A Case of Clear Cell Sarcoma Occurring on the Abdomen

Suck Joon Jung, M.D., Na Reu Seung, M.D., Eun Ju Park, M.D., Chul Woo Kim, M.D.,
Kwang Ho Kim, M.D., Kwang Joong Kim, M.D.

Department of Dermatology, College of Medicine, Hallym University, Anyang, Korea

Clear cell sarcoma is a rare malignant tumor representing about 1% of soft tissue tumors. It usually presents in the distal extremities of young adults, frequently attached to tendons or aponeuroses. This slowly progressive tumor tends to recur and results in eventual development of metastatic growth. Early recognition of the disease and prompt wide excision of tumor are essential to get a favorable outcome. We report a rare case of clear cell sarcoma in a 57 year-old female who presented with an erythematous hard nodule on her abdomen.
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INTRODUCTION

Clear cell sarcoma is a rare malignant soft tissue neoplasm that usually arises adjacent to tendons or aponeuroses¹. It generally affects young adults with a predominance in women and mostly appears in the extremities, especially the feet and ankles². The clinical course is rather slow, with repeated local recurrences followed by late metastases and eventual death³. Involvement of the abdomen is rare. We report a case of clear cell sarcoma affecting the abdomen.

CASE REPORT

A 57-year-old female visited our clinic with a chief complain of a skin lesion on her right lower abdomen of two years duration. She had a mitral valve replacement surgery 5 years ago, and is now taking warfarin for maintenance therapy. Other than this, there is nothing special in her past

medical or family history. In the physical examination we found a pea- sized glistening erythematous nodule on her right lower abdomen (Fig. 1). This nodule gradually increased in size but there was no pain or tenderness. We performed a punch biopsy on the tumor. The histologic examination revealed that the neoplastic cells were divided into well-defined nests and groups by fibrous tissue septa (Fig. 2). The cells consisted of round to ovoid vesicular nuclei and pale-staining cytoplasm. In the immunohistochemical staining the tumor cells showed positive reaction to S-100, Vimentin (Fig. 3) and negative reaction to Ki-67, CD34, SMA, Desmin. We performed a whole body PET-CT to ascertain the infiltrated depth of the tumor and the evidence of lymph node or distant metastasis. The PET-CT showed no other abnormal uptake except the 1 cm sized tumor in the right lower anterior abdomen. A radical excision was carried out and the resection margins were free of the tumor. There's no sign of recurrence for 14 months and now we are observing the progress of the disease.

DISCUSSION

Clear cell sarcoma (CCS), first described by Enzinger¹ in 1965, is a rare soft tissue tumor. Before this, this uncommon neoplasm had been misdiagnosed as fibrosarcoma, synovial sarcoma, heman-

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Reprint request to: Kwang Joong Kim, M.D., Department of Dermatology, Hallym University Sacred Heart Hospital, 896, Pyeongchon-dong, Dongan-gu, Anyang 431-070, Korea. Tel: 82-31-380-3761, Fax: 82-31-386-3761, E-mail: kkj51818@hallym.or.kr

Fig. 1. (A, B) Solitary pea sized glistening erythematous nodule with peripheral brownish patch on her right lower abdomen.

Fig. 2. Tumor cells are observed to be infiltrated in the dermis (A: H&E, × 40), and composed compact nests and fascicles (B: H&E, × 100). Tumor cells consist of round to ovoid vesicular nuclei and pale-staining cytoplasm (C: H&E, × 400).

Fig. 3. The tumor cells show reactivity with (A) S-100 protein, (B) Vimentin (Immunohistochemical stain, $\times 100$).

giopericytoma, alveolar soft-part sarcoma, and hemangioendothelioma⁴. It occurs most frequently in the feet and ankles of women in the second and third decades¹. The tumor is characterized by multiple local recurrences with late metastases and a high rate of deaths³. It usually presents as a slowly growing mass and occasionally causes mild pain or tenderness². Symptoms may persist for a long time (mean of 5 years) before the patient seeks medical attention².

Gross pathologic examination of CCS reveals a localized, tan-gray, firm and somewhat circumscribed mass, varying in size from 0.4 cm up to 14.5 cm⁵. Frequently, the mass is attached to tendons or aponeuroses, but there is no direct connection with overlying skin⁶. On microscopic analysis, epithelioid cells with pale nuclei and deeply basophilic nucleoli are aggregated in compact nests surrounded and separated from adjacent nests by fibrous tissue septa⁷. Mitoses are generally sparse and necrosis and haemorrhage are rare⁸.

On immunohistochemical examination, the tumor cells of nearly all cases express S-100 protein³. Most of them also express antigens associated with melanin synthesis (HMB-45, melanin-A, Mel-CAM)³. Cytokeratin, epithelial membrane antigen, carcinoembryonic antigen, desmin, and smooth muscle actin are usually negative⁹. In our case, staining the tumor cells showed positive reaction to S-100, Vimentin and negative reaction to Ki-67, CD34, SMA, Desmin.

Ultrastructural analysis shows tumor cells closely apposed to each other by continuous basal laminae and rudimentary cell junctions⁹. Cells show abundant cytoplasm containing numerous mitochondria and aggregates of glycogen⁹.

The differential diagnosis includes synovial sarcoma, fibrosarcoma, epithelioid forms of malignant peripheral nerve sheath tumor, spindle cell melanoma. One of the main differential diagnoses of CCS is malignant melanoma (MM). CCS and MM demonstrate significant morphologic overlap at light microscopic and ultrastructural levels, as well as similar immunohistochemical features, so the distinction may be difficult⁵. However, the behavior of CCS is decidedly different from that of MM, demonstrating a more indolent course⁷. CCS is a deep seated tumor that rarely shows the degree of anaplasia, necrosis, and mitotic activity usually associated with MM. Moreover the t(12 ; 22) translocation and EWSR1/ATF1 gene rearrangement observed in the majority of cases of CCS have never been found in MM⁵. Approximately 50% of MMs showed deletions of short arm of chromosome 9 at the interferon alfa locus⁵.

Treatment should be radical resection of the tumor, followed by chemotherapy and radiotherapy². However chemotherapy and radiotherapy have not been shown to be of benefit^{5,10}, so early recognition of this disease and prompt wide excision of tumor is essential for a favorable outcome¹¹. The prognosis for patients with CCS is generally poor, although

those with tumors smaller than 2 cm have a better prognosis¹². Poor prognosis is associated with tumor size more than 5 cm, presence of necrosis, metastasis and local recurrence⁵.

We encountered a rare case of CCS arising in the abdomen. Although CCS is usually fatal after a long clinical course, our patient is still alive without recurrence 14 months after resection.

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