Introduction/Background To report the case of a patient diagnosed with a Pagetoid vulvar lesion in the vulva with breast cancer and to conduct a literature review of the diagnosis, treatment, and prognosis in that location.

Methodology A 60-year-old patient who presented a pagetoid vulvar lesion with breast cancer to CPMC, Algiers, Algeria. The lesion was assessed on MRI and then surgically excised; histopathology showed invasive carcinoma of no special type (NST) after a mastectomy for the initial breast cancer. We reviewed PubMed for our search, all dates using the terms: breast cancer recurrence, breast cancer metastasis, vulva and breast cancer, metastatic vulvar cancer and vulvar cancer, ectopic localization.

Results Including our case, a total of 21 publications were listed including 9 cases of IDC, 5 cases of ILC, 2 cases of undifferentiated carcinomas, 2 cases not clinically described, 1 case of comedocarcinoma and 1 case of cystosarcoma phylloides. The time interval between the initial diagnosis of breast cancer and the secondary vulvar localization, ranges from 4 months to 255 months.

Conclusion Hartung, in 1872, first reported a fully formed mammary gland in the left labium majus of a 30-year-old woman. Even the ectopic breast tissue occurs along the milk lines, extending bilaterally from the mid-axillae through the normal breasts and then inferiorly to the medial groins. In women, the inferior extensions of the milk lines transverse the vulva bilaterally. In this case, Is it a secondary localization or an ectopic localization of an infiltrating breast carcinoma? Due to the rarity of this diagnosis, there are no established guidelines for the treatment of the patient. The appropriate treatment for a primary orthotopic breast cancer of a similar stage is recommended. Our patient was treated with local excision of the vulva and adjuvant.