CASE REPORT

Case Report: An incidental finding of an adrenal metastases noted in a “collision tumor” from a large malignant nerve sheath tumor of the thigh [version 1; referees: awaiting peer review]

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Abstract

In cases of peripheral nerve sheath tumors, current guidelines do not recommend routine abdominal imaging to stage the disease, as extra-pulmonary metastasis is considered rare. We report a case of large peripheral nerve sheath tumor in a 40 year-old-female with neurofibromatosis type 1 who had isolated adrenal metastasis. She underwent primary and adrenal metastasis resection.

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Introduction

Soft tissue sarcomas of the extremity are a rare, comprising less than 1% of all malignancies. These sarcomas are a histologically heterogeneous group of tumors with a predilection for hematogenous spread. Distant metastatic disease is found in approximately 20–30% of patients, with pulmonary lesions accounting for 75% of these cases. Because of the relatively low incidence of extra-pulmonary metastasis, the current guidelines from the National Comprehensive Cancer Network (NCCN) recommend to consider abdominal/pelvic CT imaging for certain subgroups of sarcomas (myxoid/round cell liposarcoma, epithelial sarcoma, angiosarcoma, leiomyosarcoma). A similar recommendation is made by the European Society of Medical Oncology (ESMO)/European Sarcoma Network Working Group.

We report a case of a malignant peripheral nerve sheath tumor of the thigh, without evidence of concomitant pulmonary metastases, which was found to have metastasized to the adrenal gland. This case raises the question if routine abdominal imaging should be performed along with pulmonary imaging in metastatic peripheral nerve sheath tumors.

Case report

A 40 year old woman with neurofibromatosis type 1 and no other significant past medical history or family history, presented for a hemorrhagic right thigh mass which had been enlarging over the past 3 months. The mass had previously been stable in size for a few years and was thought to be consistent with a neurofibroma. A CT scan of the extremities revealed a large mass within the posterior compartment of the right thigh, measuring 13.4 × 13.4 × 24.8 cm. In addition, an incidental 2 × 2cm left adrenal gland mass and 2 small lytic bone lesions (right 10th rib and right proximal femur) were also noted. Of note, a CT scan of the thorax, completed for staging purposes, did not reveal any suspicious pulmonary nodules or masses. An excisional biopsy of the thigh mass confirmed a high grade spindle cell sarcoma, negative for S100 protein, Desmin, CD31, AE1:AE3, HMB45, MelanA, SOX10 and TLE1. The lack of immunoreactivity for all performed markers for this tumor was most compatible with a malignant peripheral sheath tumor (Figure 1 and Figure 2). An MRI of the abdomen, to further evaluate the adrenal mass, revealed 2 small left adrenal gland lesions in the medial and lateral limbs. Given the known association of neurofibromatosis with pheochromocytoma, a biochemical workup showed elevated plasma free metanephrines, supporting the diagnosis of pheochromocytoma. The patient underwent a laparoscopic adrenalectomy, with gross pathological exams revealing two tumor nodules and with a histological exam revealing an intermingled “collision” tumor involving pheochromocytoma and sarcoma, consistent with metastatic peripheral nerve sheath tumor. The patient refused radiation therapy and did not follow up with oncology.

Figure 1. Section from the thigh mass (10x, H&E) shows a hypercellular tumor, with spindle cells in sheets and fascicular arrangement. The spindle-shaped nuclei have clumped chromatin. These features are compatible with a malignant peripheral nerve sheath tumor.

Figure 2. The area on the right shows two populations of tumor cells that are intermingling with each other, representing a collision tumor (20x, H&E). One population is composed of hypercellular malignant spindle cells with hyperchromatic nuclei (blue arrow) that are infiltrating the adjacent adrenal tissue. This is morphologically compatible with malignant peripheral nerve sheath tumor. The other population is composed of the nests of polygonal cells with abundant eosinophilic cytoplasm (green arrow), compatible with pheochromocytoma.
Discussion
The above case describes a patient with a malignant peripheral nerve sheath tumor (MPNST) of the thigh with pathologically confirmed metastases to the adrenal gland, yet without evidence of pulmonary metastases. The adrenal mass was found incidentally upon imaging of the thigh mass, but based on current NCCN guidelines, a screening CT of the abdomen/pelvis (A/P) would not be indicated and thus could have potentially missed the presence of metastatic disease.

Two recent case series reached contradictory conclusions regarding the benefit of abdominal/pelvic CT screening. In the first case series, King, et al. evaluated 124 adult patients with sarcoma who underwent CT chest/C/A/P imaging at their institution for staging and surveillance. Twenty (16%) of the patients had evidence of A/P metastasis, on the initial scan and 13 on the surveillance. Of note, six of the 20 patients (5% of the cohort) were found to have isolated A/P metastases without the development of pulmonary metastases during the study period. MPNST, specifically, made up 6% of the sarcomas evaluated and while no A/P metastases were found on screening, 2 patients had evidence on surveillance scans. Based on the finding that a wide variety of sarcoma subtypes were found to have extra-pulmonary disease, the authors conclude that A/P imaging should be included in the evaluation of all sarcoma subtypes. In contrast, Thompson et al. reviewed 140 patients of all ages who had a diagnosis of a malignant neoplasm of the upper or lower extremity and underwent screening and/or surveillance with a CT C/A/P. Of these patients, 14 (10%) had evidence of abdominal/pelvic metastasis, with only 4 (2.9%) with evidence of isolated A/P disease. Additionally, of the 10 patients who developed metastases to both the chest and abdomen/pelvis, none developed evidence of disease in the abdomen/pelvis prior to the chest. Based on their results, the authors offer up the contrary opinion from King in that abdominopelvic imaging is not warranted in the sarcoma population.

Our case adds a patient to the literature with a peripheral MPNST who was found to have an isolated adrenal metastasis without evidence of concomitant pulmonary disease. One striking feature of this case is the very large size of the primary tumor. Factors known to be associated with the development of metastases are tumor grade, tumor size, tumor depth, and certain histopathologies. Currently, though, guidelines only recommend CT of the chest for all patients and to consider CT A/P in patients with myxoid/round cell liposarcoma, epithelial sarcoma, angiosarcoma and leiomyosarcoma. Based on our case and the literature, we also would suggest adding screening CT A/P for large (>5cm), deep tumors of any histology.

Conclusions
Soft tissue sarcomas are a heterogeneous group of tumors with a variety of prognoses. Deciding on a unified set of guidelines will be challenging, but given the clinical significance of finding metastatic disease, adding additional parameters (size and depth) to a more complete screening process would seem prudent.

Consent
Written informed consent was obtained by the patient for publication of their clinical details. There are no potentially identifying images included in this paper.

Competing interests
No competing interests were disclosed.

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References
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