A Case of Malignant Melanoma of Soft Parts with Unusual Histopathologic Findings

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We report a case of malignant melanoma of soft parts presented with a hard mass on the dorsum of the left second toe. Malignant melanoma of soft parts has also been known as clear cell sarcoma of tendons and aponeuroses, which indicates the histologic feature of clear cell predominance and the origin of tumor cells, tendinous or aponeurotic structures. Although our case presented with many characteristic clinicopathologic features of malignant melanoma of soft parts, diagnosing this case as malignant melanoma of soft parts was made with great difficulty because of a rather unusual finding that the major cell component of neoplasm was spindle shaped cells in stead of clear cells.

Cases featuring this distinctive histopathologic finding have not been reported individually to our knowledge, but described briefly in the literature. We think this case deserves special attention because of its close resemblance to various sarcomas.

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Enzinger first described clear cell sarcoma of tendons and aponeuroses in 1965[1]. Although the histogenesis of this tumor is still not known exactly, there is little doubt that clear cell sarcoma is a tumor of neural crest origin, or soft tissue variant of malignant melanoma. So this tumor is now called by the term malignant melanoma of soft parts rather than clear cell sarcoma of tendons and aponeuroses[2,3]. Malignant melanoma of soft parts presents as a slowly enlarging mass on the extremities, especially the region of the foot and ankle. The histological findings of malignant melanoma of soft parts are usually characterized by tumor cells with clear, pale-staining cytoplasm and multinucleated giant cells[4].

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Several cases have been reported in Korea[5-9] under the title of clear cell sarcoma. we experienced an additional case of this rare tumor which showed unusual microscopic finding, that is, the paucity of clear cells.

REPORT OF A CASE

A 30 year-old-male patient visited our hospital because of hard mass on the dorsum of the left second toe which had been growing for 8 years. He experienced intermittent pain and tenderness in that area. On physical examination, he was found to have a hard elliptoid mass measuring 2 × 3cm and the skin over the mass was reddish, and keratotic(Fig. 1). He had no palpable lymph nodes at that time. The results of routine laboratory tests including complete blood count, liver function test urinalysis, and serologic tests were within normal limits or negative. An rhoentgenogram of the left foot showed soft tissue swelling and no

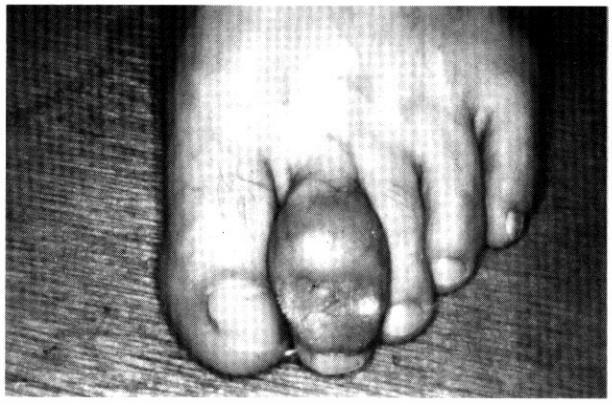


Fig. 1. A 2 × 3cm sized, hard elliptoid mass on the dorsum of the left second toe.

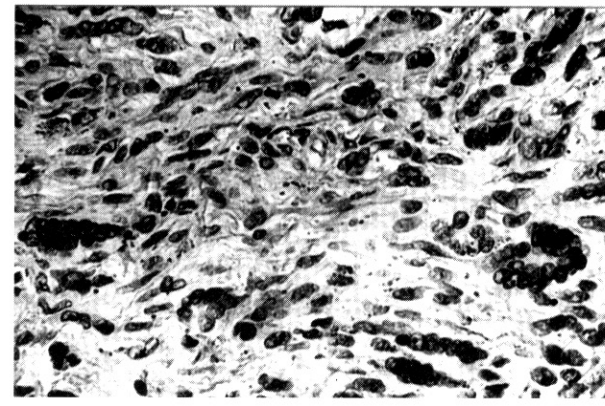
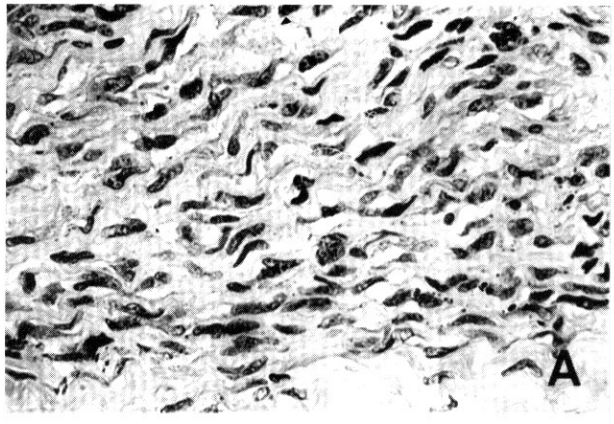


Fig. 3. The multinucleated giant cells having peripherally placed and irregularly aggregated nuclei(H&E, × 400).



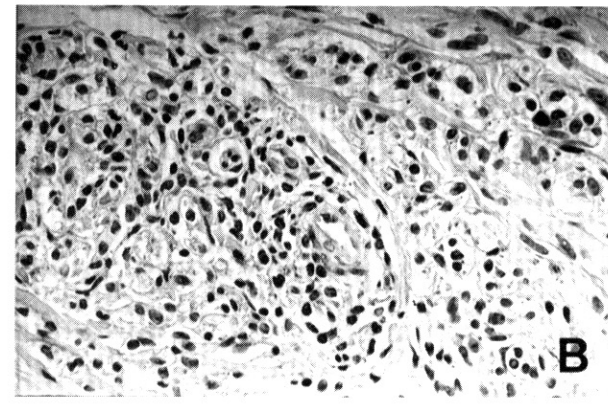


Fig. 2. The tumor was consisted of two types of tumor cells. One was spindle-shaped cells with hyperchromatic nuclei and eosinophilic cytoplasm(Fig. 2 A, H&E, × 200), the other was round or fusiform cells with a clear cytoplasm and vesicular nuclei(Fig. 2 B, H&E, × 400).

bony abnormality.

Grossly the excised tumor mass was firmly attached to the underlying tissues around the phalangeal bone. The routine histopathologic examination revealed massive proliferation of tumor cells with some heavily melanized foci in the whole dermis and deeper tissue. But there was no junctional activity in the dermoepidermal junction. The tumor mass had no encapsulation and was ill-defined with adjacent tissues. The involvement of skin appendages was not found. It consisted of two types of tumor cells. The spindle-shaped cells with hyperchromatic large nuclei and eosinophilic cytoplasm(Fig. 2 A), and round or fusiform cells with a clear cytoplasm and small hyperchromatic nuclei were observed(Fig. 2 B). Mitotic figures and cellular pleomorphism were rarely ob-

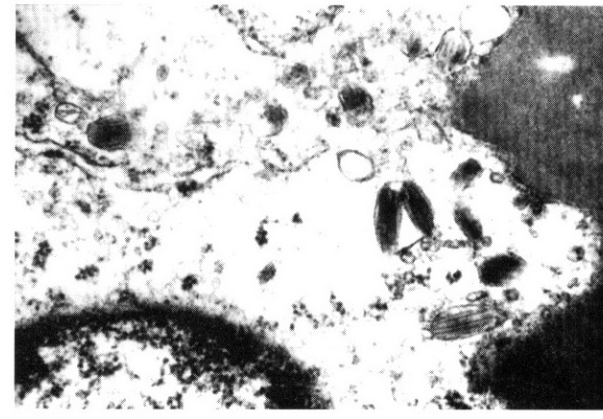


Fig. 4. Melanosomes in tumor cells could be observed by electon-microscopic examination(EM, × 30, 000).

served in both cell components. Only a small proportion of tumor cells, mainly clear cells, formed cell nests surrounded by a framework of fibrocollagenous tissue which was clearly shown by reticulin staining. There were some multinucleated giant cells(Fig. 3) having 8 to 15 peripherally placed or centrally aggregated nuclei. The Fontana-Masson staining demonstrated the presence of melanin pigments in some tumor cells. The immunohistochemical studies with anti-S-100 antibody and HMB-45 antibody showed positive results. On ultrastructural examination, melanosomes in varying stages of development, lack of fully melanized stage IV melanosome, were observed in the tumor cells(Fig. 4).

The patient was referred to the department of orthopedics and after the work-up for metastasis including careful physical examination and pelvic CT, bone scan, the disarticulation of metatarsophalangeal joint was carried out. The result of treatment is not available since he has missed the follow-up.

COMMENT

Malignant melanoma of soft parts occurs predominantly in young patients between the ages of 20 and 40, and is nearly always intimately associated with tendons or aponeuroses of the extremities with a predilection for the feet. The overlying skin tends to be uninvolved. The tumor presents as a slowly enlarging mass causing tenderness or pain in slightly more than 50% of patients[3,4]. The clinical findings of our case were very typical to those of malignant melanoma of soft parts.

Histopathologically, malignant melanoma of soft parts lacks junctional activity, and displays a uniform growth pattern characterized by tumor cells with a clear to granular eosinophilic cytoplasm that are arranged in nest-like aggregates[3,4]. The Fontana-Masson staining of the cases of malignant melanoma of soft parts ususally detect intracellular melanin in 50-70% of the cases. Immunohistochemically, the cells of nearly all cases express antigens for S-100 protein. Also melanosomes in varing stage of development are observed in most of the cases by electron microscope[4,10-12].

The histopathologic findings of our case disclosed many spindle-shaped cells with hyperchromatic nuclei and eosinophilic cytoplasm as well as a few foci composed of clear cells. The unusual proliferation of spindle-shaped cells in our case meant special effort was needed to differentiate it from other sarcomas, malignant schwannoma, and desmoplastic malignant melanoma. The results of routine and several special stainings, immunohistochemical studies and electronmicroscopic examination led us to the conclusion that this tumor should be a nevomelanocytic malignancy which had originated from soft tissue rather than skin. Many previously reported cases and original description emphasized the characteristic cellular morphology and used the term clear cell sarcoma, which has been replaced by the term, malignant melanoma of soft parts in recent reports. Our case shows that this rare tumor may be clear-cell predominant, as in previously reported cases, and spindle-cell predominant as in our case. So, the term clear cell sarcoma of tendons and aponeuroses should be discarded as the clear cell predominance is not a consistent finding.

In spite of adequate therapy, many patients with malignant melanoma of soft parts develop recurrences and metastases, sometimes 10 or more years after initial treatment. So long-term follw-up is mandatory[4].

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