



Case report

Intraabdominal dissemination of porocarcinoma; A case report

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ARTICLE INFO

Keywords:

Porocarcinoma
Intraperitoneal invasion
Peritoneal seedlings

ABSTRACT

Introduction: Porocarcinoma is a rare malignancy of dermal sweat glands commonly diagnosed in the seventh decade of life. It frequently evolves from a de novo benign poroma. These tumors present as a mass/nodule, ulcer, papule, or wart. Difficult to differentiate from other cutaneous lesions. Intraperitoneal invasion is scantily reported in the literature.

Case presentation: The authors present a case of a fifty-year-old female patient with a rare cancer of the dermal sweat glands in an unusual location and infiltration into the abdominal cavity, leading to intraperitoneal seedlings.

Discussion: Tumors of the sweat gland are rare and difficult to diagnose, often misdiagnosed as granuloma, squamous cell tumors, or warts. Surgical excision and Mohs micrographic surgery are mainstay treatment modalities in the early stages. Our patient was managed elsewhere with a diagnosis of granuloma. She was referred with a recurrence of the abdominal lesion. An appropriate diagnosis of porocarcinoma was made while she had an extensive intraperitoneal invasion and seedlings. We postulate that the previous abdominal incision had disseminated porocarcinoma cells into the abdominal cavity, causing extensive intraperitoneal dissemination.

Conclusion: Because it is rare and difficult to diagnose, there is a considerable knowledge gap in the early accurate diagnosis and appropriate management of porocarcinoma. This causes a delay in establishing a diagnosis and profoundly impacts treatment outcomes.

1. Introduction

Porocarcinoma are rare and heterogenous adnexal tumors of the dermis [1]. They originate from epithelial cells of eccrine glands, intra epidermal and dermal eccrine ducts [2,3]. Initially described as benign adenomatous poroma and confused with basal cell carcinoma until 1969, when it was termed porocarcinoma [4]. The first case of porocarcinoma was reported in 1963 by Pinkus and Mehregan; described as epidemotrophic eccrine carcinoma due to histological epidemotrophism and pagetoid nature [5]. There is an even gender distribution or a slight male predominance [6,7]. The disease commonly affects whites and is often diagnosed in the seventh decade of life [6,8,9]. High radiation dose, prolonged sunlight exposure, immunosuppression, and other pre-existing dermatological pathology are the main risk factors for porocarcinoma [6]. Although not known, the pathogenesis of porocarcinoma is linked to p53, observed in 83 % of eccrine and 73 % of porocarcinoma

[10]. Molecular analyses have revealed that the fusion of YAP1 and WWTR1 genes profoundly impacts the tumorigenic transformation of poromas [11]. Histologically porocarcinoma consists of matured ductal formation and matured intracytoplasmic lumina; other microscopic findings include comedo necrosis, diffuse necrosis, and squamous differentiation [12]. Common sites affected are the lower limbs, scalp, face, neck, and trunk [6,7,9]. This article is being reported following the SCARE criteria 2020 [13].

2. Case report

A fifty-year-old female was referred to us with the complaint of abdominal pain and distension for one year. She had a history of two abdominal surgeries two years earlier, a hysterectomy and wide local excision of the suprapubic polypoid mass. A recurring exophytic growth on her previous midline incision was painless and progressing in size.

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<https://doi.org/10.1016/j.ijscr.2022.107529>

Received 14 July 2022; Received in revised form 15 August 2022; Accepted 18 August 2022

Available online 22 August 2022

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Fig. 1. Shiny and distended abdomen with a granulomatous polypoid mass in the suprapubic region.



Fig. 2. Axial and coronal CT images show the cystic lesion's invasion deep in the abdominal cavity.

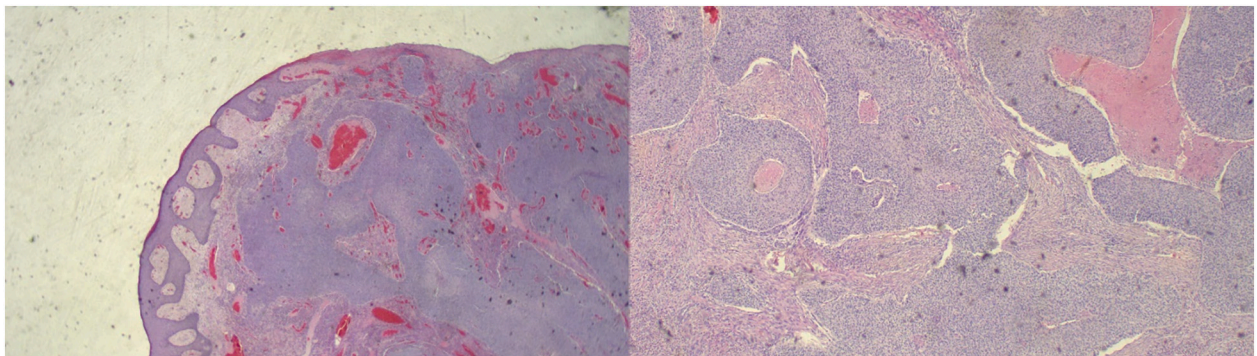


Fig. 3. Histological sections showing porocarcinoma.



Fig. 4. Cystic fluid drainage for cytology.

The tumor was easily bleeding, soaking her cloth. Clinically she had shiny abdominal skin with an indefinite cystic lesion and an exophytic mass in the suprapubic area of 4 by 3 cm (Fig. 1). Thoracoabdominal CT revealed a complex cystic lesion extending from the subcutaneous tissues into the peritoneal cavity (Fig. 2). A tissue biopsy of the lesion revealed an invasive porocarcinoma. Histologically, the tumor emerged from skin adnexa. Individual cells were epithelial, atypical, and cohesive with sweat gland differentiation (Fig. 3).

Intra-operative, a complex cystic lesion was found extending below the epidermis into the peritoneal cavity. The components of the cyst had an odorless amber-colored fluid (Fig. 4). She had mesenteric lymphadenopathy of the small bowel and colon with several peritoneal seedlings (Fig. 5). An excisional biopsy of the granulomatous lesion revealed an invasive porocarcinoma.

Immediate postoperative was uneventful; discharged a few days later with acceptable progress. She was kept under conservative therapy and best supportive care. Three months later, she succumbed.

3. Discussion

Porocarcinoma is a rare pathology with an age-adjusted incidence of 0.06 in males and 0.04 in females per 100,000 person-years [9]. There is a substantial increase in incidence beyond 70 years [9]. The tumor commonly presents as a mass/nodule (71.2 %); other features include ulcer, plaque, wart, papule, and wart [7]. Porocarcinoma is difficult to diagnose, often misdiagnosed as granuloma, squamous cell carcinoma, and viral warts [2,14]. Distant and local regional spread is found in less than one-third of the cases [7]. The most common organs of local-regional and foreign involvement are lymph nodes (57.7 %), lung (12.8 %), liver (9 %), brain (9 %), and cutaneous (5.8 %). Others include breast, stomach, and disseminated disease [7]. Surgery is the

cornerstone in managing porocarcinoma; options include wide local excision and Mohs micrographic surgery [8]. Chemotherapy and radiotherapy are used in advanced stages [7]. Primary excision in the early stages results in 70–80 % curability; however, a local-regional recurrence of 20 % and a distant spread of 20 % have been reported after a curative excision [15]. The survival rate ranges from 5 to 24 months in metastatic disease [16]. The diversity in establishing diagnosis had a profound impact on disease progression and a delay in curative treatment. Our patient had an exophytic polypoid lesion along the previous midline incision, which was managed as a granuloma. Due to tumor recurrence and its progressive growth, the diagnosis of porocarcinoma was made in advanced stages. She had abdominal wall invasion, seedlings to the peritoneal layer, and multiple mesenteric lymphadenopathies to the root of the mesentery. We postulate that a hysterectomy two years earlier had led to the dislocation of poroma/porocarcinoma cells through the abdominal wall into the peritoneal cavity, causing this unusual direction of infiltration with extensive intraperitoneal spreading. She was subjected to conservative therapy and succumbed three months later. Surgical excision is the preferred treatment modality but carries a high tendency for recurrence [7,16]. A delay in establishing the diagnosis and different tumor biology might suggest an otherwise aggressive tumor. The initial case series reported porocarcinoma as an aggressive skin cancer with mortality rates of 80 % in disseminated disease [17]. Current literature suggests an otherwise better prognosis. A five-year survival rate of 74.5 % for head and neck porocarcinoma has been reported in the U.S [6]. In Finland, a ten-year retrospective analysis revealed a single porocarcinoma-related death among 69 cases reviewed [9].

4. Conclusion

Our patient had an extensive intraabdominal invasion of porocarcinoma. The diagnosis was established late with significant morbidity and later fatality. Porocarcinoma is difficult to diagnose, with a high rate of misdiagnosis causing a delay in curative treatment. Due to the scarcity of data, there is contention in treatment modalities. There is a considerable gap in the learning curve for accurate diagnosis and management of this rare pathology. A high index of suspicion and appropriate pathological diagnosis is advocated for improved treatment outcomes.

Funding

No financial support for this work.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Registration of research studies

Not applicable.

Guarantor

All authors in the article accept full responsibility for the work, have access to the patient's information, and decision to publish.

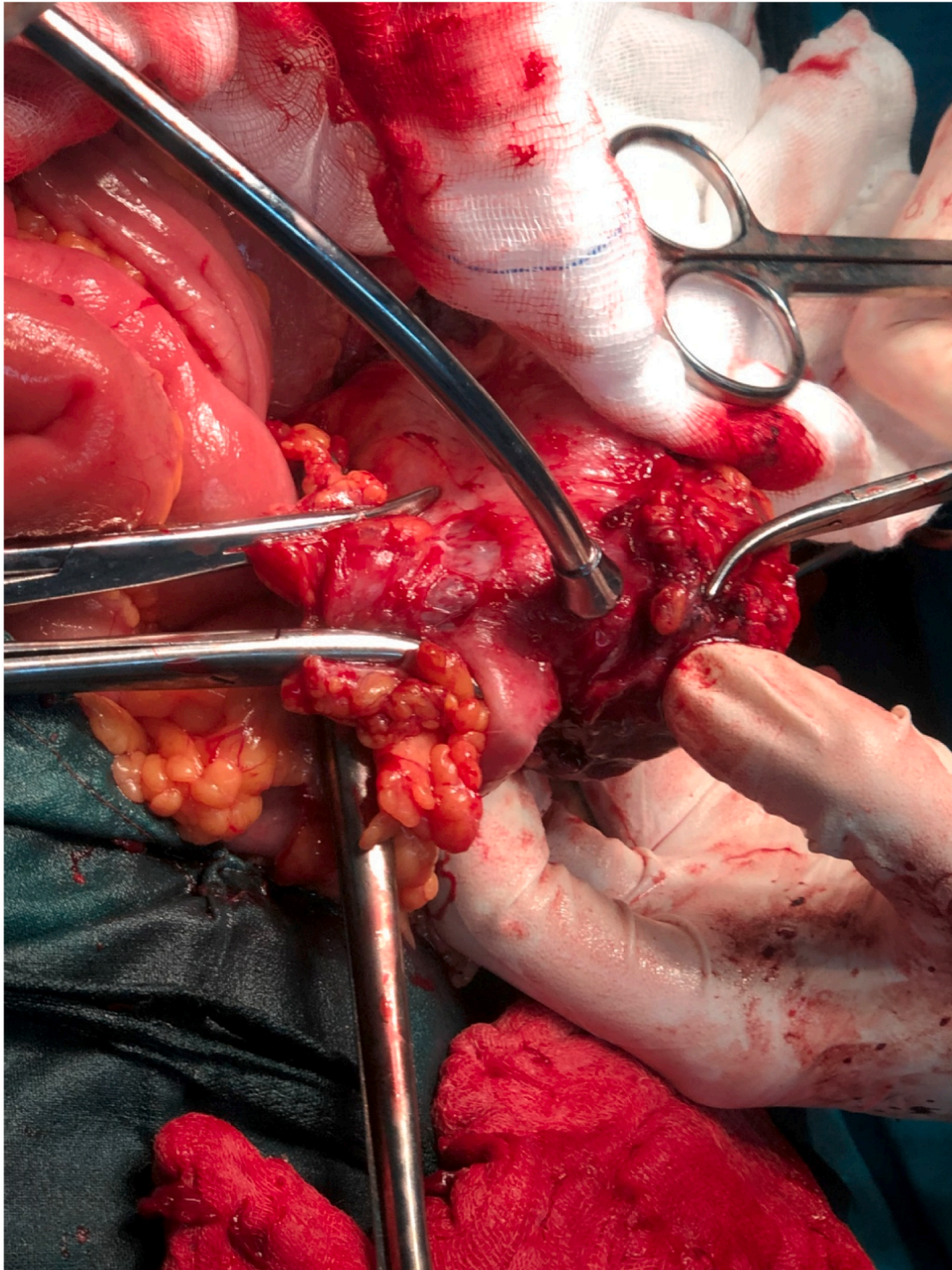


Fig. 5. A lymph node at the transverse mesocolon.

Credit authorship contribution statement

Dr. Kennedy Misso: Perioperative care, drafted and approved the final article.

Dr. Mathayo Shadrack: Perioperative care participated in the drafting and approval of the article.

Dr. Venant Ntakarutimana: Assisted the surgery, drafted and approved the final article.

Dr. Gilbert Nkya: Pathological diagnosis, drafted and approved the final article.

Dr. Murad Tarmohamed: Assisted in the procedure, drafted and approved the final article.

Dr. Kondo Chilonga: Chief surgeon, drafted and approved the final article.

Declaration of competing interest

The authors declare no conflict of interest.

Acknowledgment

The authors express their sincere gratitude to all surgical and pathology department members. To the patient for allowing us to proceed with academic publications.

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