Mantle cell lymphoma (MCL) represents 4% of lymphomas in the United States and 7%-9% in Europe. This lymphoma is diagnosed in patients with an average age of 60, and most frequently affect males (ratio 2:1). In spite of its extralymphatic involvement (digestive tract, stomach, colon, liver, skin, lacrimal glands and central nervous system), MCL located in the breasts is uncommon. We present the clinical case of a male patient with MCL of the breast, in whom surgery provided adequate local control.

The patient is a 56-year-old man diagnosed with stage IIA blastoid MCL, diagnosed by cervical and right axillary lymphadenopathies. He was initially treated with radiotherapy of the affected field (total dose 47 Gy) and rituximab (4 doses), as he refused treatment with polychemotherapy. Complete remission was reached, but 2 months after finalizing treatment he presented a relapse with supradiaphragmatic lymphadenopathies and a retroareolar mass in the right breast, with a biopsy that was compatible with blastoid MCL (Ki-67: 75%). For this reason, a second line of treatment was initiated with alternating R-CHOP/R-DHAP chemotherapy (4 cycles). When the response was evaluated after one month, evidence of partial response was observed, and PET/CT showed persistence of the right retroareolar mass (Fig. 1).

Given the partial response to treatment and due to the solitary location of the lymphoma in the right breast, we decided to resect the mass. Using a Stewart incision, we found a mass causing nipple retraction and conducted a simple mastectomy. The pathology study reported a mass measuring 4 × 2.5 × 3.7 cm, proximal to the deep resection margin, identified as an MCL (aggressive or blastoid), with 20–22 mitoses per 10 high-power fields. In the immunohistochemistry analysis, the mass expressed CD20, CD79a, Bcl-2, PAX5 and CD5 with intense cyclin D1 overexpression and showed lambda light chain restriction; the proliferative index (Ki-67) was 70%, and a moderate expression of p53 was identified.

After another evaluation of the disease one month after surgery, complete remission was observed, at which time an autologous peripheral blood transplantation was performed.

**Fig. 1 – PET-CT**: (A) computed tomography demonstrating a mass measuring 4 × 3.5 cm in the right breast; (B) positron-emission tomography showing evidence of a mass with hyperuptake in the right breast with an SUV_{max} of 12.

Please cite this article as: Ruiz Pardo J, López López V, Blanquer Blanquer M, Cabezas Herrera J, Piñero Madrona A. Afectación mamaria por linfoma de células del manto en un varón. Cir Esp. 2017;95:170–172.
Table 1 – Published Cases of Mantle Cell Lymphoma in the Breast.

<table>
<thead>
<tr>
<th>Study</th>
<th>Year</th>
<th>Sex</th>
<th>Age</th>
<th>Laterality</th>
<th>Symptoms</th>
<th>Type</th>
<th>Stage</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boullanger et al.</td>
<td>2001</td>
<td>Female</td>
<td>71</td>
<td>Bilateral</td>
<td>Lymphadenopathies</td>
<td>Secondary</td>
<td>IIE</td>
<td>Chemotherapy; Chemotherapy + autologous peripheral blood stem cell transplantation</td>
</tr>
<tr>
<td>Windrum et al.</td>
<td>2002</td>
<td>Female</td>
<td>53</td>
<td>Right</td>
<td>Breast mass</td>
<td>Primary</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Fadare and Shukla</td>
<td>2008</td>
<td>Female</td>
<td>90</td>
<td>Bilateral</td>
<td>Breast mass</td>
<td>Secondary</td>
<td>IVE</td>
<td>Surgery + chemotherapy + radiotherapy</td>
</tr>
<tr>
<td>Hill and Seale</td>
<td>2011</td>
<td>Female</td>
<td>67</td>
<td>Left</td>
<td>Breast mass</td>
<td>Primary</td>
<td>IIE</td>
<td>Radiotherapy + chemotherapy + surgery + autologous peripheral blood transplantation</td>
</tr>
<tr>
<td>Dane et al.</td>
<td>2015</td>
<td>Male</td>
<td>56</td>
<td>Right</td>
<td>Lymphadenopathies</td>
<td>Secondary</td>
<td>IIE</td>
<td>–</td>
</tr>
</tbody>
</table>

Eleven months after the transplantation, a new systemic recurrence was observed, and the patient died due to an invasive fungal infection after neutropenic fever two months later.

Primary breast lymphoma is rare, comprising 1% of non-Hodgkin’s lymphomas and less than 3% of extranodal lymphomas. The definition of primary breast lymphoma provided by Wiseman and Liao⁶ and modified by Hugh et al.⁴ requires the presence of breast tissue very close to the lymphoma, the absence of a personal history of lymphoma and no extramammary dissemination except for the ipsilateral lymph nodes. Secondary breast lymphoma, which is more frequent, is defined as the presence of systemic lymphoma with simultaneous or subsequent breast involvement. In clinical practice, it is difficult to distinguish between primary mammary involvement with secondary dissemination and primary involvement in another area with secondary mammary involvement.² In the case presented, the lymphoma was secondary.

MCL is a non-Hodgkin’s lymphoma characterized by the involvement of lymph nodes, spleen, blood and bone marrow, with a short period of remission following conventional therapies.⁷ There are 3 histological growth patterns (mantle, nodular and diffuse) and 4 cytological variations (classical, small-cell, pleomorphic and blastic).⁸ The (11:14) chromosomal translocation is the distinctive molecular alteration, with overexpression of cyclin D1. According to the MCL International Prognostic Index, low-risk patients have a 5-year overall survival of 60%, moderate-risk patients have a median overall survival of 51 months, and high-risk patients have a median overall survival of 29 months.¹

There are few case reports in the literature of MCL of the breast (Table 1),⁶–¹⁰ and, to our knowledge, this case is the first describing MCL of the breast in a male.

Optimal therapy includes R-CHOP chemotherapy and locoregional radiotherapy, which appear to reduce the risk of local recurrence.¹¹⁻¹² Surgical resection of breast lymphomas has demonstrated poor local control. In patients treated with surgery alone, in stages IE and IIE the 5-year overall survival rates are 40.5 and 20.5%, respectively. Hence, surgical resection should be avoided and is indicated only to obtain biopsies.² In the cases of breast MCL described in the literature, only one surgical resection was performed, which, combined with chemotherapy and radiotherapy, resulted in an overall survival of 73 months.¹⁰ In our case, complete remission was achieved for 11 months.

Therefore, considering the exceptional nature of this single case and even though it is in direct contrast with what is stated in the literature, in patients with MCL of the breast, surgery could be useful to prolong the period of complete remission.

References

Endosalpingiosis is an obstructive entity simulating a Sigma Neoplasm

Endosalpingiosis as an Obstructive Entity Simulating a Sigma Neoplasm

Endosalpingiosis is the presence of ciliated tubal-type epithelium in an ectopic location other than its normal origin in the Fallopian tubes.1 There have been no studies that clarify the most prevalent signs and symptoms of endosalpingiosis. Patients may present pelvic pain or chronic pelvic inflammation, or endosalpingiosis may be found incidentally during surgery.2–4 Occasionally, they may even manifest like tumors.5 Even when benign in nature, there are case reports of malignant transformation to papillary serous adenocarcinoma.6

We present the case of a 67-year-old woman with a medical history of hypertension, dyslipidemia and hysterectomy due to symptomatic uterine fibroids with postoperative pelvic pain. She was currently being studied due to constipation for several months that could not be resolved with hygienic, dietary or pharmacological measures. In addition, the patient reported an important amount of weight loss, together with rectal and vaginal bleeding.

Given the suspicion of an underlying neoplastic process, a colonoscopy was performed, which demonstrated rigidity of the rectum/sigmoid colon 15 cm from the anal margin, which impeded passage of the colonoscope. No neoplastic-appearing mucosal lesions were observed, so no colon biopsies were taken. A lower gastrointestinal series (Fig. 1) revealed stenosis of the sigmoid colon with no observed cause and absence of diverticula that could demonstrate chronic diverticular disease. Virtual colonoscopy (Fig. 2) showed the stenosis, which was probably inflammatory in origin.

Given the persistence of the subocclusive patient symptoms, we decided on surgery. During surgery, the lesion in the sigmoid colon was observed, which caused stenosis and was macroscopically suggestive of a tumor process with superficial implants. The remaining structures of the cavity appeared normal, so we conducted an oncological anterior resection of the upper rectum. Postoperative evolution was satisfactory, and the patient was asymptomatic at later follow-up evaluations. The histology study defined the lesion as endosalpingiosis occupying the entire muscular wall and the pericolic adipose tissue, with muscular layer hypertrophy and signs of fibrosis, but no signs of malignancy.

Endosalpingiosis is defined as the presence of Fallopian tube tissue in an ectopic location, unlike müllerianosis, which is the ectopic presence of at least 2 of the 3 components derived from Müllerian tissue (endometrium, Fallopian tube epithelium and endocervix).1 The pathogenesis is not clear, and there are currently 2 theories that attempt to clarify this anomaly: coelomic metaplasia,5 which is most widely accepted, and the theory of implantation (transplantation of the tubal mucosa to the peritoneal surface during surgical procedures or after inflammatory processes).2 In our case, the history of a previous hysterectomy could be the origin of the sigmoid endosalpingiosis based on the theory of implantation.

There are discrepancies regarding the clinical manifestations of endosalpingiosis. According to Prentice et al.,2 this entity appears more in postmenopausal women and presents less pelvic pain than patients with endometriosis or those without endosalpingiosis. However, there are studies that propose a greater association between endosalpingiosis and

---

Endosalpingiosis as an Obstructive Entity Simulating a Sigma Neoplasm

Endosalpingiosis como una entidad obstructiva simuladora de neoplasia de sigma

Fig. 1 – Barium enema study with sigmoid stenosis.

---