6. TG W, SE R, KD T, AB H, SR L, WR B. et al. An estimation of the global volume of surgery: a modelling strategy based on available data. Lancet (London England). 2008;372(9633):139–44. Available from: https://pubmed.ncbi.nlm.nih.gov/18582931/.
7. CE G, KG B, CM D, CB L. Systematic review of barriers to surgical care in low-income and middle-income countries. World J Surg [Internet]. 2011;35(5):941–50. Available from: https://pubmed.ncbi.nlm.nih.gov/21360305/.
8. LM K, P B, S C, T C, TE C, B D. et al. A geospatial evaluation of timely access to surgical care in seven countries. Bull World Health Organ [Internet]. 2017;95(6):437–44. Available from: https://pubmed.ncbi.nlm.nih.gov/28603310/.
9. CE G, R L, A D, N D, S R, M M. et al. Cost-Effectiveness of Two Government District Hospitals in Sub-Saharan Africa. World J Surg [Internet]. 2017;41(9):2187–92. Available from: https://pubmed.ncbi.nlm.nih.gov/28349322/.
10. BC A, MG S, AJ D, JR V, JG M. Global economic consequences of selected surgical diseases: a modelling study. Lancet Glob Heal [Internet]. 2015;3 Suppl 2:S21–7. Available from: https://pubmed.ncbi.nlm.nih.gov/25926317/.

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Breast cancer and Poland's syndrome

Síndrome de Poland y cáncer de mama

Poland's syndrome (PS) is a very rare congenital musculoskeletal disorder characterised by hypoplasia or absence of the pectoralis major muscle and may be associated with breast hypoplasia, ammastia, chest wall deformities and ipsilateral upper limb abnormalities. A higher incidence of certain malignant tumours, including infiltrating ductal carcinoma (IDC) of the breast, has been reported in these patients. We present 2 cases of PS associated with breast carcinoma operated on in our department, describing their characteristics and oncological evolution.

Case 1

A 39-year-old woman with right PS (breast hypoplasia, absence of pectoralis, shortening of the ipsilateral arm and malformations of the fingers), and with a history of a contralateral symmetry mammoplasty. She started with a 20 mm nodule in the upper outer quadrant of the right breast, with no pathological lymphadenopathies. Mammography showed a 2 cm spiculated nodule, BIRADS 5. The core needle biopsy indicated IDC. She underwent lumpectomy and sentinel node biopsy (SNB) with definitive anatomopathological results of grade III IDC, hormone receptor (HR) positive and Her2-positive enlargement, stage T1N0M0. After surgery, she evolved favourably and was discharged 24 h after surgery. She received 6 cycles of fluorouracil, epirubicin and cyclophosphamide (FEC) associated with trastuzumab for one year. In addition to radiotherapy (50 Gy + boost 10 Gy) and hormone therapy with tamoxifen for 5 years. There was no recurrence with a 10-year follow-up.

Case 2

A 48-year-old female patient with a history of right PS due to breast hypoplasia. She was assessed for pain and local discomfort in the right breast. Physical examination showed induration and retraction of the nipple, with no palpable nodules or lymph nodes. Mammography revealed small retroareolar punctate calcifications with no other alterations and a skin biopsy of the nipple was performed. The result was positive for stage T4bN0M0 IDC. She received neoadjuvant treatment with 4 cycles of doxorubicin and cyclophosphamide followed by 12 cycles of paclitaxel with subsequent skin-sparing mastectomy, SNB and immediate reconstruction with direct prosthesis. Pathological anatomy showed a 3 cm grade III IDC with free borders, HR positive, Ki-67 of 15% and negative Her2 magnification. The SNB was negative for metastasis. She had a satisfactory evolution and was discharged 48 h postoperatively. Subsequently, hormone therapy with exemestane was started, and treatment

Table 1 – Cases of Poland's syndrome and breast cancer reported in the literature.

#	Author	Age	Side	Breast involved	Histology	Hormonal receptors	Stage	Surgery	Hormonal treatment	CT	RT
1	Fukushima et al., 1999	57	Right	Ipsilateral	IDC	O+ P+ Her2 N/R	IIA T1N1M0	Mastectomy + AE	–	–	–
2	Fukushima et al., 1999	53	Left	Ipsilateral	IDC	O– P– Her2 N/R	IIA T2N0M0	Tumorectomy + AE	–	–	–
3	Havlik et al., 1999	33	Right	Ipsilateral	IDC + ILC	O+ P– Her2 N/R	IIA T2N0M0	Mastectomy + AE	–	–	–
4	Katz et al., 2001	42	Left	Ipsilateral	IDC	O– P+ Her2–	IA T1N0M0	Mastectomy + AE	–	+	–
5	Okamo et al., 2002	59	Right	Contralateral	IDC	O+ P+ Her2 N/R	IIA T1N1bM0	Tumorectomy + AE	–	+	–
6	Khandelwal et al., 2004	71	Right	Ipsilateral	IDC + ISDC	O+ P– Her2–	IA T1N0M0	Mastectomy + AE	–	–	–
7	Wong et al., 2004	51	Left	Ipsilateral	ISDC	ON/R P N/R Her2 N/R	0 TisN0M0	Mastectomy	–	–	–
8	Tamiolakis et al., 2004	53	Left	Ipsilateral	IDC	O– P+ Her2–	IA T1N0M0	Mastectomy + AE	–	+	–
9	Salhab et al., 2005	52	Left	Ipsilateral	IDC + ISDC	O N/R P N/R Her2 N/R	IA T1N0M0	Tumorectomy + SNB	–	–	–
10	Wang et al., 2008	46	Right	Ipsilateral	IDC	O+ P+ Her2–	IIB T2N1M0	Mastectomy + AE	–	+	+
11	Ji et al., 2008	58	Left	Ipsilateral	IDC	O N/R P N/R Her2 N/R	IIA T2N0M0	Mastectomy + AE	–	–	–
12	Caussa et al., 2009	43	Left	Ipsilateral	IDC	O+ P N/R Her2–	IIIA T3N1M0	Mastectomy + AE	+	+	+
13	Yesilkaya et al., 2011	39	Left	Ipsilateral	IDC	O– P+ Her2+	IIA T2N0M0	Mastectomy + AE	+	+	–
14	Zhang et al., 2011	43	Left	Ipsilateral	IDC	O+ P+ Her2+	IIIC T1N3M0	Mastectomy + AE	–	+	–
15	DeFacio et al., 2017	62	Right	Contralateral	IDC	O– P– Her2+	IIA T2N0M0	Mastectomy bilateral + SNB	–	+	–

Table 1 (*Continued*)

#	Author	Age	Side	Breast involved	Histology	Hormonal receptors	Stage	Surgery	Hormonal treatment	CT	RT
16	DeFacio et al., 2018	69	Left	Ipsilateral	IDC	O+ P− Her2-	IA T1bN0M0	Tumorectomy + SNB	−	−	+
17	DeFacio et al., 2018	37	Right	Bilateral	R: ISDC L: IDC	O R+ L− P R− L− Her2 R+ L+	R: IA T1bN0M0 I: IA T1cN0M0	Mastectomy + AE bilateral	−	−	−
18	Huang et al., 2018	74	Left	Ipsilateral	IDC	O+ P+ Her2−	IIB T3N0M0	Mastectomy + AE	+	−	−
19	Huang et al., 2018	60	Left	Ipsilateral	IDC	O− P− Her2+	IIIB T4N1M0	Mastectomy + AE	−	−	−
20	Malatay Gonzalez et al., 2020	56	Left	Ipsilateral	IDC	O+ P+ Her2+	IV T3N0M1	Mastectomy + AE	+	+	+
21	Case 1, 2007	39	Right	Ipsilateral	IDC	O+ P+ Her2+	IA T1cN0M0	Tumorectomy + SNB	+	+	+
22	Case 2, 2018	48	Right	Ipsilateral	IDC	O+ P+ Her2−	IIIB T4bN0M0	Mastectomy + SNB	+	+	−

AE: Axillary emptying; CT: Chemotherapy; IDC: Infiltrating ductal carcinoma; ILC: Infiltrating lobular carcinoma; ISDC: In situ ductal carcinoma; L: Left breast; N/R: Not reported; O: Oestrogen receptors; P: Progesterone receptors; R: Right breast; RT: Radiotherapy; SNB: Sentinel node biopsy.

has been ongoing to date, with no evidence of tumour recurrence.

To date, 22 cases of PS associated with breast carcinoma have been described (Table 1). Of the cases described, 59% were in the left breast, with malignant degeneration of the hypoplastic breast in 86.3% of the cases, with the healthy breast being affected in only 9% and only one case with bilateral involvement. Histology was IDC in 95% of cases, with oestrogen and progesterone receptor positivity in 68% and 53%, respectively, and Her2 amplification in 54%. Mastectomy with or without reconstruction was performed in most cases (77%). Our cases showed a right predominance, with involvement of the hypoplastic breast by IDC and HR expression. Only one of them had Her2 enlargement (case 1).

PS is a congenital musculoskeletal disorder with an estimated incidence of 1:20,000 to 1:32,000 births,[1] with a predilection for males.[2] Breast involvement can range from mild hypoplasia to complete absence or ammastia.[3] Seventy percent of cases affect the right side of the body.

Although the aetiology is unknown, the main defect is thought to be in the mesoderm from which the pectoral area and upper limb develop, due to decreased blood flow from the subclavian in this region during embryonic development.[4] These vascular alterations have been demonstrated in some studies.[5,6,8]

The diagnosis of this disease is mainly clinical. In some cases, the clinical features are not so characteristic and may only present breast asymmetry or slight alterations of the chest wall, so this disease may be underdiagnosed. In this case, complementary imaging tests such as chest wall ultrasound, computed axial tomography or magnetic resonance imaging are recommended.[5]

Surgical treatment of breast cancer in these patients does not differ from that of the general population,[9] although it is necessary to take extreme precautions to avoid thoracic lesions due to the possible absence of both pectoral muscles.[8] Aesthetic correction of the hypoplastic breast requires individualised treatment and may require a contralateral symmetry mammoplasty.[7] Greater caution should be exercised with the use of external radiotherapy in these patients, as it carries a higher risk of pulmonary and cardiac lesions and complications due to the absence of pectoral muscle to protect the intrathoracic organs.

Future studies are needed to establish a definitive relationship between the two pathologies, although this could be complicated due to the low incidence of this disease and the greater involvement of males.

To date, 22 cases of SP associated with breast carcinoma have been reported. Although there is no definitive association between the two, there may be an increased risk of cancer in these patients. We therefore recommend closer follow-up and a high level of suspicion when nodules or other breast alterations appear.

Conflict of interests

The authors have no conflict of interests to declare.

REFERENCES

1. Freire-Maia NF, Chautard EA, Opitz JM, Freire-Maia A, Quelce-Salgado A. The Poland syndrome: Clinical and genealogical data, dermatoglyphic analysis, and incidence. Hum Hered. 1973;23:97–104.
2. Fokin AA, Robicsek F. Poland's syndrome revisited. Ann Thorac Surg. 2002;74:2218–25.
3. Bland KI, Copeland EM. The Breast: Comprehensive Management of Benign and Malignant Diseases. Ann Surg. 1992;215(2):192.
4. Bavinck JN, Weaver DD. Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland, Klippel-Feil, and Mobius anomalies. Am J Med Genet. 1986;23:903–18.
5. Mentzel HJ, Seidel J, Sauner D, et al. Radiological aspects of the Poland syndrome and implications for treatment: a case study and review. Eur J Pediatr. 2002;161:455–9.
6. Katz SC, Hazen A, Colen SR, DF Roses. Poland's syndrome and carcinoma of the breast: A case report. Breast J. 2001;7:56–9.
7. Mojallal A, La Marca S, Shipkov C, Sinna R, Braye F. Poland syndrome and breast tumor: a case report and review of the literature. Aesthet Surg J. 2012;32(1):77–83.
8. DeFazio MV, Dervishaj OA, Bozzuto LM, Pittman TA, Olding MJ, Tousimis EA, et al. Delayed Recurrent and Bilateral Breast Cancer in Patients With Partial Poland's Anomaly: Report of 2 Rare Cases and Review of the Literature. Clin Breast Cancer. 2018;18(3):285–90.
9. Malatay Gonzalez C, et al. Cáncer de mama asociado a síndrome de Poland. Revista de Senología y Patología Mamaria. 2021. http://dx.doi.org/10.1016/j.senol.2020.09.006.

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