

## Branch Duct-type Intraductal Papillary Mucinous Neoplasm Presenting as Paraneoplastic Small Plaque Para-psoriasis

### Abstract

To present and discuss a novel association between branch duct-type intraductal papillary mucinous neoplasm and paraneoplastic parapsoriasis. We present the case of a middle-aged male presenting with skin lesions that were suggestive of parapsoriasis, resistant to treatment, and in whom a diagnosis of branch-type intraductal papillary mucinous neoplasm of the pancreas was eventually made. A curative Whipple's surgery led to complete resolution of the skin lesions within 3 weeks. Paraneoplastic parapsoriasis in association with intraductal papillary mucinous pancreatic neoplasm has never been reported before.

**Keywords:** *Intraductal papillary mucinous neoplasia, obstructive jaundice, pancreatic tumor, paraneoplastic syndrome, parapsoriasis, pruritus psoriasis*

### Introduction

Parapsoriasis (PP) are a heterogeneous group of rare dermatoses presenting, with erythematous and scaly patches of variable size and a chronic course, that are difficult to treat and mostly seen in the elderly. Small-plaque parapsoriasis (SPP) and large-plaque parapsoriasis (LPP) are the two broad categories within this group. Lesions are usually asymptomatic or associated with mild pruritus and are predominantly located on the trunk. SPP is generally considered a chronic benign condition whereas LPP is regarded as a premalignant condition with a high risk of progression to mycosis fungoides, a type of cutaneous T cell lymphoma.<sup>[1]</sup> Intraductal papillary mucinous neoplasia (IPMN) is considered precursors of pancreatic cancers with a slow growth rate that enables early diagnosis and curative surgical treatment. The World Health Organization classifies IPMN according to their malignant transformation into IPMN with low or intermediate dysplasia, high-grade dysplasia, or invasive cancer. Approximately 20% of the patients are asymptomatic whereas symptomatic patients present with nonspecific symptoms such as nausea, abdominal or back pain, and weight loss, and sometimes with recurrent pancreatitis. Patients with IPMN are most notably elderly females. Paraneoplastic

syndromes are rarely described with IPMN.<sup>[2,3]</sup> Here, we present a middle-aged male diagnosed to have treatment-resistant SPP in whom an eventual diagnosis of IPMN was made. Curative Whipple's surgery led to complete resolution of skin lesions within 3 weeks. This rare association has never been described before.

### Case Report

A 42-year-old male without chronic comorbid illnesses and a teetotaler presented with fluctuating jaundice for 2 weeks. His past history was significant for multiple crops of pruritic flat red and scaly skin lesions affecting mostly the trunk and upper thighs that were waxing and waning for 3 months. He was told to have psoriasis, even though a skin biopsy was not done, and was given short courses of steroids and topical retinoids without relief. He denied gastrointestinal bleeds, altered sensorium, abdominal distension, abdominal pain, or fevers. A significant weight loss of >10% had occurred during the last month with loss of well-being and appetite. Clinical examination revealed mild icterus without pallor, lymphadenopathy, or peripheral edema. Abdominal examination was essentially normal. General skin examination revealed multiple erythematous, round-to-oval,

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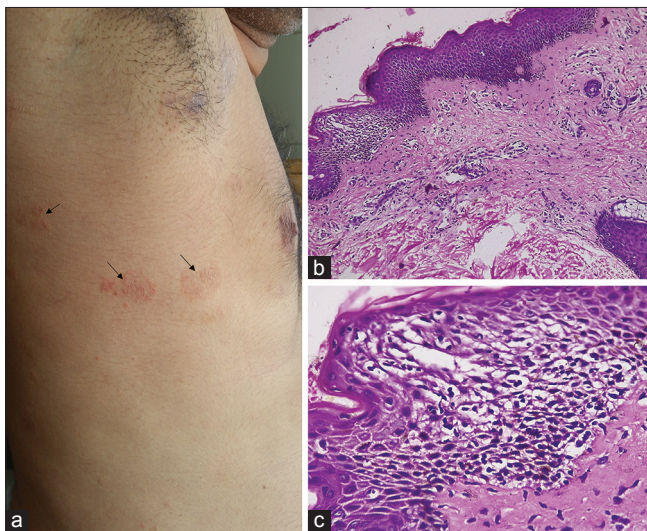
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scaly lesions with distinct borders scattered mostly over the trunk [Figure 1a, arrow] and upper chest that were provisionally thought to be SPP, cutaneous T cell lymphoma, or tinea incognito. Hemogram was normal, liver function tests showed total bilirubin of 9.4 mg/dL with direct fraction 5.8 mg/dL, alanine transaminase 76 IU/L, aspartate transaminase 98 IU/L, alkaline phosphatase 88 IU/L, and gamma glutamyl trans peptidase 228 IU/L. Tumor markers including CA19-9, carcinoembryonic antigen, alfa fetoprotein, and prostate specific antigen were within normal limits. The dermatologist ordered a skin biopsy which revealed, focal keratosis, parakeratosis, and acanthosis with upper papillary dermal fibrosis with perivascular lymphocytic infiltrates [Figure 1b], along with few areas showing spongiotic changes and neutrophilic exocytosis, consistent with PP with focal spongiotic dermatopathy [Figure 1c]. A contrast-enhanced computed tomography imaging of the abdomen revealed bilateral intrahepatic biliary radicle dilatation [Figure 2a, black arrow], cystic dilatation of the branches of pancreatic duct near the head and uncinate process [Figure 2b, white arrow], with a soft tissue mass at the distal end of the common bile duct (CBD) and extending into the distal pancreatic duct causing dilatation of both ducts, which was confirmed by a magnetic resonance cholangiopancreatogram (MRCP) [Figure 2c, white circle], as well as a normal appearing liver and absence of lymphadenopathy or ascites. A subsequent endoscopic ultrasound and fine-needle aspiration cytology from the mass lesion near the CBD revealed only benign cholangiocytes. After discussion with the hepatobiliary surgical team, Whipple's resection was performed and the resected specimens were sent for histopathological



**Figure 1:** (a) Multiple erythematous, round to oval, scaly lesions with distinct borders scattered mostly over the trunk (arrows); (b) Skin biopsy showing focal keratosis, parakeratosis and acanthosis with upper papillary dermal fibrosis with perivascular lymphocytic infiltrates (Hematoxylin and Eosin stain,  $\times 10$ ); (c) Spongiotic changes and neutrophilic exocytosis, consistent with parapsoriasis with focal spongiotic dermatopathy (Hematoxylin and Eosin stain  $\times 40$ )

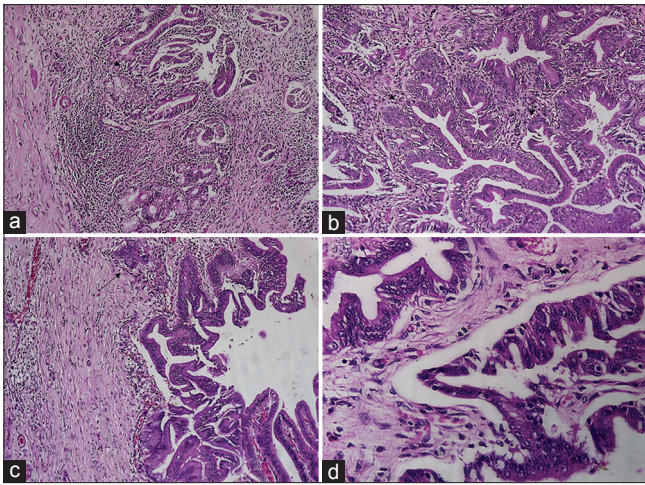
evaluation. Sections from the pancreas showed dilated main pancreatic duct showing intermediate-grade IPMN arranged in papillae, lined by pancreaticobiliary type epithelium focally showing mucinous appearance with lining cells showing focal crowding, and stratification [Figure 3a and b, arrows]. Occasional foci showed areas of microinvasion [Figure 3c, arrow] and surrounding stroma showing mild inflammatory infiltrates [Figure 3d]. A final diagnosis of paraneoplastic SPP secondary to IPMN was made. One month after the surgery on follow-up, the patient is doing well, with complete resolution of the skin lesions [Figure 4] and increased appetite and weight gain.

## Discussion

Paraneoplastic dermatoses are the second most common paraneoplastic syndrome after endocrine syndromes. Curth *et al.*, in his studies of acanthosis nigricans maligna, provided criteria to assess the causal relationship between dermatopathy and potential underlying malignancy (Curth's Criteria). Various dermatological manifestations are described in the literature that have shown strong associations with internal malignancies [Table 1].<sup>[4-7]</sup> IPMN are potentially malignant epithelial neoplasms consisting of cystic dilatations of the main pancreatic duct (MD type), side branches (BD type), or a combination of both. The BD type is mostly seen in the head and uncinate process is more localized and mass-like, but maybe multifocal with macro or microcystic in appearance.<sup>[5,7]</sup> IPMNs are classified into low-grade, intermediate-grade, and high-grade dysplasia based on cytological atypia. Based on histological features and mucin immune-phenotype, IPMN are classified into gastric, intestinal, pancreatobiliary, and oncocytic types. Fukuoka guidelines state that pancreatic cysts more than 5 mm in diameter and communicating with the main pancreatic duct, in the absence of pancreatitis



**Figure 2:** (a) Contrast CT imaging of abdomen showing bilateral intrahepatic biliary radicle dilatation (black arrow); (b) Soft tissue mass at the distal end of CBD and extending into the distal pancreatic duct (white arrow); (c) MRCP showing soft tissue mass at the lower end of CBD and distal pancreatic duct showing duct cut off signs (white circle)



**Figure 3:** (a) Pancreatic histology showing dilated main pancreatic duct with intermediate grade intraductal papillary mucinous neoplasm (IPMN) arranged in papillae, lined by pancreaticobiliary type epithelium (arrows, Hematoxylin and Eosin stain  $\times 20$ ); (b) Focal mucinous appearance with lining cells showing focal crowding and stratification (arrows, Hematoxylin and Eosin stain,  $\times 40$ ); (c) Occasional foci showing areas of microinvasion (arrow, Hematoxylin and Eosin stain,  $\times 40$ ) and (d) Surrounding stroma showing mild inflammatory infiltrates (Hematoxylin and Eosin stain,  $\times 40$ )

and/or diffuse dilation of the main pancreatic duct more than 5 mm diameter without other causes of obstruction are sufficient radiological criteria for IPMN, which were noted in our patient and proven thereafter with histopathology.<sup>[8,9]</sup> IPMN has been rarely shown to present with paraneoplastic features. Grigorescu *et al.* reported the association of IPMN with paraneoplastic rheumatoid arthritis.<sup>[10]</sup> Roch *et al.* described 220 extrapancreatic malignancies in 185 patients of IPMN compared with expected 5% in the general population. One hundred ten synchronous autoimmune diseases were found in 96 patients (11%). Systemic lupus erythematosus, rheumatoid arthritis, and inflammatory bowel disease showed statistically significant observed/expected ratios ( $P < 0.0001$ , 01, and  $< 0.0001$ , respectively) without impact of immunosuppressive treatment on the IPMN subtype and malignancy rate. They concluded that IPMNs are associated with surprisingly high rates of autoimmune diseases suggesting that IPMN might be one manifestation of a more systemic disease.<sup>[11]</sup>

Our case demonstrates two novel associations. One, the description of a paraneoplastic dermatopathy in association with IPMN, following Curth's criteria and one that improved after curative resection of the primary and two, the first description of SPP as a paraneoplasm in association with internal malignancy, especially pancreatobiliary cancers.

### Conclusion

IPMNs are rare tumors that can present with rare autoimmune systemic symptoms and with paraneoplasms, especially dermatopathies. Early identification and evaluation for pancreatobiliary malignancies are warranted in patients presenting with dermatological symptoms that are atypical and refractory to conventional treatment.



**Figure 4:** Post Whipples surgery, the skin lesions resolved completely

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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**Table 1: Curth's criteria for paraneoplastic dermatoses and commonly described paraneoplastic dermatosis and the associated internal malignancies**

Curth's criteria	Paraneoplastic dermatoses	
	Dermopathy	Associated malignancy
Both conditions began simultaneously (neoplasia and paraneoplasia)	Acanthosis nigricans maligna	Gastric neoplasia
Development of a parallel course *	Acquired pachydermatoglyphia	Gastric and pulmonary carcinoma
The skin lesion is not associated with a genetic syndrome	Erythema gyratum repens	Pulmonary, oesophageal and breast
There is a specific type of neoplasia that occurs with paraneoplasia	Bazex acrokeratosis	Aero digestive tract
The dermatosis is rare in the general population	Acquired hypertrichosis lanuginosa	Colorectal, pulmonary and breast
There is a high frequency of association between both conditions	Necrolytic migratory erythema	Glucagonoma
* Treatment of the neoplasia results in regression of the skin lesion; recurrence of the neoplasia implies recurrence of the skin lesion.	Leser-Trelat sign	Gastric and colorectal
	Paraneoplastic pemphigus	Non-Hodgkin lymphoma, chronic lymphocytic leukaemia, Castleman's disease, thymoma
	Pityriasis rotunda	Hepatocellular carcinoma, gastric and oesophageal, prostate neoplasms, chronic lymphocytic leukaemia, multiple myeloma
	Dermatomyositis	Ovarian carcinoma, bronchogenic adenocarcinoma
	Palmoplantar keratoderma	Oesophageal carcinoma
	Pyoderma gangrenosum	Myelodysplastic syndrome, myeloma, leukemia
	Sweet syndrome	Acute myelogenous leukemia, myelodysplastic syndrome

\* Major criteria

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