PAMJ Clinical Medicine
Pan African Medical Journals

Case report

Mycosis fungoides with CD30 large cell transformation accompanied by hidden adenocarcinoma of the colon: a case report

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Received: 10 Dec 2019 - Accepted: 01 Mar 2020 - Published: 13 Mar 2020

Domain: Dermatology

Keywords: Mycosis fungoides CD30+, colon adenocarcinoma, cutaneous T-cell lymphoma

Abstract

Mycosis fungoides is the most common subtype of primary cutaneous T-cell lymphoma, its evolution is indolent, except in the case of transformation into large T-cell lymphoma. A 63-year-old patient, he has a stage IB mycosis fungoid for 4 years treated by topical corticosteroids associated with methotrexate. Then he had progressed to stage IIB, treated by photochemotherapy (PUVA) and acitretin with complete remission and then relapsed, with the appearance of tumors, and adenopathy, the extension assessment had objectified a transformed CD30 mycosis fungoid, with a lymph node localization of T lymphoma, the patient is classified as stage IIIB. Thoracic-abdomino-pelvic CT had shown a thickening of the sigmoid colon wall, completed by colonoscopy, whose colon biopsy had objectified a colonic adenocarcinoma. The association of colonic adenocarcinoma and a mycosis fungoides is rare, a pathophysiology is poorly understood, and suggests a probably genetic predisposition.

Case report | Volume 2, Article 104, 13 Mar 2020 | 10.11604/pamj-cm.2020.2.104.21255

Available online at: https://www.clinical-medicine.panafrican-med-journal.com/content/article/2/104/full

Introduction

Cutaneous T-cell lymphoma is a malignant non-Hodgkin T-cell lymphoma, with mycosis fungoides (MF) and Sezary syndrome being the most common subtypes. In patients with MF, transformation to large cell histology is associated with worse prognosis [1]. The association of MF with colon adenocarcinoma is a challenge because it is rare, most often we think of a digestive metastasis of MF. We present the case of a man with MF with CD30 large cell transformation who also had a hidden adenocarcinoma in the colon.

Patient and observation

A 63-year-old man, with a 4-year history of mycosis fungoides (MF). He first presented with multiple erythematous and poikilodermas patches of various sizes on the extremities and trunk. The affected body surface area was calculated about 60%. Skin biopsy showed a dense cellular infiltration of atypical lymphocytes with large hyperchromatic and pleomorphic nuclei throughout the dermis-compatible with MF. Laboratory evaluation revealed normal complete blood count and lactate dehydrogenase level. A computed tomographic scan of his chest and whole abdomen showed no significant abnormalities. MF stage IB was diagnosed. He was treated with corticosteroid associated with methotrexate (25mg/week), and achieved partial response. One year after treatment, he noticed a new tumor, making 4*3 cm in size, in front of the right popliteal fossa, with a bilateral centimetric inguinal adenopathy (Figure 1). Histopathological analysis revealed tumor stage of MF without transformation CD30. Tomographic scan of his chest and whole abdomen was normal. MF stage IIB was diagnosed and PUVA (30 fractionated *3/week) with acitretin (25mg/day) was started. All skin lesions were almost cleared.

Nine months after treatment, his skin lesions relapsed, made of multiple erythematous patches affecting 20% of body surface and appearance of three new tumors at the right mandibular level (Figure 2), the left scapula, and in folds of the left groin, with sizes varying between 5 and 15 cm (Figure 3). Skin biopsy of tumor showed a regular epidermis with an orthokeratotic keratosis. The dermis and hypodermis are infiltrated by medium at large-sized atypical lymphocytes with dark hyperconvulated nuclei and scant cytoplasm (Figure 4). The atypical lymphocytes were CD3+, CD4+, CD8-, CD20-, CD30+(2%), CD56-ALK- to immunohistochemistry (Figure 5). Examination of ganglionic areas showed several bilateral centimetric inguinal adenopathies, and the ultrasound objectified well differentiated adenopathy seat at the level axillary, inguinal and cervical, ranging from 6 to 15 mm. Laboratory examination showed a leukopenia with lymphopenia, the CD4 / CD8 ratio was 3.23, with absence of sesary cells in the blood, a lactate dehydrogenase level of 257U/L (normal range: 125-220U/L), with positive hepatitis B serology. A computed tomographic scan of his chest and whole abdomen showed tumor thickening of the sigmoid colon (Figure 6). Colonoscopy revealed an ulcero-burging tumor of the rectal sigmoid hinge with internal hemorrhoids stage 2, biopsy confirmed adenocarcinoma with medium differentiated and infiltrating arriving at the subserosa (Figure 7). From the clinical symptoms and the laboratory findings, large-cell transformed CD30+ MF stage IIIB associated with adenocarcinoma colic was diagnosed. The patient underwent a surgical excision of adenocarcinoma, which margins were healthy, complicated after two weeks by orchiepididymitis, and peritonitis with 2 surgical revisions and the patient was hospitalized in reanimation for 15 days and died by septic shock.

Discussion

Mycosis fungoides (MF) is the most common subtype of primary cutaneous T cell lymphoma, accounting for about 50% of cases. The course of mycosis fungoides (MF) is indolent except when transformation to a large T-cell lymphoma occurs. MF transformation (T-MF) is defined on the following histopathological basis: presence of large cells(4 times the size of a small lymphocyte) exceeding 25% of the infiltrate throughout or forming microscopic nodules, able to express or not the CD30 [2]. This transformation has been shown to represent an evolution of the original malignant clone. The diagnosis of transformation may be difficult because histopathologic criteria have a low reproducibility, and several differential clinical and/or histopathologic diagnoses exist. The diagnosis of T-MF is almost always made on skin biopsies in front of a clinical progression of a tumor or infiltrated plaques. Which prognosis is always poor, a mean 5-year survival rate of less than 20% [3, 4]. The causes of transformation are not known. Our patient had presented a mycosis fungoid which rapidly evolved into a CD30 transformed lymphoma, associated with adenocarcinoma of sigmoid colon found during the workup for systemic lymphoma involvement. This association is to our knowledge never described in literature, whose pathophysiology is unknown. We then hypothesized that the sudden aggravated skin lesions might be a cutaneous manifestation of paraneoplastic syndrome, because the cytokine secretion and a triggered immune response to antigens expressed in the tumor are thought to be responsible for the deterioration of the cutaneous lesions [5]. Especially that, the patient suffered a worsening of the cutaneous lesions at the discovery of the adenocarcinoma.

According to Curth's postulates, which defines the criteria for establishing a relationship between an internal malignancy and a skin disorder there is a specific type of neoplasia that occurs with paraneoplasia and a high frequency of association between both conditions [5]. The malignancy and the skin disease run a parallel course. Successful treatment of the tumor leads to regression of the skin disease, and recurrence of the tumor leads to a return of cutaneous signs and symptoms [6]. Our hypothesis seems logical especially that the patient had a fast evolution in stage of tumor, concomitant with the discovery of adenocarcinoma in the colon. Our case is similar to the case reported by Hye-Jin Ahn *et al.* the patient had a mycosis fungoid plaque with a 40% initial surface area, then she suffered a rapid worsening of plaque which became infiltrated and ulcerated with the detection of MF CD8+ on skin biopsy. The patient underwent further examinations to determine systemic involvement of extracutaneous lymphoma, but a sigmoid colon mass was detected by abdominal computed tomography, and biopsy of the mass found an adenocarcinoma. Stage IIIC sigmoid colon cancer with stage IB of MF was finally diagnosed, and she underwent surgical resection and treatment with 12 cycles of FOLFOX chemotherapy, after colon cancer treatment, the aggravated skin lesions improved, and the vegetative plaques resolved to brownish patches, similar to their appearance during the first visit. Since then, her MF has been well controlled with phototherapy [7].

Conclusion

To our knowledge, the association of mycosis fungoid transform and colon adenocarcinoma have never been described. Physiopathology is indeterminate, necessitates more epidemiological, genetic and physiopathological studies to explain the occurrence of cancer during cutaneous lymphoma, mainly transformed mycosis fungoides.

Competing interests

The authors declare no competing interests.

Authors' contributions

All the authors have read and agreed to the final manuscript.

Figures

Figure 1: initial appearance of the tumors at the right mandibular level

Figure 2: increase in tumors size after 3 months varying between 5 and 15 cm

Figure 3: erythematous and poikilodermatous patches affecting 20% of body surface

Figure 4: A) regular epidermis with an orthokeratotic keratosis; B) the dermis is infiltrated by medium at large-sized atypical lymphocyte

Figure 5: the atypical lymphocytes were CD3+, CD4+, CD8-, CD30+(2%), ALK- to immunohistochemistry

Figure 6: tumor at the level of the sigmoid colon

Figure 7: adenocarcinoma with medium differentiated and infiltrating arriving at the subserosa

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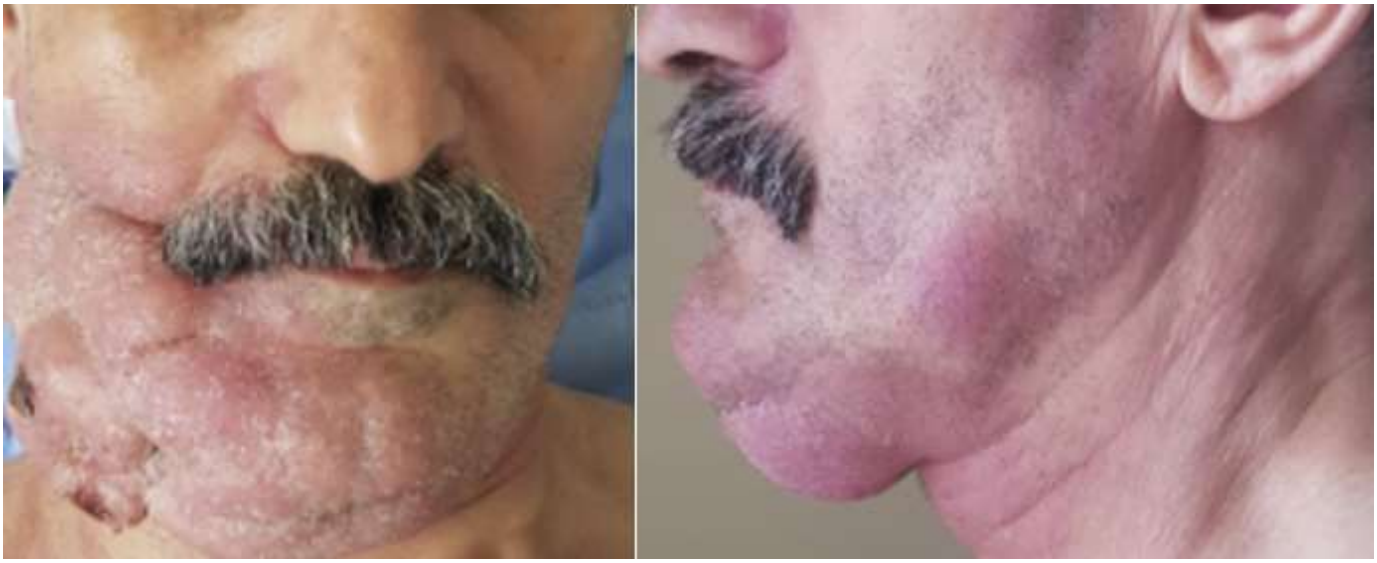

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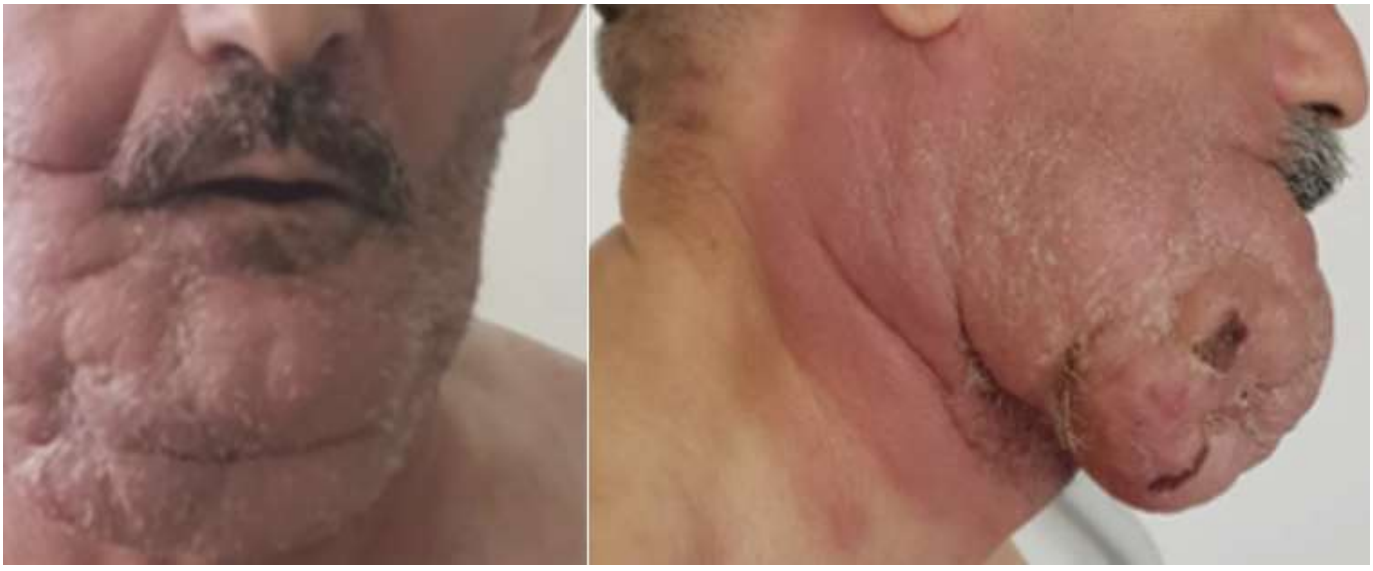
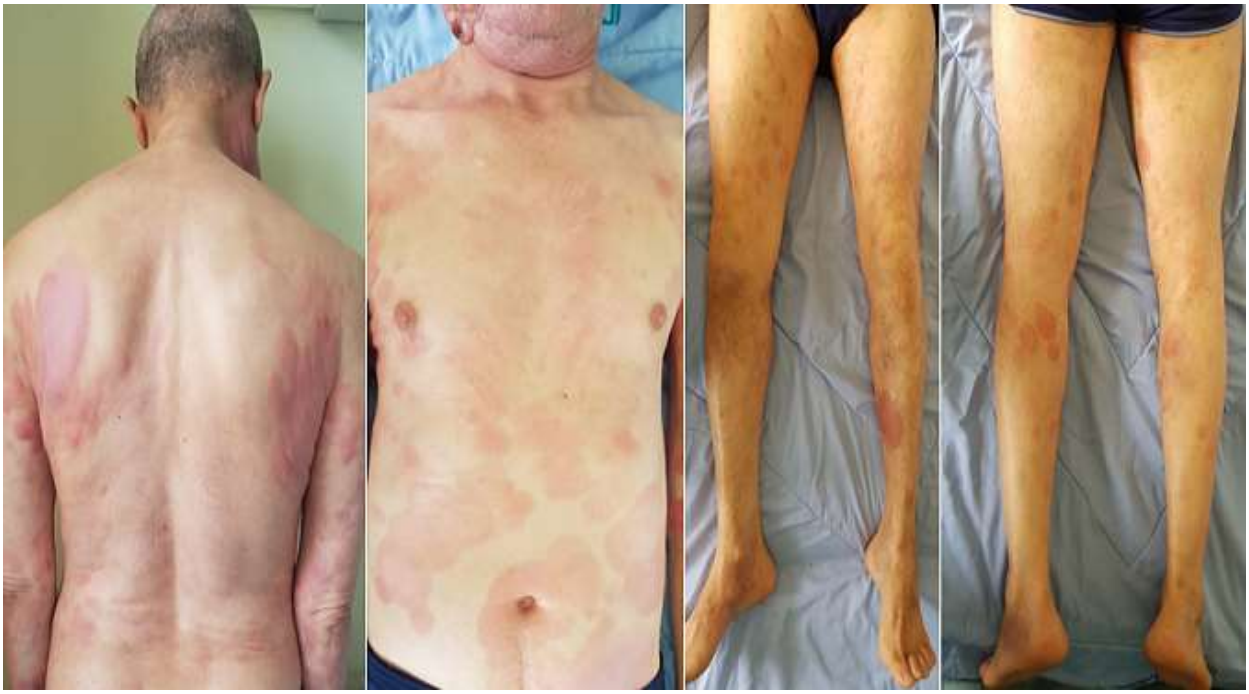


Figure 2: increase in tumors size after 3 months varying between 5 and 15 cm



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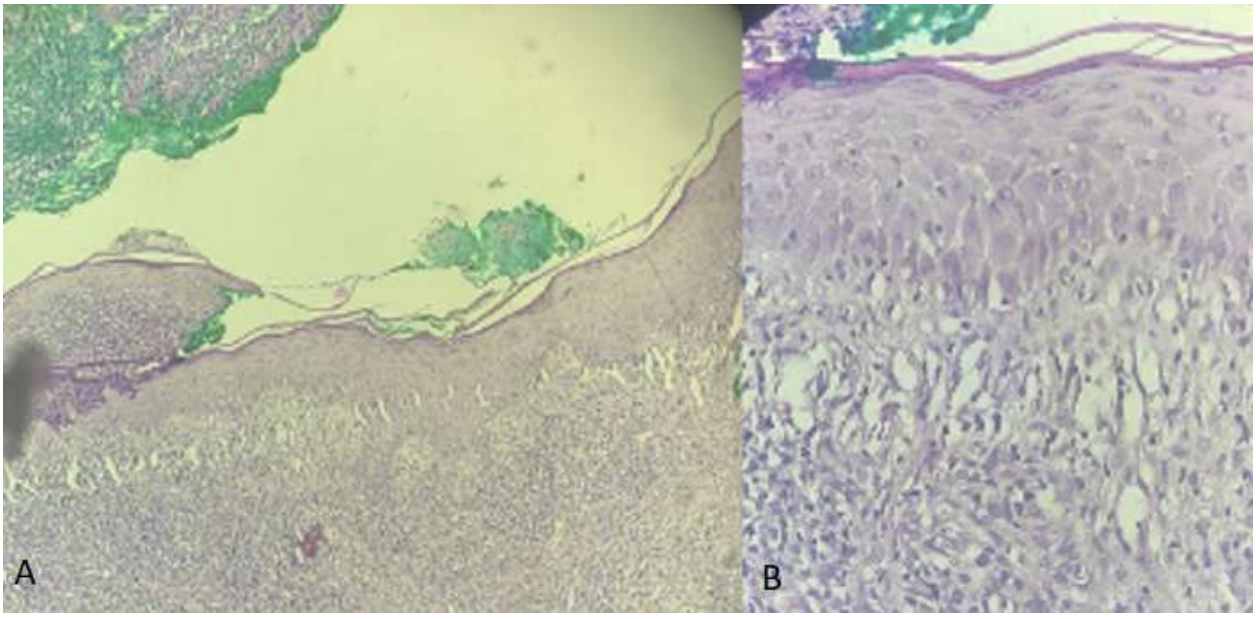

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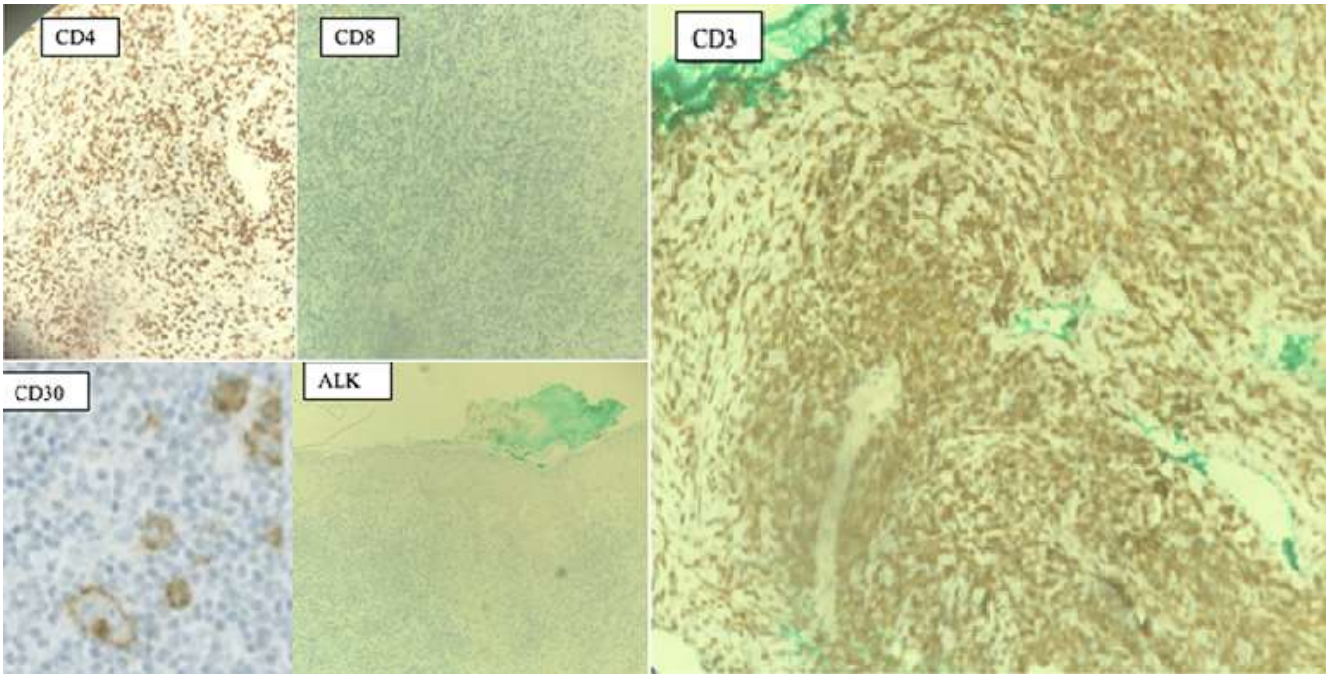

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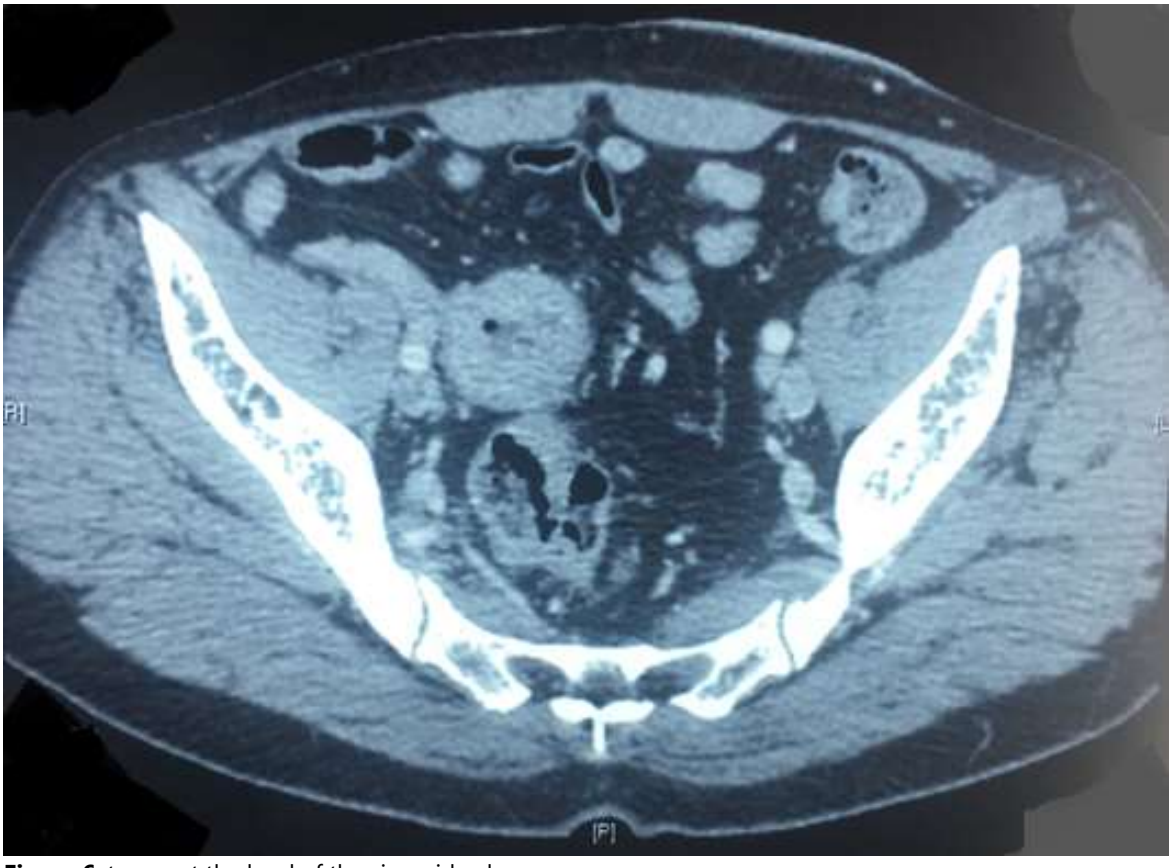

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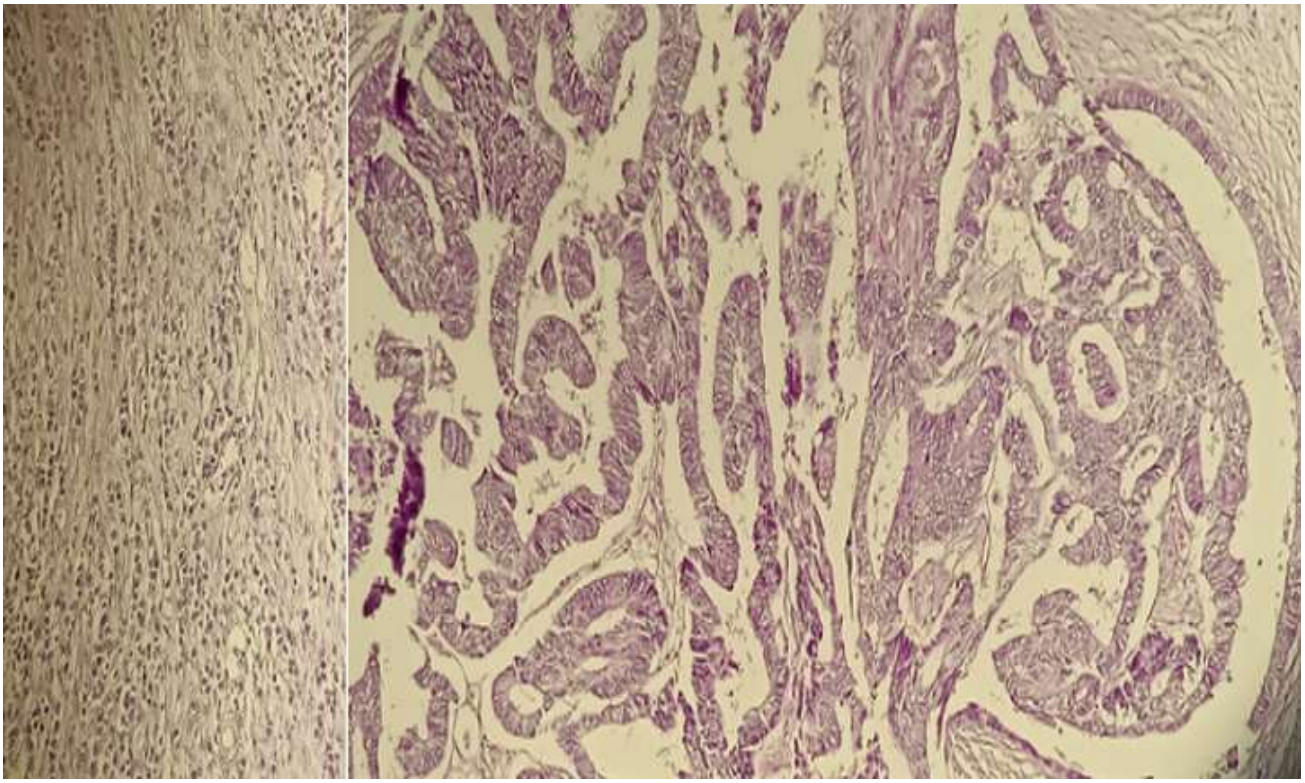

Figure 7: adenocarcinoma with medium differentiated and infiltrating arriving at the subserosa