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## Background:



- Paget's disease of the nipple is very rare , 2-3% of all breast carcinoma.
- It has a high (67-100%) association rate with additional underlying malignant lesions <sup>(1)</sup>.
- Ductal carcinoma in-situ is the commonest concurrent malignant lesion found in Paget's disease (> 90% of cases).The diagnosis is however often underestimated due to mammographically occult DCIS.
- As a result, surgeons find it difficult to determine the appropriate management as surgical options vary depending on disease extent.
- Many case studies have reported the benefits of MRI as a problem solver in detecting underlying DCIS <sup>(2,3,4)</sup>.

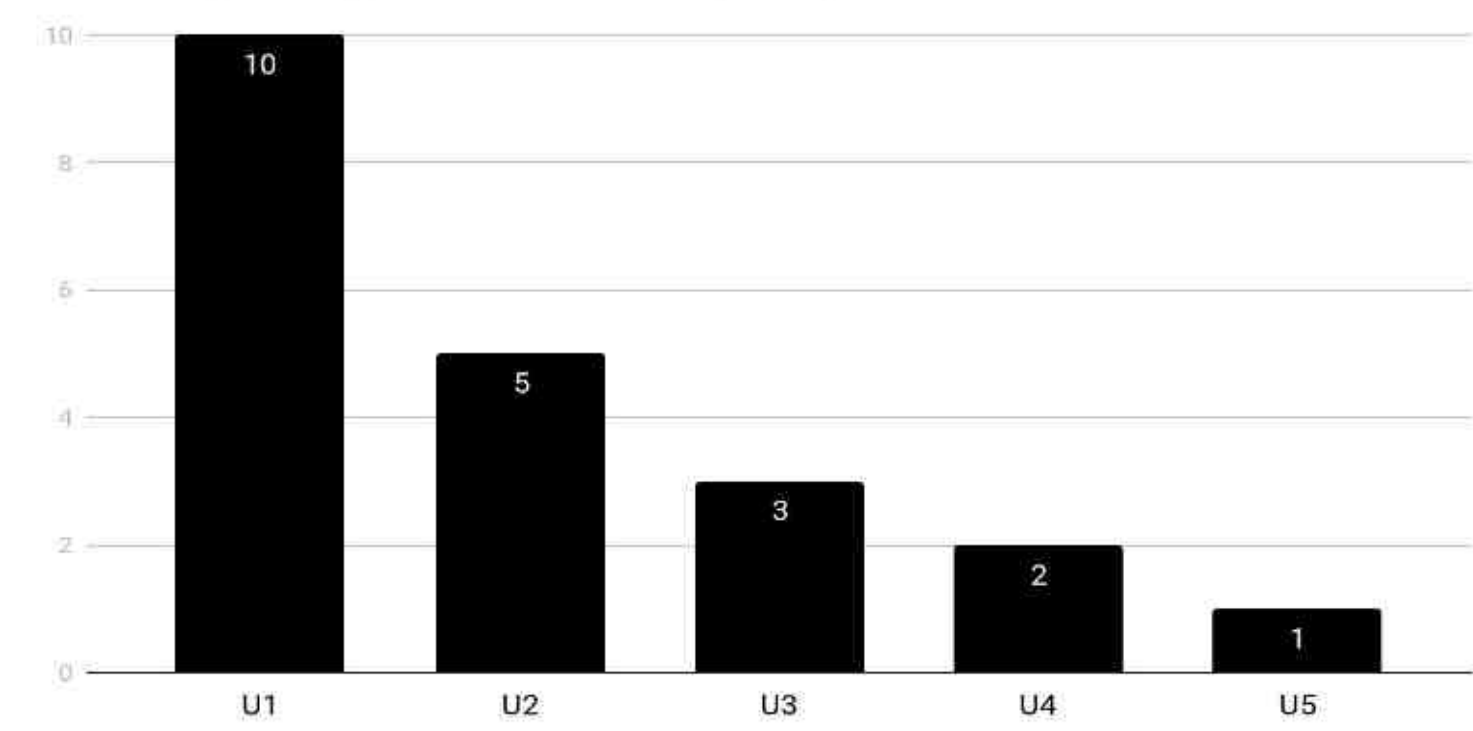
## Methodology:

Retrospective study on all patients diagnosed with Paget's diseases from 1st January 2012 to 31st March 2019 in 3 district general hospitals , where MRI is not part of routine workup for Paget's disease.

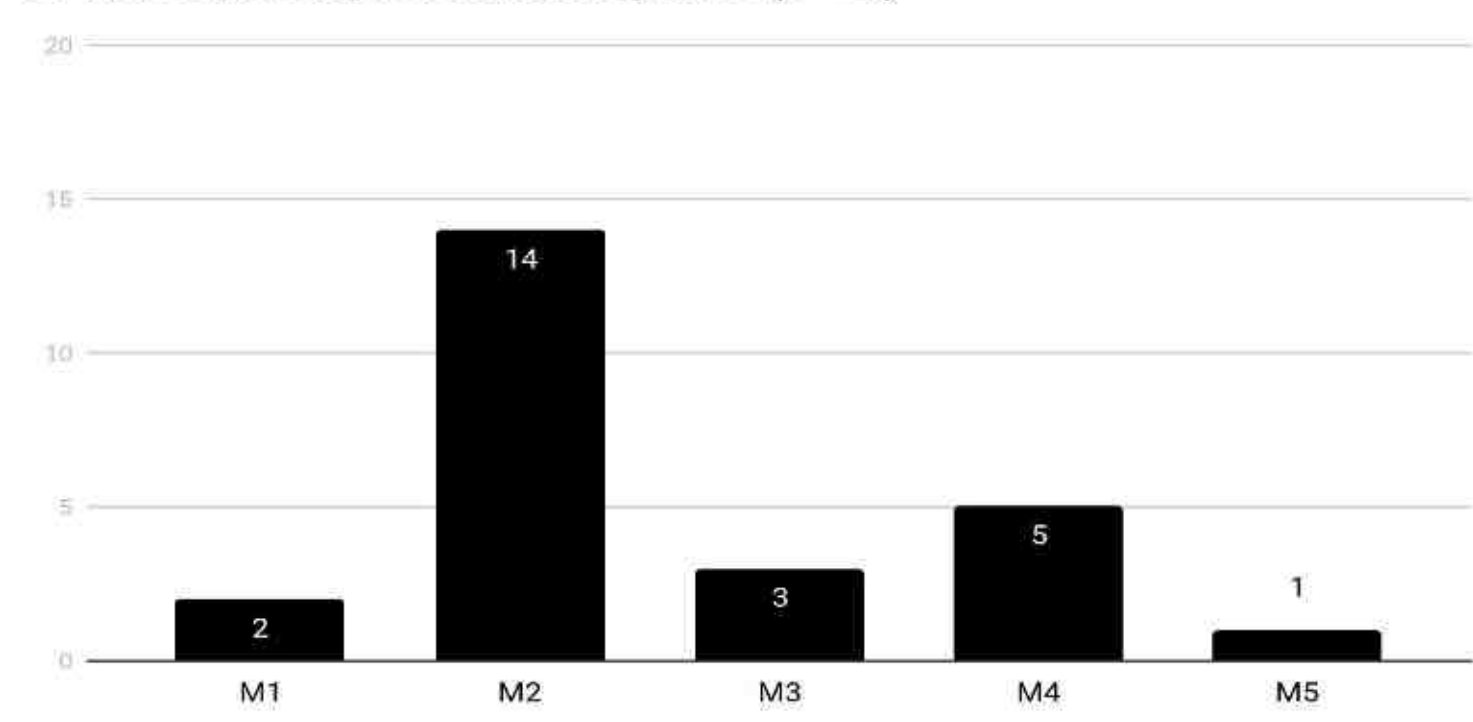
## Results:

1. 25 females with Paget's disease of nipple identified (age range 28-91; mean age 67) .
2. All patients except one were assessed with mammogram and ultrasound.

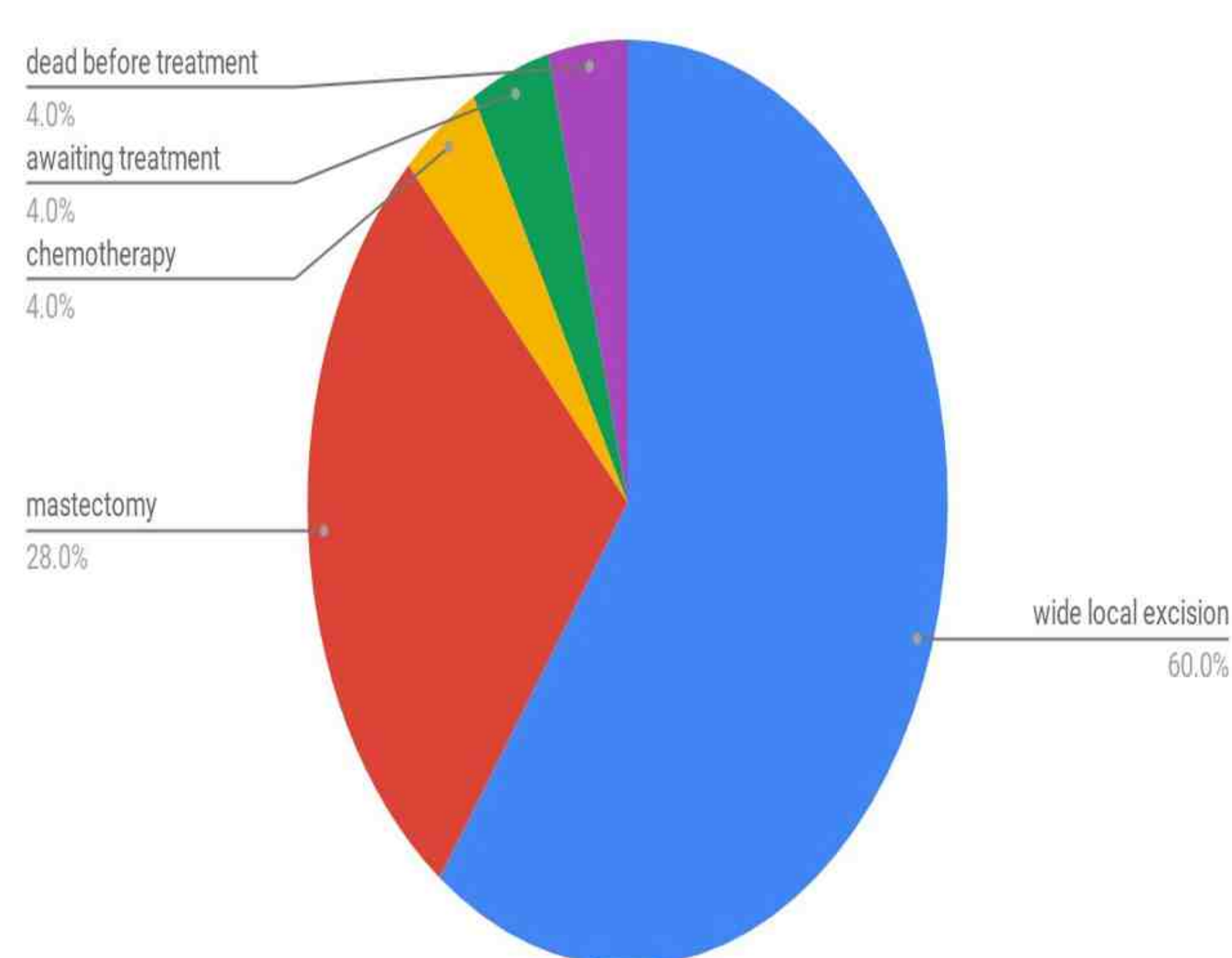
BI-RADS grading for ultrasounds (n=21)



BI-RADS grading for mammograms (n=24)



Management plans among patients with Paget's disease



3. 22/25 patients were managed surgically.15 wide local excision; 7 mastectomies.
4. Pathological analysis detected concomitant DCIS on 16/22 resected specimens (73%). 3/16 patients with DCIS demonstrated invasive disease. Out of 16 patients with DCIS, only 6 were detected on preoperative conventional imaging.
5. 3 patients needed repeat surgeries due to margin involvement, disease recurrence and widespread distribution of Paget's disease. None of them had been assessed with MRI.
6. 5 patients had preoperative MRI. Multifocal invasive carcinoma was diagnosed on MRI in one patient, with resultant mastectomy (see case 1). Another patient also had mastectomy for DCIS which was shown to be more extensive on MRI than mammogram (see case 2). Two MRIs were unable to detect underlying DCIS due to size/subtly of lesion (1mm and 15mm). One MRI confirmed Paget's disease with no additional lesion.

## Discussion:

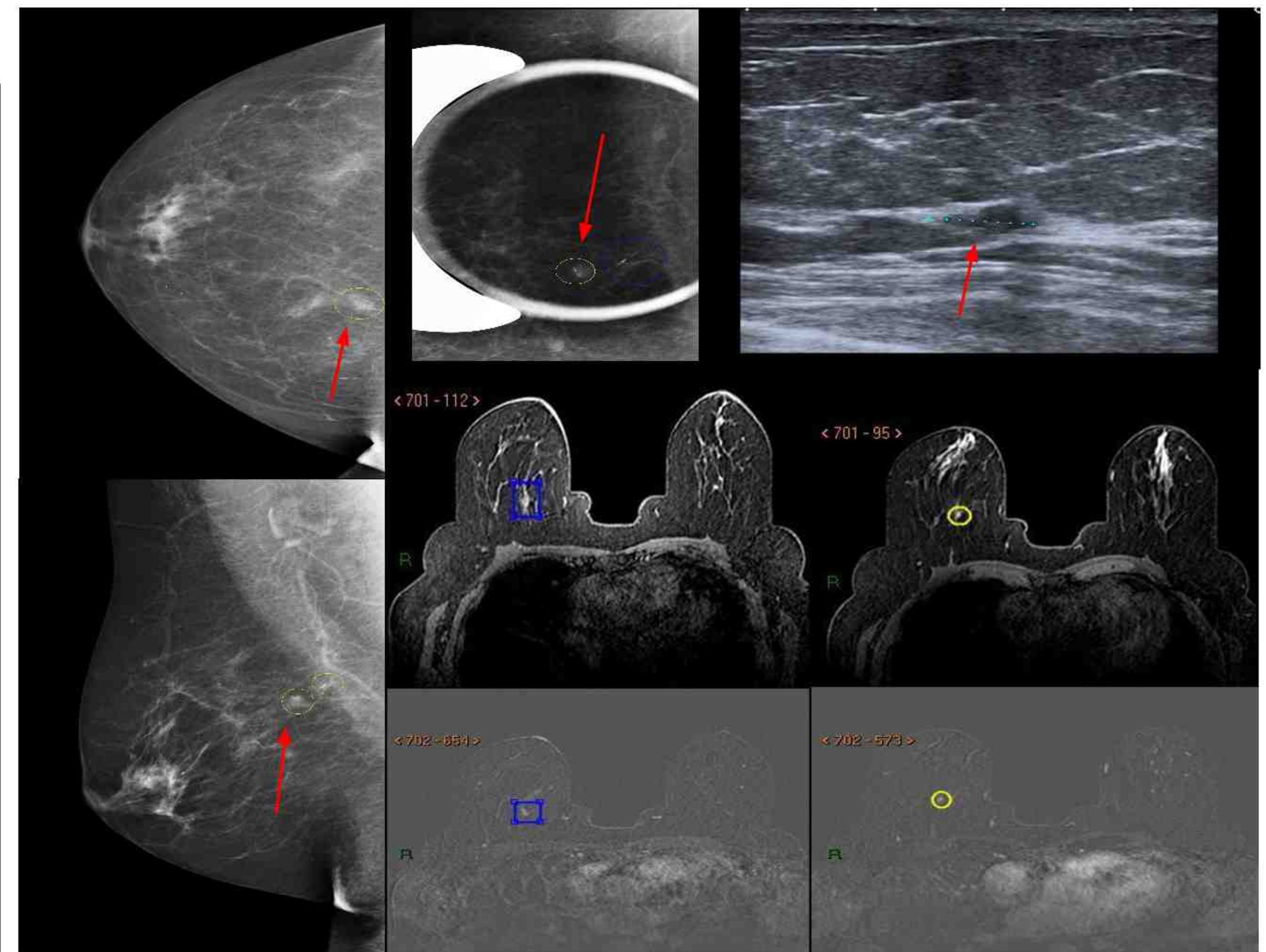
- DCIS is usually non-palpable and can be non-calcified therefore undetectable on conventional imaging.
- Based on the signal intensity and contrast enhancement pattern, MRI has higher sensitivity in detecting additional lesions and more accurately estimating the size of tumour pre-operatively. The sensitivity of MRI in detecting high-grade DCIS is reported higher than that of mammography (92% vs. 56%) according to Kuhl et al<sup>(5)</sup>, especially for high grade DCIS without necrosis (97% vs. 35%) .
- However, it is important to recognise the low specificity of MRI in differentiating benign from malignant lesions and that it still has limitations in detecting DCIS. Despite this, MRI is recommended as a supplemental tool rather than as a substitution for conventional imaging.

## Conclusion:

The benefits of MRI input were proven in our study and are felt to outweigh its limitations. Given the potential for MRI to more accurately detect and size concomitant pathology, we propose that all patients with Paget's disease have pre-operative MRI to allow appropriate surgical management from the outset.

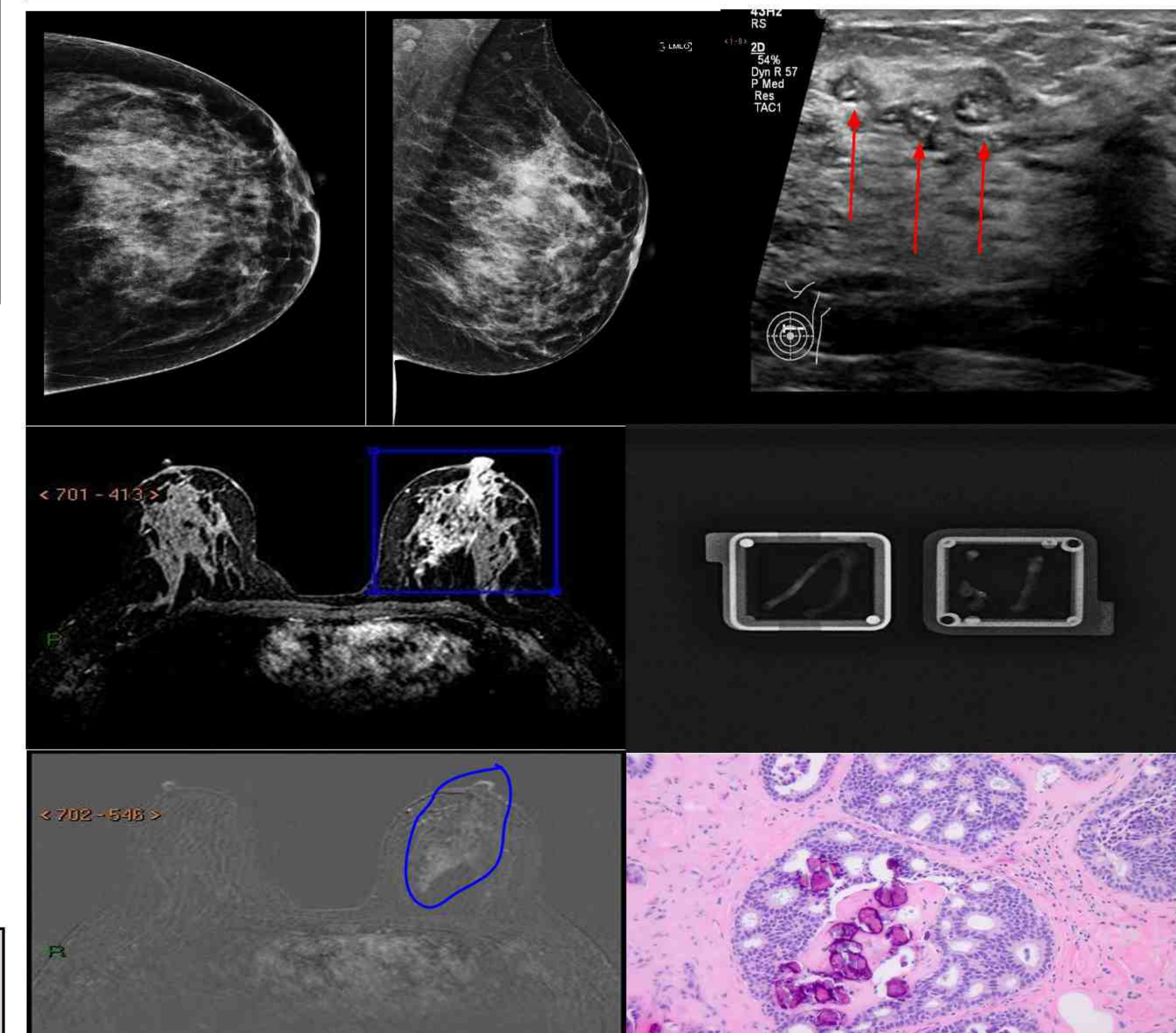
## Case 1

67 year-old patient first presented with nipple itchiness and had punch biopsy proven Paget's disease. No evidence of DCIS on punch biopsy specimen. Routine mammogram (*RCC, RML0 and magnification views provided*) showed a focal nodule with suspicious microcalcification (*yellow circle & red arrows*) posteriorly in the right upper inner quadrant. The lesion was later confirmed as invasive carcinoma, B5b, on ultrasound-guided core biopsy (*red arrow*). Patient was further assessed with contrast enhanced MRI which detected **a second invasive lesion** . *Selected slices of T1 weighted high-resolution isotropic volume examination (THRIVE) and digital subtraction sequences with both 1st lesion (blue square) and 2nd lesion (yellow circle) as attached below*. In view of multifocal invasive breast carcinoma with the distance between two focal lesions 3.1cm apart, patient was offered mastectomy instead of breast conserving surgery.



## Case 2

A 28 year old patient with Paget's disease had numerous pleomorphic microcalcifications on mammography (*LCC, LML0 views provided*).Ultrasound guided core biopsy proven DCIS (*uss with red arrows showed hyperechoic calcified lesion ; microcalcification within biopsied specimen on mmg*). She was further assessed with dynamic contrast enhanced MRI. Based on the contrast uptake, the extent of DCIS disease shown on MRI was **more extensive** than mammograms. *Selected slices of T1 weighted high-resolution isotropic volume examination (THRIVE) and digital subtraction sequences demonstrated a single extensive contrast enhancing area abutting the nipple-areolar complex*. Patient had left mastectomy with clear margin achieved. No re-excision required.



References:  
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2. Eleonora Caspari, Aurora Ricci, Valeria Liberto, Angela Lia Sciarano, Maria Fornari, and Giovanni Simonetti. "An Unusual Case of Mammary Paget's Disease Diagnosed Using Dynamic Contrast-Enhanced MRI" *Case Reports in Radiology*, Volume 2013, Article ID 206235, 5 pages  
3. Gudrun Peters, Mary Self, Anne Lynch, Lynne Brothers. "Breast MRI as a Problem Solving Tool in Paget's Disease of the Nipple". *Journal of Current Surgery* ,Volume 2, Number 3, June 2012, pages 113-115  
4. Morogh M, Morris EA, Liberman L, Van Zee K, Cody HS, 3rd, King TA. MRI identifies otherwise occult disease in select patients with Paget disease of the nipple. *J Am Coll Surg*. 2008;206(2):316-321.  
5. Kuhl CK, Schradang S, Bieling HB, Wardelmann E, Leutner CC, Koenig R, et al. MRI for diagnosis of pure ductal carcinoma in situ: a prospective observational study. *Lancet*. 2007;370:485-92.