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# Sarcomas and malignant phyllodes tumours of the breast – A retrospective study

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ABSTRACT

*Background:* Although most breast cancers are adenocarcinomas of the mammary gland, primary breast sarcomas may also arise from mammary gland mesenchymal tissue. The annual incidence of primary breast sarcoma is low and has been estimated at 45 new cases per 10 million women. These tumours are at high risk of recurrence and are known to have poor prognosis. Phyllodes tumours represent a specific subset of these breast soft tissue tumours. They are composed of a connective tissue stroma and epithelial elements. Pathological presentation ranges from grade I to malignant phyllodes tumours (grade III) where the stromal component clearly exhibits a sarcoma pattern.

*Materials and Methods:* SAPHYR (SArcoma and PHYllode Retrospective) is a retrospective study of the experience of Leon Bérard Cancer Centre (Lyon, France) from 1966 to August 2004. SAPHYR aims to describe the characteristics of primary breast sarcomas and to define potential survival factors to be evaluated in future prospective studies.

*Results:* We included 70 patients. Half of them presented at least one recurrence (35/70). Median disease-free-survival (DFS) was 1.15 years. At 3 years, median overall survival had not been reached and more than 61% of the patients were alive. Quality of surgical resection was significantly ($p = 0.036$) different whether patients were in the R0 group (72%) or not (38%). No survival difference was found between malignant phyllodes (grade III) and other primary breast sarcomas (angiosarcomas excluded). Histology revealed three significantly ($p = 0.0003$) different prognostic groups: phyllodes grade I and II (DFS = 57%), angiosarcomas (DFS = 7%) and phyllodes grade III and other primary breast sarcomas (DFS = 45%).

*Discussion:* Phyllodes tumours and primary breast sarcomas are totally different from epithelial breast cancers and should be considered as a distinct group of rare tumours. The first goal of treatment is to achieve negative margins (R0). We propose to treat the patients according to the clinical practice guidelines in use for soft tissue sarcomas and address them to a reference centre for sarcoma. Treating rare tumours in the same place should permit us to standardise pathological data and to include patients into multicentric radiotherapy or chemotherapy protocols to improve overall survival. As further prospective studies are needed, European oncology groups must join their forces to create a prospective Rare Cancer Network.



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## 1. Introduction

Although the majority of breast malignancies are adenocarcinomas developed on the mammary gland, primary breast sarcomas (PBS) may also arise from the mesenchymal tissue of the mammary gland. The variety of cells present in this tissue, such as fat cells, muscle cells, or endothelial cells, explains the heterogeneity of the histological types encountered: liposarcomas, angiosarcomas, and leïomyosarcomas. Annual incidence can be estimated at 44.8 new cases per 10 million women, and this rate remained constant over the 1973 to 1986 period.[1] Malignant phyllodes tumours represent a specific subset of breast sarcomas composed of epithelial elements with a connective tissue stroma. Malignant phyllodes tumours, initially defined according to their fleshy macroscopic appearance, encompass a wide variety of tumours with mixed epithelial and benign or malignant mesenchymal proliferation. Pathological presentation ranges from grade I to malignant phyllodes tumours (grade III) where the stromal component displays a clear pattern of sarcoma. These tumours are characterised by a high risk of recurrence and are associated with evolution towards more aggressive disease.[2]

Whatever the grade of the phyllodes tumour, the goal of surgery is to perform complete resection with safe margins. Because there is strong evidence that phyllodes tumours can be considered malignant when the stromal component exhibits a clear pattern of sarcoma, we considered that primary breast sarcoma (PBS) and malignant phyllodes (grade III) tumours should be treated with the same strategies and rules.[3,4]

As primary breast sarcoma is very rare, most published papers are reports of isolated cases that do not provide an objective view of the disease. There is no definitive consensus regarding the treatment of PBS, and even if simple mastectomy without axillary dissection is still widely regarded as a gold standard, there are major variations in the extent of local therapy, ranging from wide local excision to radical mastectomy. To improve the management of primary breast sarcoma patients, we first need to collect and observe past cases in order to define critical points of management before starting a prospective multicentric national study. We present here the SAPHYR study (SArcoma and PHYllode Retrospective), a retrospective study of the experience of Leon Bérard Cancer Centre (Lyon, France) from 1966 to August 2004. This study aims to describe the characteristics of primary breast sarcomas and to define potential survival factors to be evaluated in future prospective studies.

## 2. Materials and methods

### 2.1. Patients and database

Patients with primary breast sarcomas and phyllodes tumours treated at Leon Bérard Cancer Centre (regional comprehensive cancer centre of Lyon, Rhône Alpes region, France) between 1966 and 2004 were retrospectively analysed. Dermatofibrosarcoma protuberans and metaplastic carcinomas were excluded from the study.

A database (Access, Microsoft®) was specifically created for this retrospective study. Clinical data, including age, personal breast cancer history, pathological (histological subtype of conjunctive tumour, initial staging, and local symptoms) and treatment data (surgery, chemotherapy, hormone therapy and/or radiotherapy) were retrospectively collected from medical records. Concerning margins, when information was missing, we chose to assign patients the worst score, R2. This concerned ten mastectomies (14%) and seven wide excisions (10%).

In the future, this database will be used prospectively for systematically collecting new cases and integrating them in the French phyllodes and primary breast sarcoma network.

### 2.2. Statistical analysis

Statistical analyses were performed by SPSS software®, version 11.0, using Pearson chi-square test or Fisher's exact test when appropriate and Kaplan–Meier method for survival analysis.

Follow-up was calculated from the date of diagnosis to the date of last news. Survival time was defined as the time interval from the date of PBS to the date of death or last follow-up. For survival data, as it is a retrospective study, and as the follow-up varies with the patients, we used the most conservative strategy by excluding, at each time point (1 year, 3 years and 5 years), patients whose follow-up was shorter than the time point and whose last status was 'alive'. Disease-free survival (DFS) was calculated from the date of diagnosis to the date of the first sign of relapse. Survival curves were generated using Kaplan–Meier estimates and compared using the log rank test.

Primary outcomes were 3-year overall and disease-free survival rates.

## 3. Results

### 3.1. Patient characteristics at time of PBS diagnosis

Overall, 70 patients constituted the study group; they are summarised in Table 1. Median age at diagnosis of primary carcinoma was 48.6 years (range 39.7–60.1) with 75.7% of the patients younger than 60 years. Only 9% (6/70) of the patients were metastatic at diagnosis. There were 48.6% of right-sided sarcomas. Nineteen patients (27.1%) had a history of previous benign breast disease notified as cyst, adenofibroma or mastosis. Seventeen patients (24.3%) had a personal history of breast cancer and eight had undergone homolateral adjuvant radiotherapy previously to the current sarcoma (considered as radiation-induced). For these patients with a history of breast cancer, the mean time between previous radiotherapy and sarcoma diagnosis was 7.89 years (± 3.4).

### 3.2. Pathological data

Tumour size was described by the pathologist for 56 patients (80%); most tumours were voluminous since 88% (49/56) measured at least 2 cm and 37.5% were larger than 5 cm (Table 1). Median tumour size was 47.5 mm (range 10 to 210 mm). In most of the cases, only one tumour was seen (61/71, 86%). A mitosis count was performed for 50 patients (71%) and yielded quite homogenous results (48% had up to ten mitoses per field, 34% had less than five mitoses per field). A vast majority (67%) of the tumours presented severe cellular atypia. Unfortunately, data for vascular invasion and stromal overgrowth were missing for the majority of patients.

**Table 1 – Population characteristics**

| | |
|---|---|
| **Age at diagnosis** | |
| Mean(yr)[min-max] | 49.7 ± 15.5 [13.1–84.4] |
| < 40 yrs | 17/70 (24%) |
| < 60 yrs | 53/70 (76%) |
| **Right sided tumour** | 34/70 (49%) |
| **Initial extension** | |
| Localized | 55/70 (79%) |
| Metastatic at diagnosis | 6/70 (9%) |
| Unknown | 9/70 (13%) |
| **Personal history of breast cancer** | **17/70 (24%)** |
| With homolateral irradiation to current sarcoma | 8/16 (50%) |
| Time between R × T and sarcoma (mean ± SD) | 7.89 yrs ± 3.4 |
| **Histological type** | |
| *Phyllodes* | **26/70 (37%)** |
| Grade I | 5/70 (7%) |
| Grade II | 5/70 (7%) |
| Grade III | 16/70 (23%) |
| *Primary breast sarcoma* | **44/70 (63%)** |
| Angiosarcoma | 17/70 (24%) |
| Spindle cell sarcoma | 6/70 (9%) |
| Liposarcoma | 3/70 (4%) |
| Fibrosarcoma | 3/70 (4%) |
| Osteosarcoma | 2/70 (3%) |
| Leiomysarcoma | 2/70 (3%) |
| Rhabdomyosarcoms | 1/70(1%) |
| Mixte and others | 10/70 (14%) |
| **Histological tumour size** | |
| Median [min-max] (mm) | 47.50 [10-210] |
| > 50 mm | 21/56 (37.5%) |
| **Mitotic Index** | |
| < 5 | 17/50 (34%) |
| 5-10 | 9/50 (18%) |
| > 10 | 24/50 (48%) |
| **Cellular atypia** | |
| Absent | 11/47 (16%) |
| Mild | 7/47 (26%) |
| Severe | 29/47 (67%) |

The main histological subtype was phyllodes tumour, with 26 patients (37%), and could be delineated into five grade I (7%), five grade II (7%) and 16 grade III (23%). The second group was angiosarcoma, corresponding to 24% of the tumours ($n = 17$), and the third one was spindle cell sarcoma (9%, $n = 6$). Other histologies were marginal as shown in Table 1. Phyllodes tumours grade III and sarcomas represented 86% of the total population.

Axillary dissection was performed in 26 patients (37%). Node invasion was found in two of them (8%). Histology for the two patients with axillary nodes involvement was malignant phyllode and primary breast sarcoma.

### 3.3. *Treatment*

#### 3.3.1. *Surgery*

All patients but one underwent surgery as primary treatment (Table 2). Only one refused surgery. There were primarily 38 mastectomies (55%) and 29 wide excisions (42%). Two interventions were not specified. Among wide excisions, there were 12 re-excisions consisting of eight mastectomies and four secondary wide excisions. So, the final surgical treatment consisted of 21 wide excisions (30%) and 46 mastectomies (66%). At the end of surgical care, quality of resection was assessed as a function of margins. Most were R0 (54%), 19% were R1 and 26% were R2.

#### 3.3.2. *Adjuvant treatment*

Radiotherapy was only delivered to twenty-three patients (33%) in addition to wide excision (5/21; 24%) or mastectomy (18/46; 39%). Chemotherapy was used in seventeen patients (24.3%): fifteen received adjuvant chemotherapy and two received palliative treatment. Eleven of these 17 patients (65%) received an association of three drugs based on anthracycline and alkylating agents and three had a bi-therapy (anthracycline + alkylating agent or an association of two alkylating agents). Only two (12%) were treated by monotherapy (doxorubicin or bisantrene) and one had four drugs by adding VP16 and carboplatin to anthracycline and alkylating agent (Table 3b). The main drug association used was a combination of an anthracycline and an alkylating agent (ifosfamide) (10/17; 59%). At the end of first line treatment, response was assessed in all but six patients: 60 achieved a complete response (86%) and four had a partial response.

### 3.4. *Follow-up*

In these seventy patients, the median follow-up was 31.7 months. A vast majority of the patients were followed between 1 and 6 years. Two had a very long follow-up: one for 24 years and one for 15 years and a half. These two particular cases with a very long follow-up were phyllodes tumours grade III. The first one was R0 after surgery whereas

**Table 2 – Initial treatment**

| | |
|---|---|
| **Surgery** | |
| Wide excision | 21/70 (30%) |
| Mastectomy | 46/70 (66%) |
| Unknown | 2/70 (3%) |
| No surgery | 1/70 (1%) |
| **Axillary dissection** | 26/70 (37%) |
| Invaded nodes | 2/26 (8%) |
| **Final margins** | |
| R0 | 38/70 (54%) |
| R1 | 13/70 (19%) |
| R2 | 18/70 (26%) |
| **Adjuvant radiotherapy** | 23/70 (33%) |
| Post wide excision | 5/21 (24%) |
| Post mastectomy | 18/46 (39%) |
| **Chemotherapy** | 17/70 (24%) |
| Adjuvant | 15/70 (21%) |
| Association chemotherapy + radiotherapy | 10/15 (67%) |
| Palliative | 2/70 (3%) |
| **Initial response assessment** | |
| RC | 60/70 (86%) |
| RP | 4/70 (6%) |
| NE | 6/70 (9%) |

**Table 3 – Overall survivals at 1 year and 3 years in the SAPHYR study**

| | Survival at 1 year | | Survival at 3 years | |
|---|---|---|---|---|
| | Rate (%) | Significant | Rate (%) | Significant |
| Overall survival | 86.9% ($n = 61$) | | 61.5% ($n = 52$) | |
| < 60 years | 65.9% ($n = 47$) | $p = 0.09$ | 60.9% ($n = 41$) | $p = 0.019$ |
| > 60 years | 42.9% ($n = 14$) | | 27% ($n = 11$) | |
| R0 | 76.9% ($n = 30$) | $p = 0.025$ | 72% ($n = 25$) | $p = 0.036$ |
| R1 + R2 | 46.7% ($n = 30$) | | 38.5% ($n = 26$) | |
| < 50 mm | 62.1% ($n = 29$) | $P = 0.56$ | 57.7% ($n = 26$) | $P = 0.27$ |
| > 50 mm | 57.9% ($n = 19$) | | 46.7% ($n = 15$) | |
| Malignant phyllodes | 57.1% ($n = 14$) | $p = 0.6$ | 53.8% ($n = 13$) | $p = 0.8$ |
| Other primary breast sarcomas (PBS) | 68% ($n = 25$) | | 60% ($n = 20$) | |
| Phyllodes grade I & II | 100% ($n = 8$) | | 100% ($n = 6$) | |
| Malignant phyllodes + PBS | 64.1% ($n = 39$) | $P = 0.002$ | 57.6% ($n = 33$) | $P = 0.001$ |
| Angiosarcomas | 28.7% ($n = 14$) | $P = 0.013$ | 23.1% ($n = 13$) | $P = 0.005$ |

Mean survival at 3 years: 2.35 (2.09–2.62).

the resection quality in the second one was rated R2 in our analyses.

### 3.5. *Recurrence*

Half of the patients presented at least one recurrence (35/70). Median time to recurrence was 8.4 months, ranging from 1.33 to 85.27 months. Among the 35 patients who experienced relapse, 22 (63%) relapsed during the first 12 months and 89% during the first 2 years. Only two patients underwent a very late recurrence: one at 6 years and one at 7 years. Twenty-two patients relapsed locally whereas 11 had systemic recurrence. Two had distant and local relapses at the same time.

Risk of recurrence did not correlate to type of surgery: 14 relapses in the 29 patients with tumorectomy and 20 relapses in the 38 patients with mastectomy ($p = 0.72$). There is no significant difference either in recurrence risk whether patients have received radiotherapy or not (54.2% recurred after radiotherapy versus 47.8% radiotherapy; $p = 0.6$). Adjuvant radiotherapy was not associated with a significantly reduced risk of local recurrence.

The risk to have a subsequent relapse was 63% (22/35) after the first recurrence, rising to 68% (15/22) after the second one.

### 3.6. *Overall survival*

At 3 years (Table 3), median overall survival was not reached and more than 61% of the patients were still alive (mean survival time of 2.35 years (2.09–2.62 95% confidence interval)). The 11 patients (21%) over 60 years old represented a poor prognosis group with an overall survival rate of only 27% at 3 years, compared to 61% in the younger group ($n = 41$) (Table 3) ($p = 0.019$). Regarding quality of resection (Table 3), there was a difference in survival whether the patient was in the R0 group (survival rate = 72%) or not (survival rate for groups R1 + R2 = 38.46%). This difference was statistically significant ($p = 0.036$). Even if there was a trend in favour of the group presenting tumours measuring 5 cm or smaller (survival rate 57.7% versus 46.7%), the difference was not statistically significant. No other significant factor was found to correlate with survival. Considering final surgical strategy, no difference could be seen between wide resection and mastectomy ($p = 0.2$). Histological subtype analysis showed no difference between the survival rate of malignant phyllodes ($n = 13$) and other primary breast sarcomas ($n = 20$) with a survival rate at 3 years of 54% and 60% ($p = 0.8$), respectively. However, the survival rate of angiosarcomas (23%) strongly differed from that of other sarcomas, including malignant phyllodes (57.6%) ($p = 0.005$). Finally, the tiny group of phyllodes grades I and II ($n = 6/52$) represented a significantly good prognosis subgroup with 100% survival rate.

At 1 year, reliable information was available for 61 patients. The overall survival rate was 87% (Table 1). The following poor survival prognostic factors were found significant: positive resection margins (microscopic or macroscopic) and angiosarcoma histology. There was no significant difference in survival rate between malignant phyllodes and other primary breast sarcomas excluding angiosarcomas. Phyllodes grades I and II formed a group with good prognosis. Concerning tumour size and patient age, we did not find any significant difference between the groups.

At 5 years, the overall survival rate was 49%.

### 3.7. *Disease free-survival (DFS)*

DFS rate was 61% at 1 year, 37% at 3 years and 28% at 5 years (Fig. 1). Median DFS was 1.15 years (range 0.84–1.84). As previously shown for overall survival, patients older than 60 years (Fig. 2) had a significantly lower 3-year DFS rate (14.3%) than younger patients (46.3%) ($p = 0.004$). Concerning tumour size and resection margins, there was no significant difference ($p = 0.09$) between groups. This is probably due to our very conservative strategy of analysis which assigned the score of R2 by default when the quality of resection was not specified.

Fig. 3 shows that malignant phyllodes and primary breast sarcomas (angiosarcomas excluded) have no statistically significant difference ($p = 0.51$) in DFS and present the same

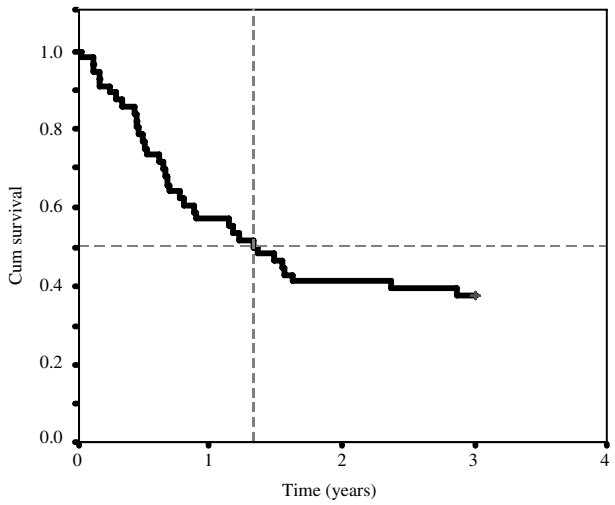


**Fig. 1 – Disease-free survival at 3 years in the SAPHYR study.**

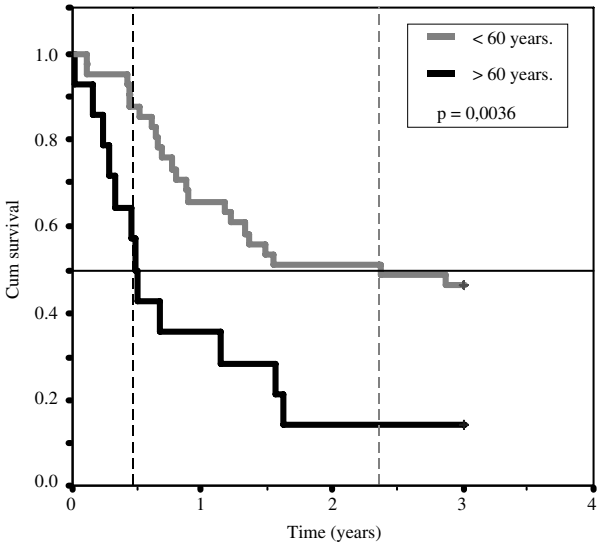


**Fig. 2 – Comparison of 3-year disease-free survival curves between age groups (±60 years).**

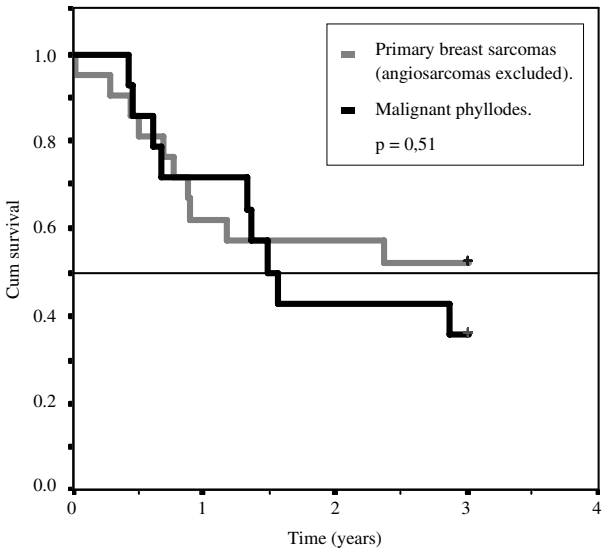


**Fig. 3 – Comparison of 3-year disease-free survival curves between histology groups: malignant phyllodes versus primary breast sarcomas (angiosarcomas excluded).**

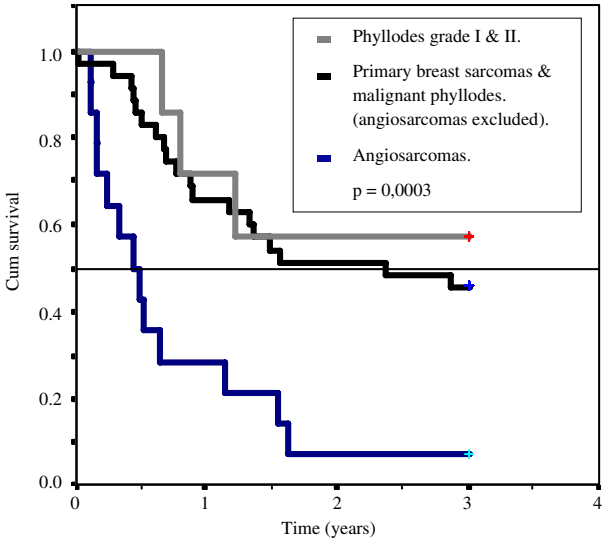


**Fig. 4 – Comparison of 3-year disease-free survival curves between three different histology groups: angiosarcomas, phyllodes grade I/II and primary breast sarcomas plus malignant phyllodes.**

behaviour at 3 years. Histology (Fig. 4) distinguished three groups with a significantly different behaviour ($p = 0.0003$): angiosarcomas ($n = 14$) with a very low 3–year DFS rate (7.1%), a second group ($n = 35$) of malignant phyllodes and other primary breast sarcomas (45.7% 3-year DFS rate) and, finally, grade I/II phyllodes tumours ($n = 7$) with the highest 3–year DFS rate (57.1%).

## 4. Discussion

This study of phyllodes tumours and primary breast sarcomas deals with a very rare disease considered to represent only 0.3 to 1.0% of all mammary tumours and 2 to 3% of fibroepithelial neoplasm of the breast.[5] Even large and famous cancer centres lack experience in managing these tumours and we have difficulties in proposing a specific treatment for these patients. Furthermore, the few studies published in the literature report a prognosis that varies from 14% at 5 years to more than 90%. More than a methodological defect of these studies, this prognosis disparity certainly reflects different subgroups. It is of major interest to identify survival prognosis factors for these tumours. The rarity of PBS precludes any prospective study, and our conclusions must be considered with caution given the limitations inherent to retrospective analyses. First, there may be a referral bias because the patients were treated in a major comprehensive cancer centre. Second, there is still no consensus regarding adjuvant chemotherapy and even radiotherapy or the extent of surgery in PBS, and the specific indications were not always discernible in this series. Another limitation of the study is that it is

retrospective and it has included patients over a very long period, nearly forty years. Obviously, during this period, medical care has progressed, medical practices have evolved and medical staff have changed. Besides, missing data have reduced the reliability of information and the power of the study.

In this study, we have identified some characteristics that distinguish primary breast sarcoma from common epithelial breast cancer. Interestingly, primary breast sarcoma may affect the whole female population from teenagers to elderly women, but the mean age at diagnosis (49.7 years) is younger than for common epithelial breast cancer. The size of the tumour is larger: median 4.7 cm. Node involvement, even for these large tumours, is quite uncommon (7.7%) but it does happen.

In our series of 70 patients, the overall survival rate was 87% at 1 year, 62% at 3 years and 49% at 5 years. Prognostic factors for survival concerned age, medical practices, histological subtype and possibly tumour size. The results of this study confirmed the current opinion that malignant phyllodes and primary breast sarcomas, excluding angiosarcomas, have the same prognosis and should be considered together.[3,4] As reported by Zeleck et al. in 2003, angiosarcomas represented a very pejorative survival prognosis factor.[3] However, no molecular analysis was delineated in this retrospective analysis. It will be fundamental for future studies to explain the difference between the survival of patients with angiosarcoma and other subgroups, as well as their response to treatment. This study seems to confirm the prognosis similarity between malignant phyllodes tumours and primary breast sarcomas (except angiosarcomas), which should further be confirmed by a prospective study including molecular analysis in the phyllodes network program.

Studies by Joshi et al. and Pandey et al.[6,7] have reported a benefit from adjuvant radiotherapy in patients exhibiting poor prognostic features. We failed to show such a benefit and to prevent local recurrence using adjuvant radiotherapy. We think that, at the moment, it is not clear whether this benefit exists and further prospective studies are needed before recommending this treatment strategy.[8]

Due to the long retrospective period of the study, tumour heterogeneity and missing pathology data, we were not able to show significant prognostic recurrence factors, contrary to Eroglu et al.[9] who identified tumour necrosis, stromal overgrowth, number of mitoses, stromal cellularity and atypia. In their study, Asoglu et al.[10] only retained stromal overgrowth as relevant.

As in our series, Blanchard et al. reported some (10%) axillary node involvements.[11] Our findings are in agreement with reports of Blanchard et al.[11] and Shabahang et al.[12] that prognosis is not influenced by adjuvant radiotherapy or chemotherapy but by the ability to achieve complete microscopic resection (R0) whatever the surgical technique or the tumour size. To influence survival, the opinion that the 'gold standard' is to achieve negative margins (R0) is shared by some other authors like Trent II et al.[13] or Khan and Badve.[14] It is also supported by other published studies.[7,8,15–18] Nevertheless, some authors like Gutman et al. propose a more aggressive approach for tumours 5 cm or larger by using neoadjuvant chemo radiation to improve local control. Unfortunately, in their study, chemotherapy and/or radiation only prolonged DFS but not overall survival. Secondly, there was only a trend toward improved local control using adjuvant radiotherapy.[19]

In conclusion, the therapeutic management of phyllodes tumours and primary breast sarcomas is a crucial problem because of their rarity and also of the specific treatment strategy that must be followed. The disease is usually diagnosed and initially treated by gynaecology surgeons. However, phyllodes tumours and primary breast sarcomas are totally different from epithelial breast cancers and require a different management. They should be considered as sarcomas. Thus the strategy logically proposed for these rare tumours is to treat them according to the 'clinical practice guidelines for soft tissue sarcoma and osteosarcoma'.[20,21] The main direction is to address these patients to a sarcoma reference centre where a multidisciplinary medical team will coordinate local evaluation by MRI and distant evaluation by chest radiograph and chest tomodensitometry before proposing the best surgical strategy permitting to achieve negative margins (R0). Furthermore, referring rare tumours to the same reference centres allows the standardisation of pathologic data and the inclusion of patients in multicentric protocols of radiotherapy or chemotherapy. This attitude is certainly appropriate to give our patients the best chances of survival and to progress in the knowledge of prognostic factors and therapeutic strategies. Previous studies of prognostic factors and overall survival in sarcoma also described multidisciplinary as a potent prognostic factor.[20–23] As further prospective studies are needed, European oncology groups will have to join their forces and create a prospective Cancer Network on these rare diseases.

## Conflict of interest statement

None declared.

## REFERENCES


1. May DS, Stroup NE. The incidence of sarcomas of the breast among women in the United-States, 1973-1986. *Plast Reconstr Surg*. 1991(87):193–4.
2. Tavassoli FA, Devilee P. *Pathology and genetics of tumours of the breast and female genitale organs. World Health Organization Classification of tumours*. IARC Press Lyon; 2003.
3. Zelek L, Llombart-Cussac A, Terrier P, et al. Prognostic factors in primary breast sarcomas: a series of patients with long-term follow-up. *J Clin Oncol* 2003;**21**(13):2583–8.
4. Terrier P, Terrier-Lacombe MJ, Mouriesse H, Friedman S, Spielmann M, Contesso G. Primary breast sarcoma: a review of 33 cases with immunohistochemistry and prognostic factors. *Breast Cancer Res Treat* 1989;**13**(1):39–48.
5. Liberman L et al. Benign and malignant phyllodes tumours. Mammographic and sonographic findings. *Radiology* 1996;**198**:121.
6. Joshi SC, Sharma DN, Bahadur AK, Maurya R, Kumar S, Khurana N. Cystosarcoma phyllodes: our institutional experience. *Australas Radiol* 2003;**47**(4):434–7.
7. Pandey M, Mathew A, Kattoor J, et al. Malignant phyllodes tumor. *Breast J* 2001;**7**(6):411–6.

8. Barrow BJ, Janjan NA, Gutman H, et al. Role of radiotherapy in sarcoma of the breast—a retrospective review of the M.D. Anderson experience. *Radiother Oncol* 1999;**52**(2):173–8.
9. Eroglu E, Irkkan C, Ozsoy M, Eroglu F. Phyllodes tumor of the breast: case series of 40 patients. *Eur J Gynaecol Oncol* 2004;**25**(1):123–5.
10. Asoglu O, Ugurlu MM, Blanchard K, et al. Risk factors for recurrence and death after primary surgical treatment of malignant phyllodes tumours. *Ann Surg Oncol* 2004;**11**(11):1011–7.
11. Blanchard DK, Reynolds CA, Grant CS, Donohue JH. Primary nonphylloides breast sarcomas. *Am J Surg* 2003;**186**(4):359–61.
12. Shabahang M, Franceschi D, Sundaram M, et al. Surgical management of primary breast sarcoma. *Am Surg* 2002;**68**(8):673–7. discussion 677.
13. Trent II JC, Benjamin RS, Valero V. Primary soft tissue sarcoma of the breast. *Curr Treat Options Oncol* 2001;**2**(2):169–76.
14. Khan SA, Badve S. Phyllodes tumours of the breast. *Curr Treat Options Oncol* 2001;**2**(2):139–47.
15. Pandey M, Mathew A, Abraham EK, Rajan B. Primary sarcoma of the breast. *J Surg Oncol* 2004;**87**(3):121–5.
16. Soumarova R, Seneklova Z, Horova H, et al. Retrospective analysis of 25 women with malignant cystosarcoma phyllodes–treatment results. *Arch Gynecol Obstet* 2004;**269**(4):278–81.
17. Berg JW, Decrosse JJ, Fracchia AA, Farrow J. Stromal sarcomas of the breast. A unified approach to connective tissue sarcomas other than cystosarcoma phyllodes. *Cancer* 1962;**15**:418–24.
18. Chaney AW, Pollack A, McNeese MD, et al. Primary treatment of cystosarcoma phyllodes of the breast. *Cancer* 2000;**89**(7):1502–11.
19. Gutman H, Pollock RE, Ross MI, et al. Sarcoma of the breast: implications for extent of therapy. *The M.D. Anderson experience. Surgery* 1994;**116**(3):505–9.
20. Philip T et al. Standards, options and recommendations (SOR) for diagnosis, treatment and follow-up of osteosarcoma. Groupe de travail SOR. *Bull Cancer* 86.2 1999:159–76.
21. Pisters PW et al. Analysis of prognostic factors in 1,041 patients with localized soft tissue sarcomas of the extremities. *J Clin Oncol* 14.5 1996:1679–89.
22. Ray-Coquard I, Thiesse P, Ranchere-vince D, et al. Conformity to clinical practice guidelines, multidisciplinary management and outcome of treatment for soft tissue sarcomas. *Ann Oncol* 2004;**15**(2):307–15.
23. Wiklund T. The importance of a multidisciplinary group in the treatment of soft tissue sarcomas. *Eur J Cancer* 32A.2 1996:269–73.