Chronic itching unmasking extra mammary Paget disease in a celibate patient

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Background: Paget's disease, described by Sir James Paget in 1874 is classified as mammary and extra mammary. This later has been described initially by Crocker in 1889 and confirmed by Dubreuil in 1901, characterized by the presence of intra epithelial mucin producing neoplastic cells or apocrine gland bearing skin cells especially those located in the perineum of both sexes, axilla, groin, thigh, eyelid, external ear and umbilicus. It accounts for 10 % of Paget disease. It occurs mostly in postmenopausal Caucasian women without excluding men whom are mostly touched by the disease in Asian countries. It could be primary or secondary to intestinal and urogenital malignancy. Extra mammary Paget 's disease is often multifocal, and in many cases, it has been demonstrated extending beyond the visible lesion. Diagnosis is based on having high clinical index of suspicion, confirmed by the presence of Paget cell on histopathology study and immunohistochemistry staining. Herein, we report a case of extra mammary Paget disease associated with urogenital malignancy discovered at the invasive stage in a celibate patient.

Case Report: This 86-year-old celibate lady was referred to tertiary care with a chronic complain of pruritus. She was known to have chronic arterial hypertension, chronic coronary artery disease treated by percutaneous angioplasty and recurrent pulmonary embolism without evidence of deep vein thrombosis. She denied any history of tobacco smoking, alcohol, or using illicit drugs. The sole treatment over the counter she used to take is pain killer. She reported a history of erythematous lesion appearing on the external part of her labia majors evolving into scaring and super imposed eczematous scale. She had previously been treated empirically with oral and topical antibiotics, topical antifungal agents and topical glucocorticoids without improvement. She was celibate and has never had any sexual activity. She was afebrile and stable hemodynamically. The rest of physical examination was not contributory. Patient has had a full radiological investigation including upper and lower gastro intestinal endoscopy that was without abnormal finding apart of sigmoid diverticulosis. Chest x-ray was showing mild cardiomegaly with free costo-phrenic angles and ultrasound of the breast was normal. However, ultrasound of the abdomen and pelvis showed a polypoid bladder mass with variable echogenicity and thickened wall of the bladder and three hypo echogenic liver masses evoking metastasis. Kidney sizes were appropriate to the age of the patient without stone formation or dilation. There was no post voiding residual volume on ultrasonography. Patient was referred to urology clinic for evaluation and TDM and MRI abdomen and pelvis confirmed the previous findings with extension to lymph nodes in the retroperitoneal space and groins. Urinary cytology didn't show any neoplastic cell; however, cystoscopy confirmed a neoplastic nature of the tumor. Laboratory investigation was marked by grade I inflammatory anemia with high erythrocyte sedimentation rate at 114, CRP at 55.4mg/L (N<5) and increased ferritin level at 1413 ug/l (N<150). There was abnormal liver function tests in favor of cholestatic hepatitis (GGT =579 U/L and alkaline phosphatase at 442 U/L) and impaired kidney function test characterized by a creatinine clearance at 41 ml/min /1.73m2 with tubular proteinuria at 740 mg/24hours. TSH mildly elevated and lactic dehydrogenase = 695 U/l. Plasma protein electrophorese showed monoclonal gammopathy of undetermined significance and tumor markers were positive for CEA = 683(N<5), CA19.9 at 129, 5 (N <39). The rest of laboratory data was non-significant. Cystoscopy with biopsy of both lesions showed picture of Paget cells and immunohistochemistry stained for CK7, CEA, GATA -3 and negative for CK20, PS100 and MelA. Histology of bladder confirmed the presence of infiltrate carcinomatous proliferation with focus of necrosis and embolism of lymph node. The retained diagnosis was infiltrating urothelial carcinoma of the bladder with high-grade invasion (pT2 of UICC).

Discussion: Mammary and extra mammary Paget's disease (EMPD) is uncommon intra epithelial adenocarcinomas. Both conditions have similar clinical features and they are characterized by the presence of large oval or polyhedral intra epithelial cells that have pale cytoplasm and large nuclei with prominent nucleoli. They can be visualized using hematoxylin and eosin staining. The most common presenting symptom in extra mammary Paget's disease is pruritus. In many circumstances it can be asymptomatic slowly progressive, presenting as plaque, patch or just a red lesion or complicated appearance justifying ruling out eczema, chronic local infection, inflammatory conditions and tumors. Lesions occasionally showed hyperpigmentation or hypopigmentation. Unfortunately, our patient was having longstanding pruritus and failed many therapeutic attempts associated with chronic

complicated vulvar patches in the absence of further clinical examination. We never expect the nature of the disease until receiving the histopathological and immunohistochemistry. Herein, EMPD was found to be secondary urogenital malignancy with distal invasion, classified as type III according to reported authors. Treatment options include surgery, radiotherapy, photodynamic therapy, topical immunomodulator (Imiquimod 5%) with or without Trastuzumab. Our patient was unfit for these trials as long as the disease was extended, preferring symptomatic treatment.

Conclusion: chronic rebel itching may arise from multifactorial possibility including tumoral etiology that included extra mammary local or invaded Paget 's disease. High clinical index of suspicion with a thorough work up should be undertaken to rule out secondary.

Speaker Biography

H Y Fanomezantsoa is currently working in the department of Secondary Care in Louis Jaillon General Hospital, Saint Claude, France. Her research work includes Paget disease associated with urogenital malignancy discovered at the invasive stage in a celibate patient

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