

Poland's Syndrome and Breast Cancer: Case Report and Review of The Literature

Ali İmran KÜÇÜK ^{*o}, Cem ERSİN ^{**}, Yakup İRİAĞAÇ ^{***}

Poland's Syndrome and Breast Cancer: Case Report and Review of The Literature

SUMMARY

Poland's syndrome is a rare congenital disorder characterized by congenital musculoskeletal anomalies. These patients usually have hypoplasia or absence of the pectoralis major muscle and anomalies of the chest wall and upper extremities. Breast cancer may occur despite hypoplastic breast tissue. To date, a total of 22 cases associated with breast cancer have been reported in the literature. In this study, we will discuss the case of a 68-year-old female patient who developed breast cancer in the hypoplastic breast on the basis of carcinoma insitu.

Keywords: Breast and endocrine surgery, breast cancer, poland's syndrome

Poland Sendromu ve Meme Kanseri: Vaka Takdimi ve Literatürün Gözden Geçirilmesi

ÖZ

Poland's sendromu konjenital kas iskelet sistemi anomalileri ile karakterize nadir görülen konjenital hastalıktır. Bu hastalarda genellikle pektoralis majör kasının hipoplazisi veya yokluğu, göğüs duvarı ve üst ekstremité anomalileri mevcuttur. Hastaların meme dokusu hipoplazik olmasına rağmen meme kanseri görülebilmektedir. Bugüne kadar literatürde meme kanseri ile ilişkili toplam 22 vaka bildirilmiştir. Bu çalışmada 68 yaşında bayan hastanın hipoplazik meme tarafında karsinoma insitu zemininde gelişen meme kanseri vakasını literatür eşliğinde tartışacağız

Anahtar Kelimeler: Meme Kanseri, Meme Ve Endokrin Cerrahi, Poland's Sendromu

Received: 13.01.2024

Revised: 16.01.2024

Accepted: 02.02.2024

^{*} ORCID: 0000-0002-7068-0895, University of Health Sciences Türkiye, Balıkesir Atatürk City Hospital, Department of Surgical Oncology, Balıkesir, Türkiye

^{**} ORCID: 0009-0001-7494-8537, Balıkesir Atatürk City Hospital, Department of Pathology, Balıkesir, Türkiye

^{***} ORCID: 0000-0001-7411-1705, Balıkesir Atatürk City Hospital, Department of Medical Oncology, Balıkesir, Türkiye

^o Corresponding Author; Ali İmran KÜÇÜK, M.D.

Adress : Balıkesir Atatürk Şehir Hastanesi, Cerrahi Onkoloji Kliniği, Altıeylül-Balıkesir TÜRKİYE

E-mail : drkucuk64@hotmail.com

Tel: +90 5071682648

1. INTRODUCTION

Poland's syndrome is a congenital musculoskeletal developmental anomaly characterized by unilateral chest wall hypoplasia, usually on the right side, and hand anomalies on the same side. General features include absence of the pectoralis major and/or minor muscle, hypoplasia of the breast tissue and ribs, and hand deformities such as syndactyly [1]. Not all features of the syndrome are seen in all patients in the same way. It is also known that the incidence of malignant diseases such as lymphoma, leukemia, neuroblastoma and Wilm's tumor is increased with this syndrome [2]. Poland's syndrome is very rarely associated with breast cancer. In this article, we reviewed a case of breast cancer in a patient with Poland syndrome in the light of the literature.

2. CASE REPORT

A 68-year-old woman was referred to a different tertiary health care institution after a lymph node of approximately 1 cm in the left axilla was detected. Breast USG revealed suspicious lesions in the left breast upper quadrant and malignant-looking lymph nodes in the left axilla. The patient then underwent bilateral dynamic breast MRI. MRI revealed a 10 mm lesion in the upper outer quadrant of the left breast 2 cm from the nipple, 6 mm and 7 mm lesions in the lateral neighborhood of this mass, and a 13 mm lesion approximately 5 cm from the nipple. In addition, lymph nodes of 14 mm in the left axillary region and 13 mm in the infraclavicular region were observed [Figure 1].

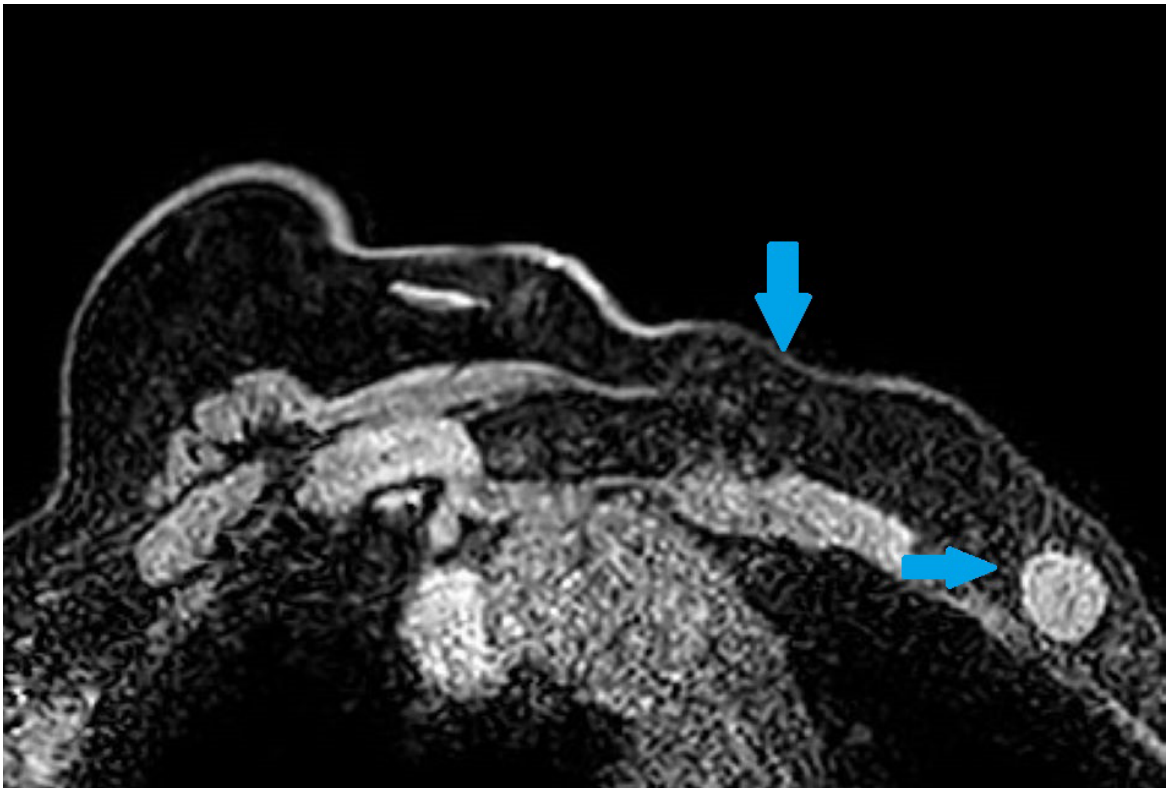


Figure 1. MRI image of chest wall deformity and axillary metastatic lymph node.

A tru-cut biopsy was performed with these findings. The biopsy result was ductal carcinoma insitu (masses at 1 and 3 o'clock), solid and comedo pattern, grade 2. Immunohistochemical study showed estrogen receptor -, Cerb B2 score 3 +, SMA +, P40 + and increased proliferation with Ki-67. Fine needle aspiration biopsy performed from the suspicious lymph node in the left axilla was reported as malignant findings. The patient presented to our outpatient clinic with these findings.

On physical examination, the left breast was hypoplastic and there was deformity in the left costochondral joint. The nipple had a normal appearance. There was no palpable mass in the left breast. Multiple lymph nodes, the largest of which was

approximately 1.5 cm, were seen in the left axilla. PET CT of the patient showed a mass of approximately 15 mm (SUV max: 6.92) in the anterior neighborhood of the left first costa in the upper inner quadrant of the left breast and a mass of approximately 17 mm (SUV max: 4.82) in the upper outer quadrant of the left breast at the level of the axillary tail. In addition, millimeter-sized lymph nodes, the largest of which was 7 mm in diameter, were seen at the level of the left axilla and axillary tail without significant FDG uptake. With these findings, modified radical mastectomy was decided (with the decision of the oncology council). The patient underwent modified radical mastectomy. The pectoralis minor and major muscles and serratus anterior muscle were absent [Figure 2].

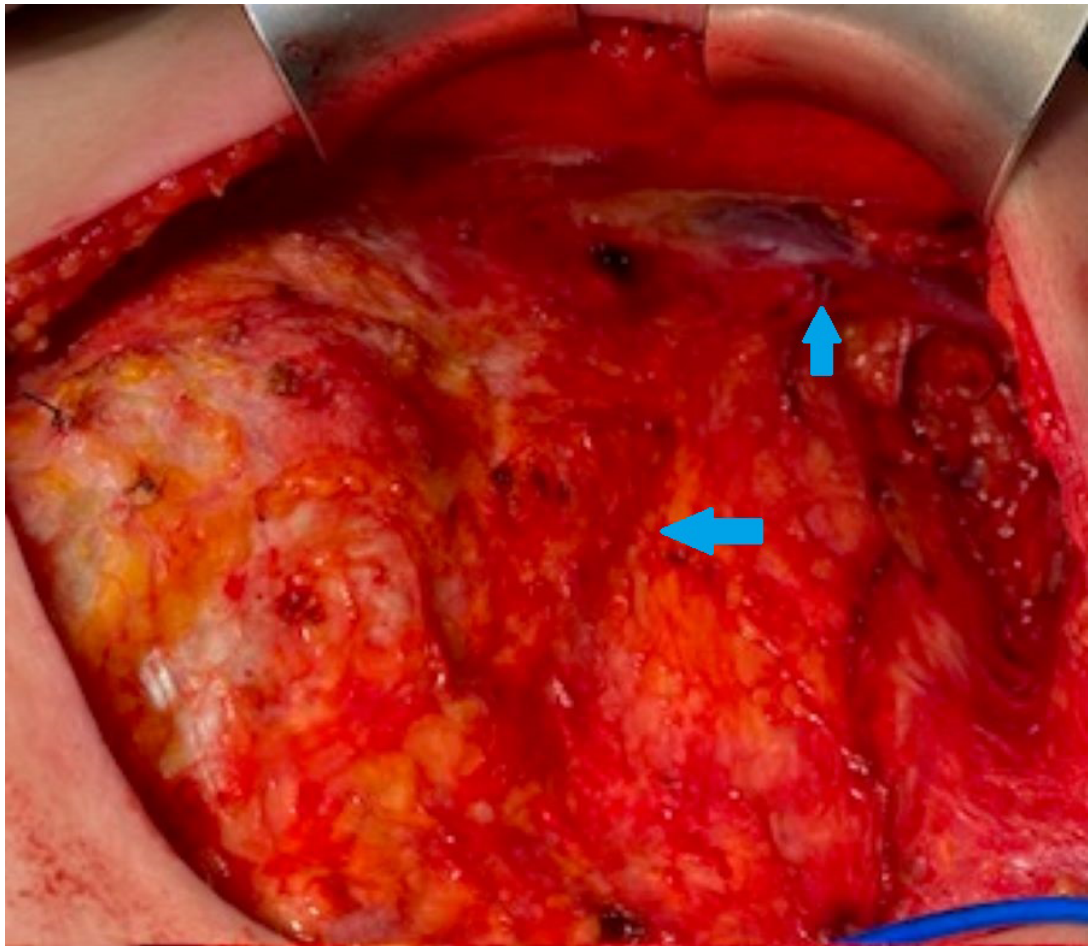


Figure 2. Modified Radical Mastectomy operative image showing the chest deformity without the pectoralis major, pectoralis minor and serratus anterior muscles and the area where the axillary vein enters the chest wall.

There were two conglomerated lumps in the axilla. Postoperative pathology result was invasive ductal carcinoma grade 2 developing on the background of

ductal carcinoma insitu. The tumor diameter was 1.8 cm in the DCIS area with multiple invasive foci, the largest of which was 2 mm [Figure 3].

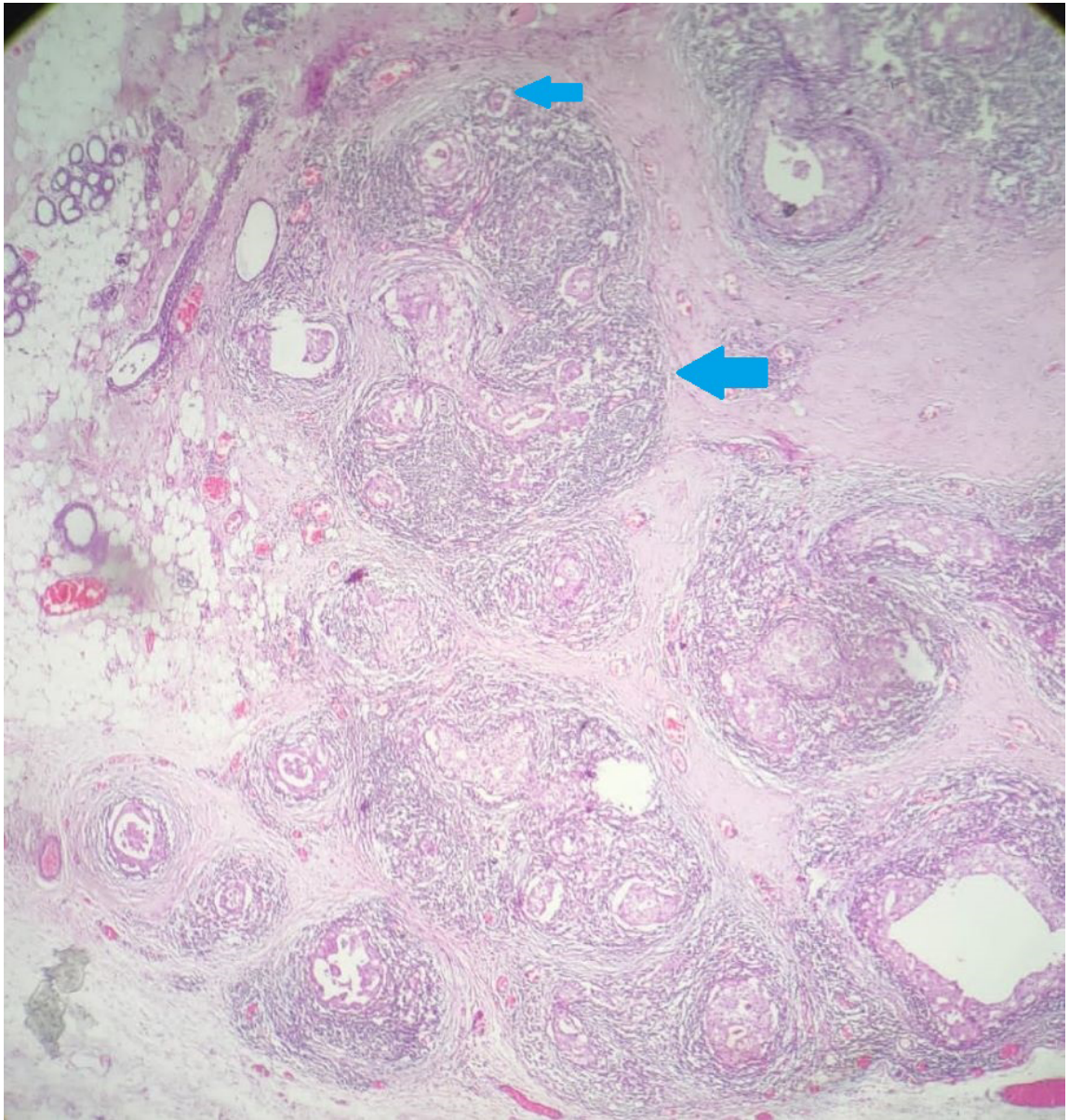


Figure 3. H&E 40X. Invasive Ductal Carcinoma areas developing on the background of Ductal Carcinoma Insitu.

Metastasis was observed in three of the 17 lymph nodes removed. Immunohistochemical findings were ER+ (2%), PR-, CerbB2 +(3), Ki-67 25%, Pan CK+,

E-cadherin + [Figure 4]. The patient was discharged on the third postoperative day without any complication.

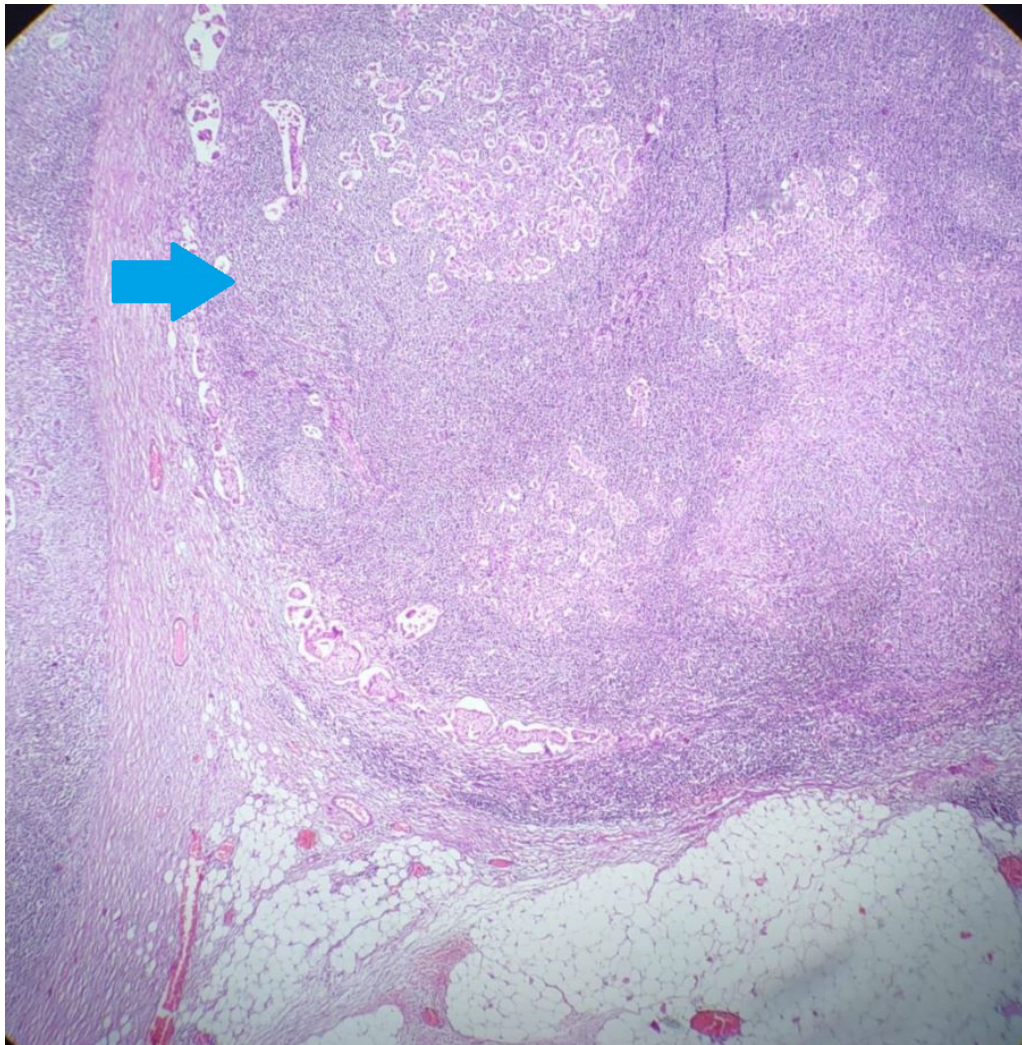


Figure 4. H&E 40X. Pathologic image of lymph node metastasis.

3. DISCUSSION

The incidence of Poland syndrome is approximately 1/20000 and it is more common in males. 70% of the cases occur on the right side of the body [1]. The etiology is not known exactly, but the most important cause is the disruption of embryonic blood flow. Gene defects, trauma, smoking and infections are also blamed [3].

After the first cases reported by Fukushima et al. in 1999, 22 cases have been reported until today [4]. Our case is the 23rd case in the literature. When we analyzed all cases; 60.8% of the cases were seen in the left breast while 39.1% were seen in the right

breast. Only one case was bilateral. While 86.9% of the cases developed on the hypoplastic breast side, approximately 6% of the cases developed on the intact breast side.

Invasive ductal carcinoma (IDC) was seen as the histologic type in almost all cases. Isolated ductal carcinoma insitu (DCIS) was seen in only one case and IDC + ILC (invasive lobular carcinoma) was seen together in one case [5,6]. In our patient, IDC developed on the background of DCIS. When we look at the immunohistochemical features, ER + is seen in 68.4% of cases, while PR + is seen in 61.1% of cases. The rate of Cerb B2+ is 50%. Isolated Cerb B2+ was seen in only one patient [table 1].

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

References	Age	Side	Breast Involved	Histology	Hormon Receptors	Stage
[4]	57	Right	İpsilateral	IDC	ER+PR+	T1N1M0
[4]	53	Left	İpsilateral	IDC	ER-PR-	T2N0M0
[6]	33	Right	İpsilateral	IDC+ILC	ER+PR-	T2N0M0
[7]	42	Left	İpsilateral	IDC	ER-PR+Her2-	T1N0M0
[8]	59	Right	Contralateral	IDC	ER+PR+	T1N1bM0
[9]	71	Right	İpsilateral	IDC+DCIS	ER+PR-Her2-	T1N0M0
[5]	51	Left	İpsilateral	DCIS		TisN0M0
[10]	53	Left	İpsilateral	IDC	ER-PR+Her2-	T1N0M0
[11]	52	Left	İpsilateral	IDC+DCIS		T1N0M0
[12]	46	Right	İpsilateral	IDC	ER+PR+Her2-	T2N1M0
[13]	58	Left	İpsilateral	IDC		T2N0M0
[14]	43	Left	İpsilateral	IDC	ER+Her2-	T3N1M0
[15]	39	Left	İpsilateral	IDC	ER-PR+Her2+	T2N0M0
[16]	43	Left	İpsilateral	IDC	ER+PR+Her2+	T1N3M0
[17]	62	Right	Contralateral	IDC	ER-PR-Her2+	T2N0M0
[17]	69	Left	İpsilateral	IDC	ER+PR-Her2-	T1bN0M0
[17]	37	Right	Bilateral	R:DCIS L:IDC	ER+PR-Her2+ ER-PR-Her2+	T1bN0M0 T1cN0M0
[18]	74	Left	İpsilateral	IDC	ER+PR+Her2-	T3N0M0
[18]	60	Left	İpsilateral	IDC	ER-PR-Her2+	T4N1M0
[19]	56	Left	İpsilateral	IDC	ER+PR+Her2+	T3N0M1
[20]	39	Right	İpsilateral	IDC	ER+PR+Her2+	T1cN0M0
[20]	48	Right	İpsilateral	IDC	ER+PR+Her2-	T4bN0M0
This report	68	Left	İpsilateral	IDC+DCIS	ER+PR-Her2+	T1aN1M0

IDC: İnvazive ductal carcinoma; **ILC:** İnvazive lobuler carcinoma; **DCIS:** Ductal carcinoma in situ;
ER: Estrogen receptor; **PR:** Progesterone receptor.

When we evaluated the clinical features of the cases; Poland syndrome is common in males, while all cases were seen in the female gender. In addition, most of the breast cancer cases developed from the hypoplastic side, but there are cases in which the opposite breast or both breasts were affected. The rate of axillary metastasis is high in the majority of patients with invasive tumors (31.8%). The reason for this may be the hypoplastic breast tissue and easy access to the lymph nodes due to the absence or underdevelopment of anatomical structures. Although the invasive foci were millimetric in size, as in our case, carcinoma metastasis was demonstrated in three lymph nodes.

Although Poland's syndrome is much rarer in women, breast cancer can be seen in syndromic

women, especially on the hypoplastic side. In these patients, care should be taken during breast examination for diagnosis and complications should be avoided during interventional procedures due to the thin breast tissue. In addition, because important anatomical structures are missing or hypoplastic, great care should be taken during the operation and injuries should be avoided.

4. CONCLUSION

Women with Poland's syndrome have a higher risk of breast cancer than the general population. When a patient with Poland's syndrome presents for treatment, imaging modalities may be useful in the evaluation of suspicious lesions. Because of the presence of chest wall deformities and lack of important anatomical

structures, surgeons should be very careful in these patients.

Conflict of Interest:

The authors wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

Author Contributions:

Conceptualization, A.İ.K.; methodology, A.İ.K.; software, A.İ.K.; investigation, A.İ.K., C.E, Yİ; resources, A.İ.K., C.E, Yİ.; data curation, A.İ.K.; writing—original draft preparation, A.İ.K.; writing—review and editing, A.İ.K.; visualization, A.İ.K., C.E.; supervision, A.İ.K.; project administration, A.İ.K. All authors have read and agreed to the published version of the manuscript.

5. REFERENCES

1. Fokin AA, Robicsek F. Poland's syndrome revisited. *Ann Thorac Surg* 2002; 74: 2281-25.
2. Ahn MI, Park SH, Park YH. Poland's syndrome with lung cancer. A case report. *Acta Radiol* 2000;41:432-4.
3. Martinez-Frias ML, Czeizel AE, Rodriguez-Pinilla E, Bermejo E. Smoking during pregnancy and Poland sequence: results of a population- based registry and a case-control registry. *Teratology* 1999;59:35-8.
4. Fukushima T, Otake T, Yshima R, et al. Breast cancer in two patients with Poland's syndrome. *Breast Cancer* 1999;6:127-30.
5. Wong TC, Lim J, Lim TC. A case of ductal carcinoma in situ of breast with Poland syndrome. *Ann Acad Med Singapore* 2004;33:382-4.
6. Havlik RJ, Sian KU, Wagner JD, Binford R, Broadie TA. Breast cancer in Poland syndrome. *Plast Reconstr Surg* 1999;104:180-2.
7. Katz SC, Hazen A, Colen SR, Roses DF. Poland's syndrome and carcinoma of the breast: a case report. *Breast J* 2001;7:56-9.
8. Okamo H, Miura K, Yamane T, Fujii H, Matsumoto Y. Invasive ductal carcinoma of the breast associated with Poland's syndrome: report of a case. *Surg Today* 2002;32:257-60.
9. Khandelwal A, O'Hea BJ, Garguilo G. Breast cancer in a patient with Poland's syndrome. *Am Surg* 2004;70:491-5.
10. Tamiolakis D, Venizelos D, Antoniou C, Tsiminikakis N, Alifieris E, Papadopoulos N. Breast cancer development in a female with Poland's syndrome. *Onkologie* 2004;27:569-71.
11. Salhab M, Al Sarakbi W, Perry N, Mokbel K. Pneumothorax after a clinical breast fine-needle aspiration of a lump in a patient with Poland's syndrome. *Int Semin Surg Oncol* 2005;2:14.
12. Wang X, Ning L. : Breast carcinoma associated with Poland's syndrome: One case report and literatures review . *Clin Oncol Cancer Res.* 2008, 5:223-5.
13. Ji J, Zhang S, Shao C, Xu M, Chen S, Lu C, et al: Poland's syndrome complicated with breast cancer: Mammographic, ultrasonographic, and computed tomographic findings . *Acta Radiol.* 2008, 49:387-90.
14. Causa L, Kirova YM, Campana F, Fourchette V, Salmon RJ.: Poland syndrome, breast cancer: The importance of the radiotherapy technique after mastectomy *Radiother Oncol.* 2009;91:138-9. *Radiother Oncol.* 2009, 91:138-9.
15. Yesilkaya Y, Dizdar O, Altundag K. : Ipsilateral breast cancer in a patient with Poland's syndrome *Am Surg.* 2011;77:234-5. *Am Surg.* 2011, 77:234-5.
16. Zhang F, Qi X, Xu Y, Zhou Y, Zhang Y, Fan L, et al.: Breast cancer and Poland's syndrome: A case report and literature review *Breast J.* 2011;17:196-200. *Breast J.* 2011, 17:196-200.

17. DeFazio MV, Dervishaj OA, Bozzuto LM 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.
18. Huang Y, Pang H, Jin S, Han X, Liu X, Yang L, Zuo S, Li J, Meng D.: Clinical characteristics of Poland's syndrome associated with breast cancer: Two case reports and a literature review. *J Cancer Res Ther.* . 2018, 14(7):1665-1669.
19. Malatay Gonzalez C, Apolo Carrion C, Wong-Achi X.: Breast cancer associated with Poland syndrome. *Revista de Senología y Patología Mamaria - Journal of Breast Science* . 2022, 35:130-133.
20. Cruz Cuadrado C, Fernandez-Pacheco BAC, Hernandez AAT, Benitez VV, Hernandez JR.: Breast cancer and Poland's syndrome. *Cirugía Española* . 2023, 101:228-231.