Spontaneous resolution of recalcitrant generalized prurigo nodularis after resection of an ampulla of Vater tumor

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Case report

A 63-year-old retired male teacher presented with generalized pruritic papules for 4 years. The prurigo was recalcitrant to treatment and he was admitted for further evaluation and treatment. His medical history was significant for thyroidecmy for hyperthyroidism 23 years ago with normal thyroid function after surgery. He had hyperuricemia with gouty arthritis but received no treatment. He also complained of epigastric discomfort for several decades. He denied any history of atopy or drug allergies, and was not on medications before the onset of skin lesions. A blood examination showed peripheral eosinophilia (1040/mm³). The serum IgE level was within the normal range. There was no hyperbilirubinemia. A stool examination showed no evidence of parasite infection. After discharge, he received treatment at other clinics in his hometown for 7 years.

In 2001, he returned to our clinic because of worsening of the skin lesions. An examination revealed many lichenified and juicy papules disseminated on his trunk and extremities (Figures 1A–1C). A biopsy taken from a lichenoid papule showed compact hyperkeratosis, hypergranulosis, epidermal hyperplasia and fibrotic papillary dermis with dilated blood vessels, consistent with lichen simplex chronicus or prurigo histopathologically. The diagnosis of generalized prurigo nodularis was made. The prurigo showed a poor response to treatments, including topical high potency steroids, oral antihistamines and narrow band UVB phototherapy. New lesions continued to erupt despite 18 months of intensive treatments. Follow-up eosinophil counts were within normal limits. The patient finally gave up and declined further treatments. One year after giving up treatments for prurigo, he underwent an endoscopic retrograde cholangiopancreatography to evaluate his epigastric discomfort. It revealed a tumor at the ampulla of Vater, which was shown to be an adenoma pathologically (Figure 2A). Magnetic resonance imaging (Figure 2B) showed a tumor in the ampulla of Vater region. The patient had no signs of cholestasis. His serum levels of bilirubin, amylase and lipase were within the normal range. The tumor was excised and the final pathological diagnosis was villotubular adenoma with moderate dysplasia. Surprisingly, the prurigo showed spontaneous improvement and disappeared completely 2 years after an operation (Figures 3A–3C). He remained free of prurigo at 4 years follow-up.

Discussion

We report a case with chronic, recalcitrant widespread prurigo nodularis that resolved spontaneously after resection of an ampulla of Vater tumor. Adenomas of the ampulla of Vater are rare neoplasms with an incidence of 0.04–0.12% in a postmortem series.¹ The tumor can obstruct normal drainage of pancreatic or bile secretions and cause jaundice, cholangitis and pancreatitis. More importantly, these adenomas have the potential for malignant changes.¹ Patients with adenoma of the ampulla of Vater may complain of non-specific upper abdominal discomfort. Pruritus usually occurs in patients with obstructive jaundice and is relieved by removal of the tumor.²

Prurigo nodularis is characterized by a papulonodular eruption with intense pruritus. Although the acute form is often induced by insect stings, most of the subacute and chronic forms appear to be idiopathic.³ Chronic lesions are usually difficult to treat and cause frustration to both patients and physicians. Prurigo nodularis is sometimes associated with atopy, pregnancy, systemic diseases, malabsorption, or...
Resolved prurigo after ampulla Vater tumor excision

Prurigo nodularis has been reported as an initial presentation of internal malignancies, including hepatocellular carcinoma, metastatic transitional cell carcinoma, Hodgkin's disease, adult T-cell leukaemia/lymphoma and gastric cancer. Prurigo may represent a paraneoplastic sign of hepatocellular carcinoma in which the skin lesions resolve after tumor removal. To the best of our knowledge, generalized prurigo nodularis associated with adenoma of the ampulla of Vater without obstructive jaundice, as seen in our patient, has never been reported.

The prurigo lesions in the present case were preceded by chronic upper abdominal discomfort for several decades.

Figure 1  (A–C) A 63-year-old male had progressively worsening generalized prurigo nodularis.

Figure 2  (A) An endoscopic retrograde cholangiopancreatography for evaluation of patient's chronic epigastric discomfort reveals a tumor at the ampulla of Vater (arrow). (B) Magnetic resonance imaging (T1 phase) shows a tumor in the ampulla Vater region (arrow).

Figure 3  (A–C) The skin lesions were chronic and recalcitrant to various treatments, but resolved spontaneously in 2 years after resection of a villotubular adenoma with moderate dysplasia of the ampulla of Vater.
The finding of moderate dysplasia in a villotubular adenoma suggests that the adenoma might have been longstanding and had undergone malignant transformation, and this might be associated with the development of prurigo lesions chronologically.

The mechanism of prurigo in our patient remains elusive. Neuropeptides, including neuronal growth factors, calcitonin gene-related peptide and substance P, have been proposed as the pathogenesis of prurigo nodularis. Further study is required to clarify whether the tumor in the present case secreted pruritogenic neuropeptides. Nevertheless, the spontaneous resolution of the recalcitrant prurigo after tumor excision suggests that there may have been a causal relationship between the recalcitrant prurigo nodularis and the ampulla Vater adenoma.

In summary, we report a patient with refractory generalized prurigo nodularis, which resolved spontaneously after excision of a dysplastic ampulla of Vater tumor. We recommend a thorough survey of internal malignancy for patients with recalcitrant prurigo as this may be a sign of internal malignancy on rare occasions, and an ampulla of Vater tumor should not be overlooked in patients with chronic epigastric discomfort.

References