.

Unlike head lice, body lice can be vectors for blood-borne diseases such as typhus and trench fever. In the United States, body louse infestation mainly affects homeless populations, said Dr. Elston of Geisinger Medical Center, Danville, Pa.

Body lice infestations are treated by removing the clothing, laundering it in hot water, drying it on high heat, or pressing it with a hot iron. However, these tactics are often not feasible in outbreaks in other parts of the world, Dr. Elston said. In some settings, treating clothing with DDT, permethrin, or fumigants is useful. In addition, single-dose ivermectin has shown promise for mass treatment, and pediculicides may be effective against body lice.

Pubic lice (*Phthirus pubis*) are distinct in appearance from head and body lice; they have short crablike bodies. While they are most frequently found in the pubic region of the in-

festated person, where they can cause intense itching and redness, they may also be found in other areas, such as in facial hair or eyelashes. In fact, Dr. Elston said, "I am amazed at how often it is misdiagnosed. When you see eyelash nits, you should be looking south, not north," he stressed.

Pubic lice infestation occurs mainly through sexual contact, but may also be acquired by sharing a bed. Children usually contract pubic lice from an adult, which should be investigated, he said.

However, pubic lice help to solve crimes "There is enough blood in a single louse to identify a rapist's DNA by [polymerase chain reaction]," he said. Because the pubic louse egg "is relatively impermeable, the best way to get to it is through the [host's] blood," Dr. Elston said. Toward that end, oral sulfa drugs as well as ivermectin have been used successfully. ■