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Intra-Abdominal Metastasis After Pelvic Primary Synovial Sarcoma Resection: Case Report

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Abstract

Introduction: Synovial sarcoma is a malignant soft tissue sarcoma. It occurs predominantly in young adults. 90% of synovial sarcomas occur in the extremities, they have been reported in other sites, including the abdomen and pelvis. Up to 40% of all patients with soft tissue sarcomas develop distant metastasis, despite adequate local disease control. **Aim of the article:** To provide an update on one of the rarest and most aggressive tumors, to affirm the importance of early management and especially surveillance, and to encourage the codification and the management of these tumors. **Presentation of cases:** Here, we discuss a case of a 78-year-old female patient, operated 10 years earlier for a pelvic Synovial sarcoma, who presented with intra-abdominal mass to our hospital that eventually turned out to be a metastatic intra-abdominal synovial sarcoma following biopsy. **Conclusion:** It is important to be aware that primary intraabdominal tumors can occur and delayed metastasis is more typical of synovial sarcoma than of many other

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sarcomas and should be considered in determining the appropriate frequency and duration of follow-up imaging.

Keywords: Synovial sarcoma; soft tissue sarcoma, metastatic sarcoma

1. Introduction

Synovial sarcoma is a malignant mesenchymal neoplasm accounting for about 5 to 10% of all soft tissue sarcomas. It occurs predominantly in young adults (15-40 years) with a slight male preponderance (male/female 1.2) (Thway, 2014) (Fisher, 1998). Primary intraabdominal synovial sarcoma is rare

Approximately 90% of synovial sarcomas occur in the extremities, and fewer than 5% occur in a joint or bursa. Rarely, they have been reported in other sites, including head and neck (5%), thoracic wall and cavity (8%), abdomen and pelvis (7%) (Bakri et al., 2012) (Ferrari et al., 2004), male and female genitourinary tracts, gastrointestinal tract, bone, and nervous system (Thway, 2014) (Fisher, 1998).

25 to 40% of all patients with soft tissue sarcomas develop distant metastasis with propensity for hematogenous spread, despite adequate local disease control (Pisters, 1996).

Here, we discuss a case of a 78-year-old female patient, operated 10years earlier for a pelvic Synovial sarcoma, who presented with intra-abdominal mass to our hospital that eventually turned out to be a metastatic intra-abdominal synovial sarcoma following biopsy.

2. Case Report

Here, we present a case of a 78-years old female, operated twice, 50years prior for an acute appendicitis and 10 years prior for a pelvic grade 2 FNCLCC (French Federation of Cancer Centers Sarcoma Group System) Synovial sarcoma followed by 18 radiotherapy sessions, who came to our hospital complaining about right hypochondrium heaviness-like pain for 18 months with deterioration of general state, no vomiting nor cholestasis were stated.

Physical examination showed a PS 2 patient with a BMI at 26.17kg/m², a distended abdomen with collateral circulation in the right flank, presence of a painful mass in the right hypochondria, measuring 8 cm long axis mobile at breathing, presence of a painful mass in the right iliac pit, measuring 5 cm long axis, fixed to the deep plane and mobile to the superficial plane. Presence of dullness of the flanks. Pelvic examination shows no abnormalities.

A Thoraco-abdomino-pelvic CT exam showed a solid-cystic tissue process, site of calcifications, interposing between the liver and the right diaphragm, internal convex limit, forcing the liver inside measuring

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166x80mm (Figure1), with a mass of the same appearance at the level of the right iliac pit, at depends on the root of the mesentery on the right measuring 89x88mm (Figure2).

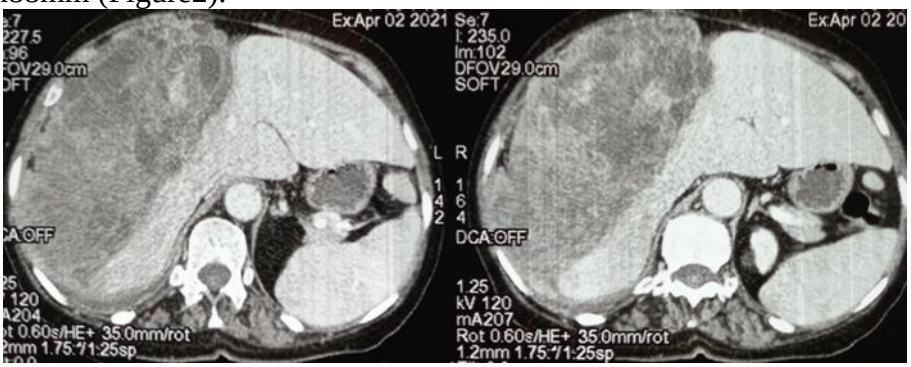

Figure 1: axial CT slice showing the solid-cystic tissue process interposing between the liver and the right diaphragm

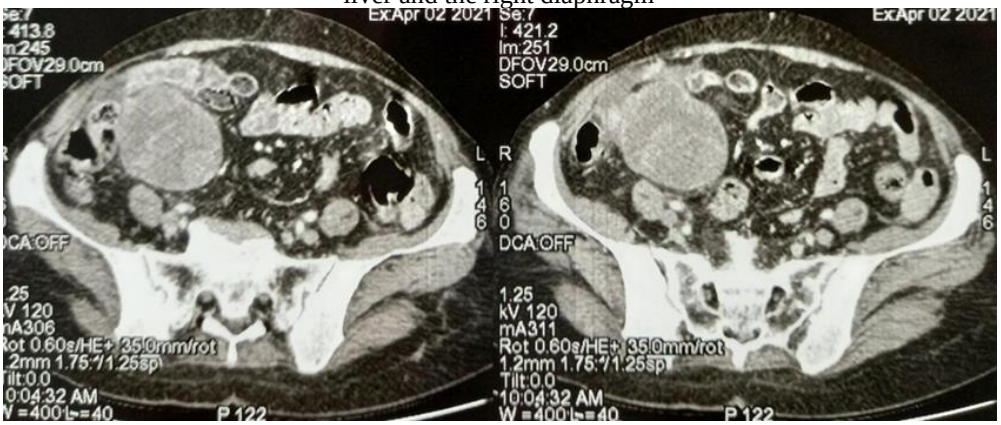

Figure 2: axial CT slice showing the solid-cystic tissue process of the right iliac pit

Blood investigations showed no abnormalities

Exploratory laparoscopy revealed the presence of a medium-abundance peritoneal effusion made of haematic-looking liquid (taken), the presence of a fixed interhepatodiaphragmatic mass measuring 20x10cm (biopsied), the presence of a fixed mass at the right iliac pit measuring 8x5 cm; peritoneal carcinosis nodules throughout the peritoneal cavity (Figure 3,4, and 5).

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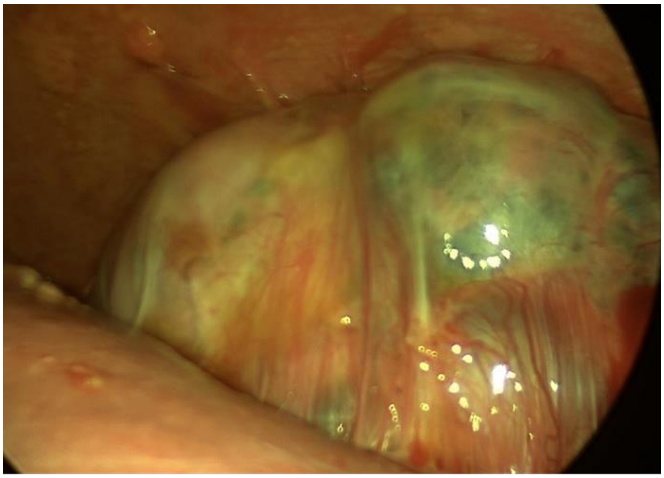

Figure 3: A laparoscopic image showing the interhepatodiaphragmatic mass

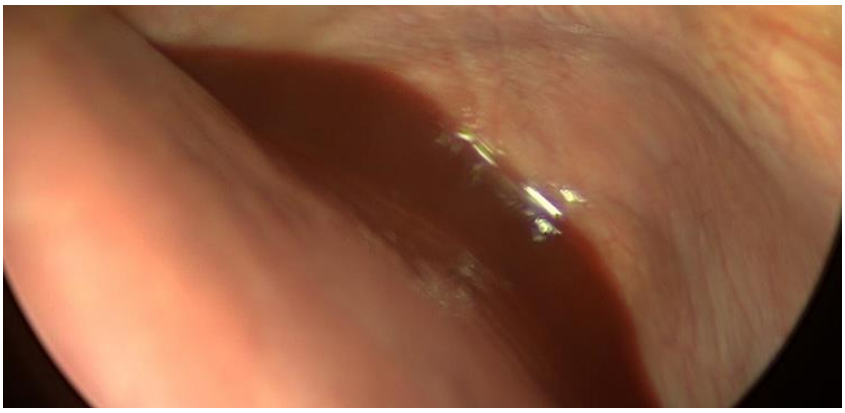

Figure 4: A laparoscopic image showing the hematic effusion

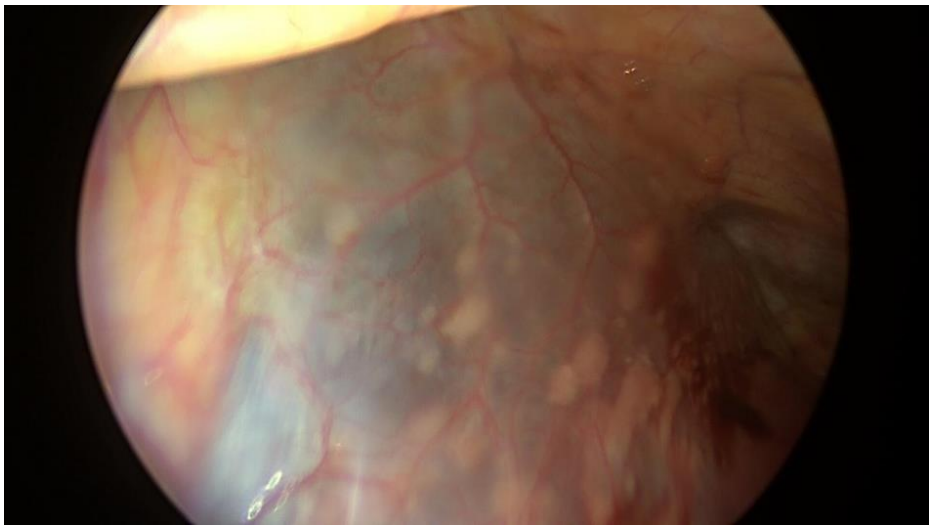

Figure 5: A laparoscopic image showing the peritoneal carcinosis nodules

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Histopathology revealed a fusiform cell monophasic FNCLCC Seconde grade Synovial sarcoma (Figure 6).

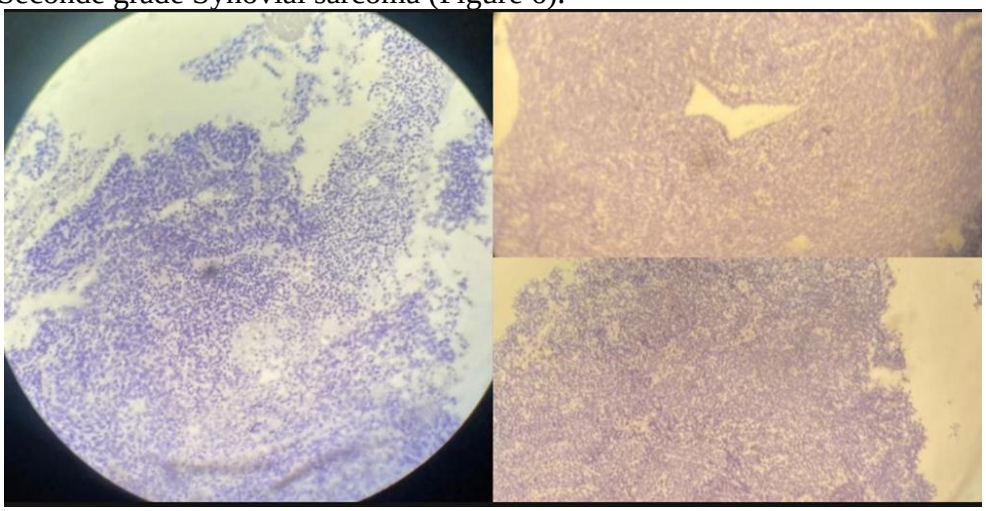

Figure 6: Histopathological examination

An immunohistochemical study revealed the expression of the AML, BCL2, EMA, CD99, and CKAE1/AE2 tumoral markers. Peritoneal effusion showed no malignant cells. The patient was put on chemotherapy.

3. Discussion

Synovial sarcoma is a misnomer because the tumor does not arise from the synovium; it only resembles synovial tissue at light microscopy. It appears to arise from as yet unknown multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures and lack synovial differentiation (Brahma, 2013). Synovial sarcoma occurs most commonly in the young, representing about 8% of all soft tissue sarcomas but about 15–20% of the cases occur in adolescents and young adults (Weiss, 2001).

There are three main histologic subtypes of synovial sarcoma: biphasic, monophasic, and poorly differentiated. Biphasic synovial sarcoma represents 20–30% of lesions and has both a mesenchymal spindle cell component and an obvious epithelial component as seen at light microscopy (Murphy, 2006). Monophasic synovial sarcoma represents 50–60% (the most common subtype) of all lesions, and in this subtype, the mesenchymal spindle cell component predominates (Murphy, 2006). Poorly differentiated synovial sarcomas are generally epithelioid in morphology and have high mitotic activity (usually 15–20/10 high-power field) with geographic necrosis. This subtype represents up to 15–25% of all synovial sarcomas (Murphy, 2006).

Synovial sarcomas, by definition, have been considered to be high-grade sarcomas. This contrasts with most soft tissue sarcomas that tend to have both a high and low-grade version. It is important to appreciate, however, that

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some groups in Europe and Asia believe that synovial sarcomas can and should be graded as intermediate and/or high-grade sarcomas (Guillou, 2004) (Hasegawa, 2002). Several recently published articles from these groups show improved disease-free and metastasis-free survival for patients with intermediate (Grade 2) synovial sarcomas (Trassard, 2001) (Guillou, 2004) (Hasegawa, 2002) (Hasegawa, 2001). The system typically used to stratify synovial sarcomas into intermediate (Grade 2) and high-grade (Grade 3) sarcomas is the French Federation of Cancer Centers Sarcoma Group System (FNCLCC) (Guillou, 1997). Mitotic count and percentage of tumor necrosis are used to stratify tumors in this system (Trassard, 2001) (Hasegawa, 2001). As this topic remains controversial, it is generally felt that it is best to consider all synovial sarcomas as high-grade sarcomas until the prognostic importance of grading synovial sarcomas is resolved (Hasegawa, 2002) (Hasegawa, 2001).

Unlike some soft tissue histologies, synovial sarcoma has no identifiable etiological agent or genetic condition that predisposes an individual to develop this malignancy (Brennan, 2005).

Synovial sarcomas can arise anywhere in the soft part of the body, generally as a progressively expanding mass. The most common clinical presentation is a slow-growing mass in the soft tissues. Patients present with a palpable, slowly growing, sometimes painful mass. Because of the insidious onset, there is often a delay in diagnosis. In one study (Brennan, 2005), the mean duration of symptoms before the patients sought medical attention was 2.5 years. Lesion size at diagnosis is variable, largely depending on the location. Although most tumors are larger than 5 cm, peripheral lesions are usually smaller, attributed to earlier discovery, often leading to diagnostic confusion with benign lesions (Kransdorf, 1995) (O'Sullivan, 2008) (Jones, 1993).

Primary intraabdominal synovial sarcoma is rare, with retroperitoneal lesions accounting for fewer than 1% of all cases of primary synovial sarcoma (Kransdorf, 1995). The typical imaging features of extremity primary tumors are heterogeneous T2 signal intensity, fluid-fluid levels, and areas of high T1 signal intensity suggestive of hemorrhage. The lesions may be larger than extremity primary lesions at presentation, resulting in displacement of adjacent structures such as the kidney. Retroperitoneal organs can also be sites of primary disease. Fewer than 50 cases of primary renal synovial sarcoma have been reported in the literature. CT and MRI may show the lesion as primarily cystic with a peripheral nodular component; however, a rapid growth pattern may be most suggestive of a more aggressive primary lesion than renal cell carcinoma (Perlmutter, 2005).

Although as many as 50% of synovial sarcomas recur locally within 2 years, late local recurrences and metastasis more than 5 years after initial

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diagnosis are common, potentially requiring longer follow-up (Krieg, 2011). Various prognostic factors have been reported, including patient age, tumor grade, and histologic subtype, but only a tumor size larger than 5 cm is consistently associated with a poor outcome (Krieg, 2011).

Like other soft-tissue sarcomas, synovial sarcoma metastasizes mainly to the lung. Unlike most other soft-tissue sarcomas, synovial sarcoma carries a small risk of spread to lymph nodes. Soft-tissue sarcomas originating from extremities rarely metastasize to the liver. One study (Jaques, 1995) showed a 75:1 ratio of the lung to the liver as sites of distant spread. However, the subset of patients with hepatic metastasis and primary disease of the abdomen was unusually high at 22%. Similarly, metastasis to the brain is also rare, even in widely disseminated diseases. There is also a high incidence of late metastasis. One prospective study showed almost 50% of deaths occurred within 5–10 years of diagnosis, emphasizing the importance of long-term follow-up (Singer, 1996). Like primary lesions, locally recurrent and metastatic lesions tend to calcify, a noteworthy characteristic from an imaging standpoint.

In our case, late metastasis was described after 10 years with the presence of peritoneal carcinosis

Intra-abdominal/hepatic metastasis are as expected, rare. Uniquely, synovial sarcomas have been shown to develop nodal metastases more commonly than most soft tissue sarcomas. Although the exact incidence of lymph node metastasis from synovial sarcoma varies in the literature, the pooled data from available studies puts the incidence at about 10–12% compared to about 3–5% for soft tissue sarcomas in general (Skinner, 1996).

For tumors that can be completely excised, surgical resection with or without radiotherapy has been found effective in establishing local control. Metastatic disease is more difficult to treat. Certainly, tumor response has occurred with first-line chemotherapy regimens consisting of ifosfamide-based chemotherapy (with or without doxorubicin). However, whether there is a substantial effect on long-term survival is uncertain. The toxicity and side effects must be weighed against potential benefits (Eilber, 2008).

Surgery has a much more limited role in the treatment of metastatic synovial sarcoma and requires careful patient selection. Patients are best selected by extent of disease, longer disease-free interval, and favorable response to systemic chemotherapy (Brennan, 2005).

The same rationale for the use of adjuvant radiation therapy in soft tissue sarcomas applies to synovial sarcoma. Because synovial sarcomas are all considered high-grade sarcomas, adjuvant radiation therapy is used in patients with tumors 5 cm in size (Brennan, 2005). It can be administered in a number of ways such as external beam therapy (neoadjuvant or adjuvant), brachytherapy, and intensity-modulated radiation therapy (IMRT). Each modality has its advantages and disadvantages, and no one modality

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specifically has been proven better for synovial sarcoma. Regardless of the type of radiation therapy employed, it has been proven to improve the local control rate in patients with high-grade sarcomas, such as synovial sarcoma (O'Sullivan, 2008)

Nevertheless, our patient went under radiotherapy, though, metastasis was discovered.

Synovial sarcoma has been regarded as a particularly chemosensitive soft tissue sarcoma. This was initially based on several studies that demonstrated impressive responses to ifosfamide-based chemotherapy in the treatment of metastatic and pediatric synovial sarcomas (Rosen, 1994) (Kampe, 1993) (Landenstein, 1993) (Okcu, 2003). These early findings have been supported by subsequent studies and thus ifosfamide-based chemotherapy (þ/ doxorubicin) is generally considered the first-line treatment for patients with metastatic synovial sarcoma (Ferrari, 2004) (Spurell, 2005). Although ifosfamide-based chemotherapy has been shown to produce notable responses in the treatment of metastatic synovial sarcomas, its effect on the survival of adult patients with primary disease has been unclear (Broowicz, 1993) (Gortzak, 2001).

Conclusion

It is important to be aware that primary intraabdominal tumors can occur, suggested by imaging features more typical of sarcoma than common primary tumors in those locations. Delayed metastasis is more typical of synovial sarcoma than of many other sarcomas and should be considered in determining the appropriate frequency and duration of follow-up imaging.

Declaration for Human Participants: This study has been approved by the head of the Department of general surgery of the IBN ROCHD University Hospital of Casablanca, Hassan II University of Casablanca, and the principles of the Helsinki Declaration were followed.

Conflict of Interest: The authors reported no conflict of interest.

Data Availability: All data are included in the content of the paper.

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