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Surgical treatment in Paget's disease of the breast

A particular type of nipple pathology is Paget's disease and it constitutes approx. 1-2% of all malignant neoplasms of the breast (10). In 1874 James Paget was the first to describe the lesions similar to eczema or ulceration which in time may develop into breast carcinoma. Nowadays the two forms of Paget's disease are identified: Paget's disease in its preinvasive form (with intraductal component) and Paget's disease as invasive carcinoma of lesser malignancy. The latter may spread along the superficial epithelium from the inside of the breast to the nipple as well as from the nipple to the inside of the breast beyond the basement membrane, as invasive carcinoma (7). Paget's disease has the characteristic histopathological feature, which is the occurrence of large, single, light cells and groups of cells called Paget's cells among the proper epidermal or epithelial cells. These cells are PAS positive, they have atypical nuclei and abundant cytoplasm. Most histopathology experts consider them to be neoplastic by nature (4, 9).

The most common therapy in Paget's disease is surgical treatment. The extent of the surgery is broad and it comprises mastectomy with the axillary nodes dissection, simple mastectomy, and breast conserving therapy. Breast conserving therapy has successfully been practised for years. It was first introduced by Curie Institute in Paris, which pioneered to apply tumorectomy with radiotherapy in breast cancer treatment as early as in 1960. Any doubts and uncertainty surrounding this method should be dispelled by the results of metaanalysis comparing various methods of Paget's disease treatment presented in 1995 by Early Breast Cancer Trialist Collaborative Group. These results confirmed the equal effectiveness of radical mastectomy and breast conserving therapy since the numbers of long-term survivals and the numbers of cases of local regrowth were almost identical in both methods (6). The primary objective of breast conserving therapy is to ensure a desired cosmetic and functional effect, which accords with the current trends in oncological surgery to preserve organs and simultaneously maintain the highest possible curability (5). Breast conserving therapy, however, has its limitations and breaching

these increases the risk of local regrowth (3, 4, 6). One of these limitations is the central location of tumour. The removal of the nipple-areola of the mamma-breast tissue complex, which is necessary in breast conserving therapy may lead to an ill cosmetic effect. The patient's approval and preserving the proper disease-free margins in histopathological examination is the indication for this type of therapy.

RESULTS

In the years 1989-1995, 2,261 female patients with breast cancer received surgical treatment in the Surgery Department of the Centre of Oncology of the Lublin Region. The patients were treated with breast conserving therapy followed by radiotherapy, and the patients from the increased risk group received chemotherapy. The analysed group of patients is presented in Table 1.

Table 1

Year	n	Average age	Paget's disease with intraductal component	Paget's disease with infiltration	Radical mastectomy	Breast conserving therapy
1989	2	53	2	0	2	0
1990	4	49	3	1	4	0
1991	6	56	5	1	6	0
1992	7	58	7	0	7	0
1993	4	52	3	1	3	1
1994	5	50	3	2	3	2
1995	7	54	5	2	4	3
Total	35	53.1	28	7	29	6

In 35 of the patients, which is 1.51%, Paget's disease was identified; 29 of the patients received radical mastectomy, and 6 – breast conserving therapy. The patients' ages varied from 42 to 71, and the average age was 53.1 years. 28 of the patients were diagnosed to have Paget's disease with intraductal component and 7 – invasive form of the carcinoma.

Table 2

	n	Metastases to lymph nodes	Incidence of local regrowth	5-year survivals in %
Paget's disease with intraductal component	28	0	1	89.76
Paget's disease with infiltration	7	4	4	49.32

Radical mastectomy was performed in 29 patients and breast conserving therapy – in 6 patients.

5-year survivals amounted to 89,76% with the patients with intraductal component, and 49.32% with the patients with infiltrating component. Local regrowth was diagnosed in 4 patients with invasive form of the carcinoma who had undergone radical mastectomy and in 1 patient with intraductal component after breast conserving therapy. Metastases to the axillary lymph nodes were found in 4 patients with the infiltrating form of Paget's disease.

DISCUSSION

The decision making process in relation to the extent of surgical treatment in Paget's disease is still very complicated due to limited possibilities of diagnosing the advancement of carcinoma. Planning the extent of an operation may hardly be assisted by mammography whose diagnostic value in Paget's disease with non-palpable indurations was assessed to be 64% (1, 8). All the surgeons specialising in breast conserving therapy agree that this type of therapy should be applied only in the patients with whom it is possible to achieve satisfying cosmetic effect without exposing them to the increased risk of local regrowth (2, 3, 4, 6). The central location of the carcinoma is not a counter-indication for this type of therapy, however, it involves another operation to reconstruct the nipple and its areola (3). In Paget's disease the difficulties to diagnose cancer-free margins in intraoperation examination are encountered both in the intraductal and the infiltrating component (4). It should be noted that excessive removal of breast tissue spoils the aesthetic effect of an operation (5). Acquiring experience in breast conserving therapy leads to a greater number of patients qualified for for this type of treatment. In 1993 we performed only one breast conserving operation, but in 1995 – three.

CONCLUSIONS

Breast conserving therapy in Paget's disease presents itself as favourable alternative type of therapy, which is more beneficial for a patient than mastectomy. Planning an operation in Paget's disease it is also advisable to consider simple mastectomy preserving the skin followed by the breast reconstruction with endoprosthesis or autologous tissues.

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SUMMARY

The extent of the operation in Paget's disease of the breast depends largely on the degree of clinical advancement of neoplasm as well as counter-indications for breast conserving therapy. Despite the increasing use of breast conserving therapy, rather few such operations of this kind have been performed in our department. We have carried out 29 mastectomies and only 6 breast conserving operations. No matter what type of therapy was applied the percentages of 5-year survivals for the patients with non-invasive component and for the ones with invasive component were respectively 89.76% and 49.32%.

Leczenie operacyjne choroby Pageta gruczołu piersiowego

Zakres operacji w chorobie Pageta gruczołu piersiowego zależy od stopnia zaawansowania nowotworu oraz przeciwwskazań do leczenia oszczędzającego. Mimo coraz powszechniejszego stosowania operacji z zaoszczędzeniem piersi w naszym materiale było ono niewielkie. Wykonaliśmy bowiem 29 mastektomii i tylko 6 operacji oszczędzających pierś. Bez względu na sposób leczenia przeżycia 5-letnie dla chorych z komponentą nieinwazyjną wyniosły 89,76%, a inwazyjną 49,32%.