

Subcutaneous axillary and scalp metastases from non-gynecological retroperitoneal leiomyosarcoma: an unusual presentation after surgical resection

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Abstract

Retroperitoneal leiomyosarcomas are rare sarcomas, with an incidence of less than 2 per million population. Cutaneous metastases from sarcoma account for only 1-2.6% of metastatic skin lesions. Cutaneous and subcutaneous metastasis from retroperitoneal leiomyosarcoma is a very rare entity. We present a case of 72-year-old male with scalp nodule and subcutaneous swelling in left posterior axillary fold. Fine needle aspiration cytology from both these sites revealed a sarcoma, which was positive for Smooth Muscle Actin and negative for S100 on cell block immunohistochemistry (IHC). The past history revealed surgical resection of a retroperitoneal mass in 2010 which was diagnosed on histopathology and IHC as leiomyosarcoma. A final diagnosis of metastatic deposits from leiomyosarcoma was made. Retroperitoneal leiomyosarcoma presenting as scalp and subcutaneous metastasis is an unusual presentation. Adequate clinical history and a high index of clinical suspicion is required to detect cutaneous and subcutaneous metastatic deposits occurring five years after surgical resection.

Introduction

Leiomyosarcomas are rare tumors of smooth muscle origin with incidence of less than two per million people and account for about 10-37% of all retroperitoneal sarcomas.^{1,2} Hematogenous spread is common and they usually metastasize to lung, liver, kidney and brain. Cutaneous metastasis of leiomyosarcoma is an unusual occurrence.³ We report a case of metastatic scalp and subcutaneous tissue deposits from retroperitoneal leiomyosarcoma of non-gynecological origin, resected 5 years back.

Case Report

A 72-year-old man presented with a firm, mobile, non-tender nodule in the left posterior axillary fold which he noticed two months ago. He also gave history of a scalp swelling since one year. On examination, the scalp swelling and the posterior axillary fold swellings measured three and 2 cm in diameter respectively which were subjected to fine needle aspiration cytology (FNAC). The smears from both the sites were cellular and composed of cohesive clusters of spindle shaped overtly malignant cells displaying marked nuclear atypia, elongated cigar shaped nuclei with blunt ends and abundant granular cytoplasm which was positive for Smooth Muscle Actin (SMA) and negative for S100 on cell block immunohistochemistry (IHC; Figure 1A-C). Abdominal ultrasonography revealed a heterogeneous lesion, measuring 3.4×3.5 cm, in the right lobe of liver which was also suspected to be metastatic deposit.

The patient's past record revealed history of a retroperitoneal tumor measuring 23×15×12 cm, for which surgical excision had been done in 2010 (Figure 2). The tumor was also seen to involve the hilum of the right kidney, hence right sided nephrectomy was also performed along with tumor resection. The histopathological examination showed interlacing fascicles of spindle cells exhibiting marked cytological atypia and more than 5 mitotic figures/10 HPF (Figure 1D). The diagnosis of leiomyosarcoma was based on morphology and confirmed by IHC which demonstrated positivity for Vimentin, Desmin and SMA while S100, Chromogranin, NSE, HMB-45, CD-117, CD-34, Cytokeratin and EMA were negative.

Thus, keeping the microscopic findings and past history of retroperitoneal leiomyosarcoma in mind, the scalp and the subcutaneous axillary fold swellings were diagnosed as metastatic deposits from leiomyosarcoma.

Discussion

The occurrence of cutaneous metastasis is reported in only 0.7-9% of cases of all malignancies.⁴ Metastatic skin deposits from sarcomas are rarer still, accounting for 1-2.6% of all metastatic skin tumors.⁵ Among sarcomas, leiomyosarcomas are more likely to give rise to cutaneous metastatic deposits and scalp is a common site of involvement, due to its rich vascularity.⁶ However, to the best of our knowledge, there are only three other instances where scalp and subcutaneous metastatic deposits have been reported from a non-gynecological (retroperitoneal) leiomyosarcoma.⁷⁻⁹

Most of the cases of cutaneous metastatic

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deposits from leiomyosarcomas also have concomitant metastatic deposits in other internal organs which signifies advanced disease and hence, poor prognosis.¹ This finding was also present in our patient who had radiological evidence of liver metastasis. According to American Joint Committee on Cancer (AJCC) grading, the tumor was in Stage 4.

Retroperitoneal leiomyosarcomas commonly occur in fifth to seventh decade, as also seen in our patient.¹⁰ These tumors generally acquire large dimensions, the median size at the time of diagnosis being around 15 cm.¹¹ The retroperitoneal tumor in our case measured 23×15×12 cm, consistent with this observation. The retroperitoneum provides a widely expansile area conducive for tumors to attain large sizes before the patients become symptomatic due to mass effects produced by the tumor.¹² Complete surgical resection is the treatment of choice. However, proximity to vital organs and major vessels in the abdomen may preclude complete excision of the tumor.¹³ Local recurrence rates range from 40-77%.

Distant and widespread metastases can occur years after surgical resection, as seen in our patient, and herald poor prognosis.¹⁴ Leiomyosarcomas with distant metastases are

categorised as Stage IV according to AJCC guidelines, and have five year survival rates less than twenty percent.¹⁵ Other prognostic markers include tumor site, size, grade and

mitotic figures. Metastatic leiomyosarcomas are treated with surgery and/or adjuvant chemotherapy, but no standard treatment has been established so far.

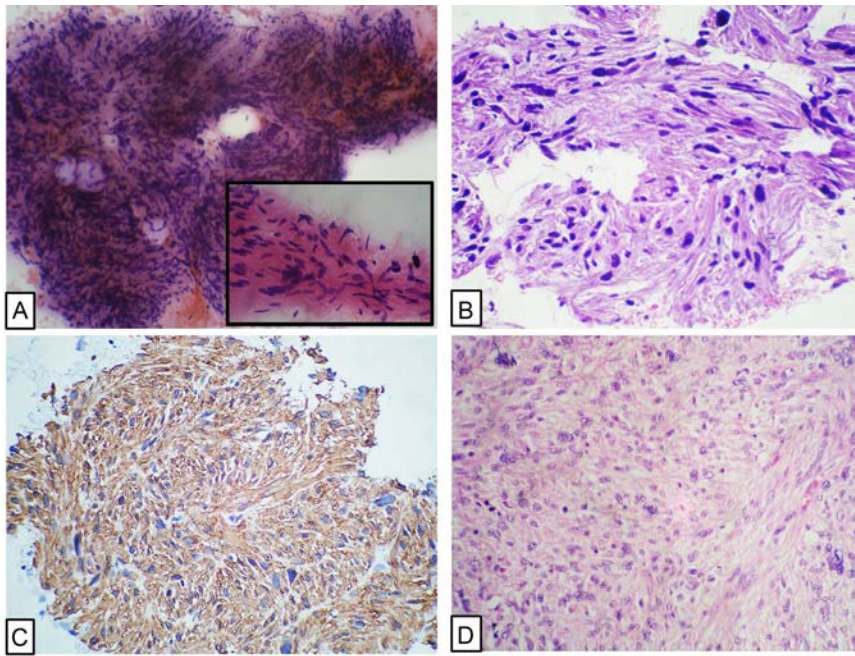


Figure 1. A) Photomicrograph of fine needle aspiration smear from axillary swelling showing cohesive cluster of spindle shaped malignant cells with nuclear atypia (Papanicolaou stain, 100×). Inset showing tumor cells having abundant granular cytoplasm and elongated cigar shaped nuclei with blunt ends (Hematoxylin & Eosin, 400×). B) Cell block from same axillary swelling showing cohesive clusters of malignant spindle cells with blunt ended nuclei and moderate nuclear atypia (Hematoxylin & Eosin, 100×). C) Cytoplasmic positivity for Smooth Muscle Actin. D) Histopathology section of the original retroperitoneal tumor showing interlacing fascicles of spindle cells exhibiting marked cytological atypia and more than 5 mitotic figures/10 HPF (Hematoxylin & Eosin, 100×).

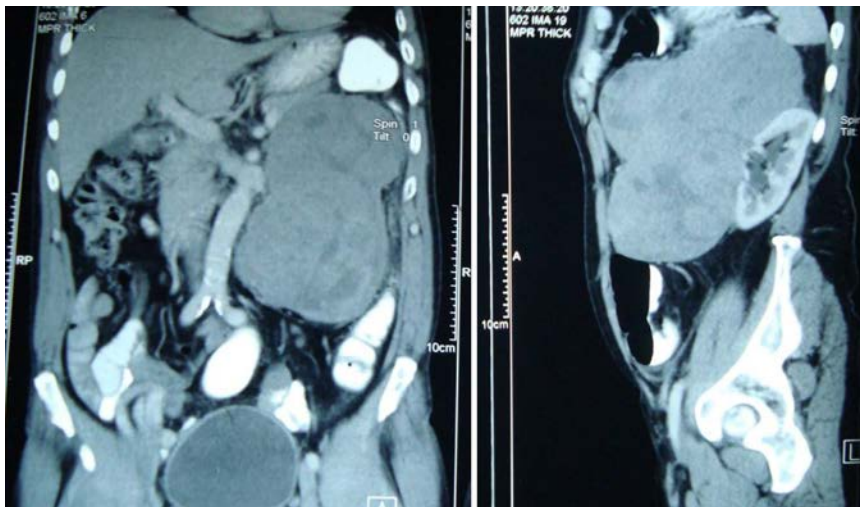


Figure 2. Computed tomography scan of abdomen showing the original retroperitoneal leiomyosarcoma as a large multilobulated retroperitoneal homogenous enhancing mass in right hypochondrium and lumbar region, with few non enhancing foci suggestive of necrosis.

Conclusions

Retroperitoneal leiomyosarcoma presenting as subcutaneous axillary and scalp metastases, 5 years after surgical resection, is an unusual scenario. Our case report highlights the significance of adequate clinical history and a high degree of clinical suspicion to detect and correctly diagnose metastatic deposits occurring late in the course of disease. These patients need to be kept on long term follow up and any new lesion anywhere in the body, even in an odd and distant location, needs to be investigated thoroughly to rule out metastasis.

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