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Clear cell sarcoma originating in a paraspinous tendon: Case report and literature review

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To the Editor

Sarcomas are amongst the least common tumors clinical oncologists encounter; however even amongst soft tissue sarcomas, subsets requiring distinct treatment plans exist. Gastrointestinal stromal sarcomas (GIST) have undergone a revolutionary paradigm shift in treatment with the discovery of targeted kinase inhibitors; other less treatable soft tissue sarcomas also require distinct treatments, even as regimens continue to evolve [1]. Although clear cell sarcoma, being an uncommon entity, is not in the differential of common paraspinous tumors this rare presentation illustrates several important aspects of truncal sarcoma management for the practicing oncologist.

We report a rare paraspinous clear cell sarcoma. A 38-year-old African-American fireman was admitted to the hospital for severe pain beginning in the left subcostal region and radiating to the left upper quadrant. A magnetic resonance imaging (MRI) scan showed a paraspinous mass 2.0×5.0 cm in size (Figures 1 and 2)

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The patient underwent a thoracotomy from a left-sided approach for diagnosis and resection because of the tumor's predominantly left-sided location. Non-necrotic margins displayed a clear cell pattern, initially felt to be consistent with a renal primary. Follow-up immunocytochemical panel included strongly positive results for S100, HMB45, and periodic acid-Schiff (PAS) reaction (Figure 3). No melanin pigment was seen; a clear cell sarcoma was suspected. Subsequently, the EWS-ATF-1 fusion gene was detected.

Subsequently, the patient underwent a course of Cisplatin, Dacarbazine, interleukin-2, and interferon. Briefly, the patient received Dacarbazine 250 mg/m^2 intravenously on days 1, 2 and 3; Cisplatin 25 mg/m^2 on days 1, 2 and 3; interleukin-2 18 million units/m^2 days 6–10 and 13–15 and interferon 5 million units/m^2 on days 6, 8, 10, 13 and 15. After two full cycles of chemotherapy, the patient was noted to have completely responded. No new lymphadenopathy was seen. The tumor subsequently recurred.

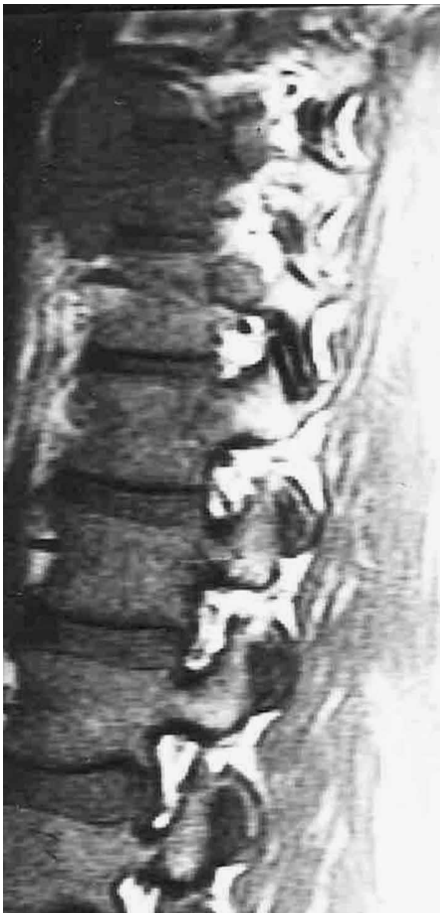

Figure 1. Sagital T1-weighted MRI of the thoracic spine showing marrow infiltration, endplate destruction, and paravertebral mass at T5 and T6 level. Note involvement of anterior longitudinal ligament.

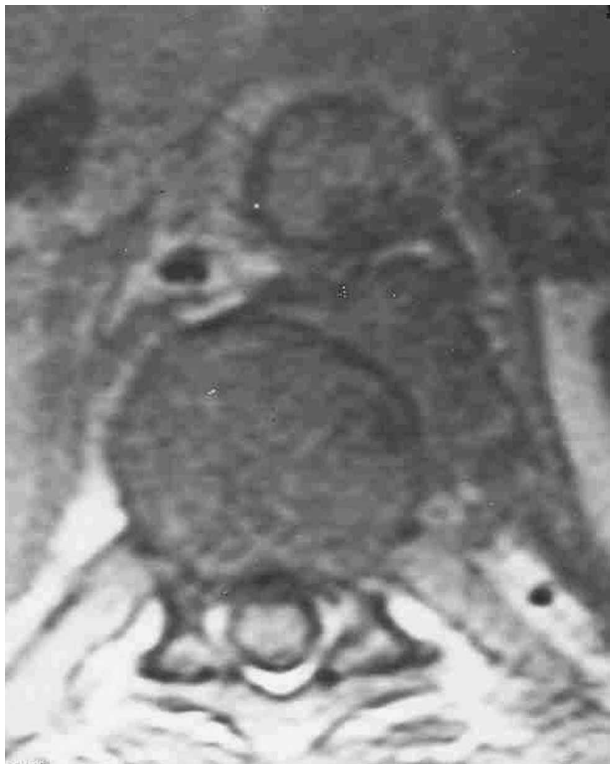

Figure 2. Axial view.

Clear cell sarcoma is an uncommon neoplasm first described by Enzinger in 1965 as a distinct type of soft tissue sarcoma [2]. The tumors are primarily diagnosed in the tendons or aponeuroses of the extremities. Because of its histological and immunochemical similarity to melanoma, it has also been called melanoma of the soft parts; like melanoma, the clear cell sarcoma is believed to originate from migrated neural crest cells, and hence, it has been thought that these tumors might

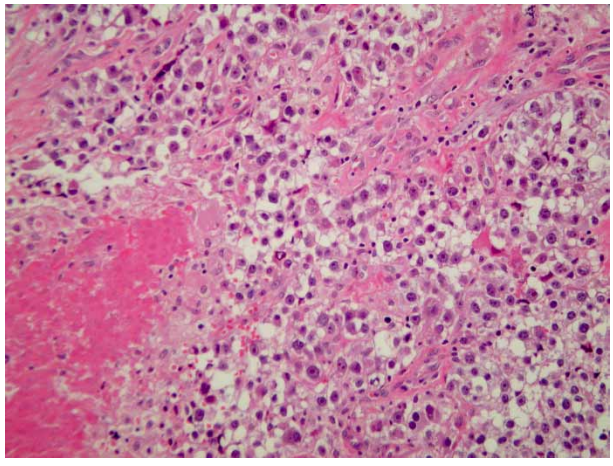

Figure 3. Paraspinal tumor. The clear cell sarcoma is comprised of polygonal cells with abundant clear to pale cytoplasm bordered by thin fibrous septae. A small amount of associated necrosis is seen in the lower left hand corner. (H & E section, 200X.)

respond better to biochemotherapy regimens used typically for malignant melanoma, rather than adriamycin and ifosfamide based regimens used more commonly in the treatment of soft tissue sarcomas [2–6]. Since 1965, less than five hundred cases have been reported, most of which have demonstrated an aggressive malignant behavior with rapidly disseminating disease.

The diagnosis of clear cell sarcoma can be challenging. Histologically, the clear cell sarcoma is characterized by fibrous septa and uniform cells which appear to have a clear eosinophilic cytoplasm, vesicular nuclei, and large basophilic nucleoli [7]. This has led to great difficulty in diagnosis due to a large differential that may include fibrosarcoma, malignant melanoma, synovial sarcoma, and, as with our case's initial diagnosis, renal cell carcinoma. Like lymphomas, however, many soft tissue sarcomas are characterized by translocations. Cytogenetic analysis has revealed a unique genetic marker in these tumors, a translocation from chromosome 12 to chromosome 22 [8–14].

The most effective treatment of a clear cell sarcoma is the complete surgical resection of the tumor. Like epithelioid sarcomas, angiosarcomas, and some liposarcomas, clear cell sarcomas are generally resistant to chemotherapy. The use of standard sarcoma regimens has not been successful. However, the remissions of two metastatic clear cell sarcomas after the use of interferon-alpha 2b (with chemotherapy in one case) have been reported [15–16]. Hence, interferon was included in the metastatic melanoma regimen we prescribed for our patient who had a transient, albeit complete response [17].

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