Introduction: Mucinous cystadenocarcinoma (MCC) is a rare primary breast tumor, first described in 1998. It was removed from the WHO classification in 2012, due to lack of consensus about its definition. To our knowledge, only 25 cases have been published in the literature. Objective: Case report and discussion of a case of breast MCC diagnosed in Centro de Doenças da Mama in Curitiba. Case report: Premenopausal 51 year-old patient, complaining of pain and nodule in the left breast. The ultrasound showed a palpable nodule of approximately 2 cm, in the UOQ of the left breast. The PET-Scan did not show extramammary site capture. Conserving surgery and sentinel lymph node were performed, with breast reconstruction using the geometric compensation technique. Macroscopy showed a cystic and solid mass, of mucinous content, measuring 4.0x3.5 cm. The histological status was suggestive of MCC, with 2 negative sentinel lymph nodes. The IQ showed mammaglobin expression, CK7 and ER, negative expression of c-erbB-2, CK20 and CK5/6. The patient was submitted to radiotherapy and hormone therapy. Discussion: The primary breast MCC needs to be distinguished from ovarian and pancreatic metastasis. The IQ for CK7 and CK20 can be useful, considering that the pancreatic and ovarian MCC have concomitant expression of CK7 and CK20; the breast MCC expresses only CK7. In the 26 cases described in the literature, including this study, mean age was 62 years (41-96), and the tumor size was variable (0.8–19 cm). Only 4 cases presented positive lymph nodes. Most described cases did not express ER. The reported cases were associated with good prognosis. Conclusion: A consensus on the histological nomenclature and longer follow-up time are necessary to better understand this variant.