



Case report

Solitary fibrous tumor of the vulva resulting in spinal metastasis: A case report

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1. Introduction

Solitary Fibrous Tumors (SFTs) are rare mesenchymal tumors. They were historically described as originating from the pleura and were thought to be a mesothelial tumor, but now known to originate throughout the body from fibroblasts (Vallat-Decouvelaere et al., 1998). Despite the diversity of presenting sites, SFT involvement of the vulva is reported rarely (Biedrzycki et al., 2007; Fukunaga, 2000; He et al., 2010; Lee, 2016; Nielsen et al., 1997; Taki et al., 2012; Nag and Rao, 2015). SFTs typically follow indolent courses (Fukunaga, 2000). No reported cases of vulvar SFTs have metastasized to the central nervous system (Nielsen et al., 1997). In this case report, we present the course of a patient who, after a long indolent course, presented with a vulvar SFT notable for aggressive spread into the spinal cord.

2. Clinical scenario

A 64-year-old female with prior medical history notable for depression, hypothyroidism, and obesity was seen in September 2015 for a right labial mass that she had noticed for 15 years. It had been felt to be a lipoma and, given its stability on interval examination, it was observed for a number of years prior to definitive workup. When she was seen by a gynecologic oncologist, physical examination in clinic revealed a large 8 cm mass on the right side of midline on her mons pubis, with an otherwise normal labia, vagina, and cervix. She underwent a transvaginal ultrasound revealing a right adnexal mass measuring 3.7 × 3.0 × 4.0 cm, but was otherwise unremarkable. A CT of the abdomen and pelvis demonstrated no evidence of metastatic disease. The patient desired definitive management and was consented for

excision. The patient did not have symptoms of back pain or neurologic disease nor did she have pre-operative imaging of her cervical or thoracic spine or chest. Notably in November 2014, she developed a cough prompting a chest CT showing a groundglass lesion. Biopsy demonstrated evidence of sarcoidosis and subsequent chest imaging should resolution of the groundglass opacity.

In October 2015, the patient underwent exam under anesthesia, total laparoscopic hysterectomy, bilateral salpingo-oophorectomy and mass excision. Her estimated blood loss from the hysterectomy was approximately 75 ml, while that of her mons pubis mass resection was 500 ml. The tumor involved 85% of the specimen volume and tumor was present at the margins. It was approximately 9.8 cm in its largest extent, histologically grade 2, with mitotic rates ranging from 0 to 1 mitoses up to 12 mitoses per 10 high power fields in a focal area. There was no lymphovascular space invasion and necrosis involved 5% of the mass. The tumor predominantly appeared bland with only 5–10% of the tumor volume displaying malignant features. The pathology was reviewed at the Johns Hopkins Hospital, with agreement in the diagnosis. The adnexal mass was found to be a serous cystadenoma.

Upon discussion at a multidisciplinary tumor board conference, recommendations were made to perform a radical repeat resection to obtain clear margins, which she underwent in November 2015. The surrounding skin and subcutaneous tissue showed changes consistent with prior resection and no residual disease was found; her right inguinal lymph node was also negative for tumor.

She completed a course of 50 Gy to the operative bed in 25 fractions of 2 Gy each over a 39 day period in February to March 2016. She was unable to receive a planned 10 Gy boost as she was found, in March, to have metastatic disease in the spine causing pain and impairing her

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ability to walk. An MRI showed an infiltrative lesion in the L1 vertebral body, and a biopsy demonstrated the presence of metastatic sarcoma from her vulva. Given no other metastatic disease and significant functional and pain control issues, the patient was referred for definitive resection.

In April 2016, she underwent an L1 decompression and subtotal corpectomy. The tumor returned as a malignant spindle cell neoplasm associated with necrosis, consistent with metastasis from the patient's previously diagnosed extrapleural solitary fibrous tumor of the vulva. It stained positive for CD34, vimentin and CD99, as had the patient's primary tumor. Subsequent positron emission tomography scanning performed in May demonstrated new areas of FDG avidity concerning for new lung metastasis as well as residual disease versus post-surgical changes at L1. No further metastatic disease was noted.

The patient did well through the summer of 2016, but in September, she was noted to have increased pain and weakness, and was found on magnetic resonance imaging to have recurrent metastasis at the T10/L1 and S2 levels. She received palliative radiation to both areas and was started on daily temozolomide. In October, she developed worsening pain, lower extremity weakness, and urinary retention. She was diagnosed with cord compression from her metastatic disease at T6. However with symptoms present for > 24 h prior to admission, she was evaluated by the neurosurgical services and deemed not a candidate for additional surgical intervention. She received steroid therapy and additional palliative radiation. In November, her course was further complicated by pulmonary thromboembolic disease and she initiated anti-coagulation. She elected to transition her care to outpatient hospice, at which point temozolomide was discontinued. In January 2017, she became acutely anuric and obtunded, was admitted to inpatient hospice and subsequently died, 15 months from the original diagnosis and approximately 10 months from her spinal recurrence.

3. Discussion

In this article, we describe an aggressive vulvar solitary fibrous tumor notable for extensive spinal metastatic disease which was refractory to multimodal oncologic management. To our knowledge, this is the first case report of a vulvar SFT resulting in a recurrence after excision. Furthermore, while reports of primary spinal sites of solitary fibrous tumors exist in the literature, reports of spinal metastasis from SFTs of any primary site are rare (Jallo et al., 2005).

SFTs are typically indolent, and this patient's course was notable for speed of progression. Reports have described features within SFTs suggesting increased risk for recurrence including increased mitotic activity (> 4 mitotic figures per 10 high-powered fields), tumor necrosis, tumor size, increased cellularity and nuclear pleomorphism (Vallat-Decouvelaere et al., 1998). The treatment for SFTs involves resection by wide local excision. Adjuvant therapy remains controversial, but resection can be followed by radiation for high recurrence risk or tumor features worrisome for recurrence (Nag and Rao, 2015). This patient's tumor did demonstrate some high-risk findings, particularly high levels of mitotic figures in focal areas of the lesion. Based on these findings, the patient had prompt repeat resection to ensure complete removal of the primary tumor and underwent adjuvant therapy with radiation therapy to the excision bed.

Due to the rarity of solitary fibrous tumors with malignant transformation, no guidelines have been established to guide adjuvant therapy. Although some studies suggest that radiation therapy is equivalent to surgery alone in comparison of its morbidity, this option may be reserved for patients who are not appropriate surgical candidates or refuse surgery (Yang et al., 1998; Kawamura et al., 2007). Case reports report radiation use, in addition to adjuvant chemotherapy, in patients with multiple sites of SFTs. In evaluations of adjuvant chemotherapeutic regimens, treatments that have shown no benefit for SFTs include ifosfamide with mesna, paclitaxel, and doxorubicin with cisplatin (Gold et al., 2002). The data has been contradictory for

temozolomide with bevacizumab with some studies showing significant antitumor activity and others demonstrating no benefit compared to single-agent temozolomide or dacarbazine (Stacchiotti et al., 2013; Park et al., 2011).

Despite these efforts, the patient developed worsening metastatic disease, refractory to surgical resection, as well as adjuvant radiation and chemotherapy. While excision of the spinal metastasis was not pursued with curative intent, the lesion at L1 was the only recurrence detected. Given this patient's recurrence was in a location that was significantly affecting quality of life, mobility, and functional capacity, resection, even within 6 months of initial surgery, was reasonable.

We note the 500 ml estimated blood loss from the primary excision. Other case reports have noted excessive bleeding associated with SFT resections, particularly with deeper tumor invasion. Some have recommended contrast enhanced imaging and consideration of embolization preoperatively, particularly for intraabdominal tumors (Yokoyama et al., 2015). Given the location and relatively smaller size of this tumor, this was not viewed as necessary. Consideration may be made of judicious local infiltration of vasoconstrictive agents, such as epinephrine.

This case highlights a rare presentation for an SFT, in that the patient presented with a longstanding vulvar growth, had it resected with positive margins, and despite re-excision and radiotherapy, had metastatic disease to her spine. While preoperative comprehensive imaging of the spine and the chest and abdomen may have detected pre-existing metastases, she was without symptoms. Others have found tumor size > 13 cm to be a predictor of metastatic potential, and this may be a reasonable threshold to consider imaging (Vallat-Decouvelaere et al., 1998). She underwent adjuvant therapy with radiation as well as systemic chemotherapy given her distant recurrence. Whether these therapies influenced her overall survival cannot be determined. However, given the rarity of this disease, her situation adds to the little information we have on this topic and serves to add to our repertoire of therapies that may guide management in the future for other patients. Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

- Biedrzycki, O.J., Singh, N., Habeeb, H., Wathen, N., Faruqi, A., 2007. Solitary fibrous tumor of the female genital tract a case report and review of the literature. *Int. J. Gynecol. Pathol.* 26 (3), 259–264.
- Fukunaga, M., 2000. Atypical solitary fibrous tumor of the vulva. *Int. J. Gynecol. Pathol.* 19 (2), 164–168.
- Gold, J.S., Antonescu, C.R., Hajdu, C., Ferrone, C.R., Hussain, M., Lewis, J.J., et al., 2002. Clinicopathologic correlates of solitary fibrous tumors. *Cancer* 94 (4), 1057–1068.
- He, Y., Yang, K.X., Yang, F., Wu, X.L., 2010. Primary solitary fibrous tumor of the vulva: a case report. *J. Reprod. Med.* 55 (9–10), 452–456.
- Jallo, G.I., Roonprapunt, C., Kothbauer, K., Freed, D., Allen, J., Epstein, F., 2005. Spinal solitary fibrous tumors: a series of four patients: case report. *Neurosurgery* 57 (1), E195 (discussion E).
- Kawamura, S., Nakamura, T., Oya, T., Ishizawa, S., Sakai, Y., Tanaka, T., et al., 2007. Advanced malignant solitary fibrous tumor in pelvis responding to radiation therapy. *Pathol. Int.* 57 (4), 213–218.
- Lee, D.H., 2016. Solitary fibrous tumor arising in the mons pubis: a case report. *Eur. J. Gynaecol. Oncol.* 37 (3), 423–425.
- Nag, G., Rao, S.R., 2015. A rare mesenchymal neoplasm at unusual location: solitary fibrous tumor of vulva. *Gynecol. Oncol. Rep.* 12, 52–54.
- Nielsen, G.P., O'Connell, J.X., Dickersin, G.R., Rosenberg, A.E., 1997. Solitary fibrous tumor of soft tissue: a report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultrastructural data. *Mod. Pathol.* 10 (10), 1028–1037.
- Park, M.S., Patel, S.R., Ludwig, J.A., Trent, J.C., Conrad, C.A., Lazar, A.J., et al., 2011. Activity of temozolomide and bevacizumab in the treatment of locally advanced, recurrent, and metastatic hemangiopericytoma and malignant solitary fibrous tumor. *Cancer* 117 (21), 4939–4947.
- Stacchiotti, S., Tortoreto, M., Bozzi, F., Tamborini, E., Morosi, C., Messina, A., et al., 2013. Dacarbazine in solitary fibrous tumor: a case series analysis and preclinical evidence vis-a-vis temozolomide and antiangiogenics. *Clin. Cancer Res.* 19 (18), 5192–5201.
- Taki, M., Baba, T., Mandai, M., Suzuki, A., Mikami, Y., Matsumura, N., et al., 2012. Solitary fibrous tumor arising slowly in the vulva over 10 years: case report and

- review. *J. Obstet. Gynaecol. Res.* 38 (5), 884–888.
- Vallat-Decouvelaere, A.V., Dry, S.M., Fletcher, C.D., 1998. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. *Am. J. Surg. Pathol.* 22 (12), 1501–1511.
- Yang, J.C., Chang, A.E., Baker, A.R., Sindelar, W.F., Danforth, D.N., Topalian, S.L., et al., 1998. Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas of the extremity. *J. Clin. Oncol.* 16 (1), 197–203.
- Yokoyama, Y., Hata, K., Kanazawa, T., Yamaguchi, H., Ishihara, S., Sunami, E., et al., 2015. Giant solitary fibrous tumor of the pelvis successfully treated with preoperative embolization and surgical resection: a case report. *World J. Surg. Oncol.* 13, 164.