Umbilical endometriosis elucidates cause of recurrent pneumothorax

To the Editor: Endometriosis is characterized by ectopic endometrial tissue. Cutaneous and thoracic variants are rare and challenging. We report a case of both cutaneous and thoracic endometriosis in the absence of surgery or prior endometriosis. Cutaneous findings secured the diagnosis of catamenial pneumothorax.

A 44-year-old woman with no medical or surgical history developed dyspnea. Chest radiograph showed right-sided tension pneumothorax consistent with spontaneous pneumothorax that resolved after chest tube placement. Twelve months later, a bleeding umbilical papule developed. Pathologic study revealed endometrial tissue (Fig 1). She had no history of endometriosis, menstrual, or fertility issues.

Another right-sided pneumothorax occurred 6 months later during menses. The cutaneous findings of endometriosis established a diagnosis of catamenial pneumothorax. Continuous oral contraceptives were initiated but a third right-sided pneumothorax occurred. Thoracoscopy identified diaphragmatic endometrial tissue (Fig 2). Leuprolide acetate was initiated. A fourth pneumothorax occurred 6 months later. The patient underwent bilateral oophorectomy with no recurrence at 44 months.

Endometriosis, defined by endometrial tissue outside the uterus, affects 5% to 10% of reproductive aged females. Endometriosis, an estrogen-dependent inflammatory process, typically presents with dysmenorrhea, dyspareunia, and subfertility. Ectopic endometrial deposits usually occur on pelvic surfaces and are rare outside the pelvis. Cutaneous endometriosis accounts for 1% of endometriosis and is most prevalent in surgical scars from cesarean section, hysterectomy, and laparoscopic procedures due to physical transplantation of endometrial cells during surgery. Umbilical endometriosis is rare but can occur in women without a history of endometriosis or surgery, perhaps from lymphatics transporting endometrial tissue.

Catamenial pneumothorax occurs within 72 hours of menses due to ectopic endometrial tissue eroding into the pleura. Rare, it affects women in their third and fourth decades. Thoracic endometriosis typically presents with pneumothorax, but hemothorax, hemoptysis, and lung nodules are also reported. In 95% of cases, pneumothorax is right-sided. The mechanism of catamenial pneumothorax is unclear. The metastatic theory involves retrograde implantation of endometrial tissue from the pelvis to the lungs via lymphatics or congenital fenestrations in the diaphragm. Once implanted, endometrial tissue bleeds with menses, causing a pneumothorax. The right hemidiaphragm contracting against the solid, fixed liver on the right side may allow for ectopic endometrial migration and implantation. The anatomic model postulates the dissolution of the cervical mucus plug during menses, allowing air to travel from the vagina to the uterus, through the fallopian tubes, and into the peritoneum. If congenital fenestrations of the diaphragm are present, intraperitoneal air ascends into the pleural cavity, causing pneumothorax.

Estrogen drives endometriosis. Treatment focuses on estrogen inhibition via hormonal (oral contraceptives, GnRH analogues, danazol, and aromatase inhibitors) or surgical treatments. Hormonal treatment alone is often insufficient to completely suppress estrogen. Due to estrogen production via peripheral conversion of androgen, surgical treatment alone is not always definitive. Recurrences are common and require combination treatment.

The diagnosis of catamenial pneumothorax was secured after correlating the rare diagnosis of
umbilical endometriosis with pneumothorax associated with menses, emphasizing the importance of skin findings in the diagnosis of systemic disease.

Christen Mowad, MD,a Cassondra Andreychik, BA,b and Timothy Murphy, MDa

Geisinger Medical Center,a Danville, and Commonwealth Medical School,b Scranton, Pennsylvania

Funding sources: None.

Conflicts of interest: None declared.

Correspondence to: Christen Mowad, MD, 115 Woodbine Lane, Danville, PA 17822
E-mail: cmowad@geisinger.edu

REFERENCES

http://dx.doi.org/10.1016/j.jaad.2014.02.024

Generalized granuloma annulare after varicella infection: Wolf isotopic response?

To the Editor: The etiopathogenesis of granuloma annulare (GA) is unknown, but a delayed-type hypersensitivity reaction to an unknown antigen has been hypothesized. Several inciting factors have been reported, including trauma, ultraviolet radiation, and viral infections.1 Among the latter, GA developing at the site of previous herpes zoster (HZ) has frequently been reported in a Wolf isotopic response (WIR).1,5 However, GA developing after varicella infection, to the best of our knowledge, has not been reported except in a single report of localized GA developing 6 weeks after uncomplicated varicella in a 4-year-old child.1 Here we report generalized GA developing in previously healed lesions of varicella 2 months after infection clearance.

A 5-year-old girl presented with a 2-month history of an asymptomatic skin eruption on her trunk and extremities. This eruption was preceded by an uncomplicated varicella infection that occurred 2 months earlier and resolved spontaneously. The parents indicated that the new lesions may have arisen at the same sites of previous varicella lesions. The patient was otherwise healthy. Skin examination revealed multiple firm skin-colored papules on trunk and extremities, some of which were grouped in a “herpetiform” arrangement (Fig 1). Punch biopsy revealed a palisaded granulomatous dermatitis with focal collagen degeneration and mucin deposition (Fig 2). These features were diagnostic of generalized GA, which probably developed at the sites of previous varicella lesions. Systemic workup was negative. The parents elected not to treat, and the lesions resolved spontaneously in 11 months.

WIR is characterized by the development of a distinctive cutaneous disease at the site of another unrelated and healed disease after a variable period ranging from days to years.1,5 All described cases have occurred after herpes simplex or zoster

Fig 1. Granuloma annulare. Multiple firm skin-colored papules in a “herpetiform pattern” on the abdomen (A) and right foot (B). Note that the pink to red background is because of the use of aqueous solution of eosin 2% by the patient prior to presentation.