CLEAR CELL SARCOMA WITH MELANIN

Report of Two Cases

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Two cases of clear cell sarcoma, one with many melanin pigmented cells and the other scanty, are described. Both are associated with tendons of the lower extremities of young Chinese adults. Their clinical behaviour differed vastly; one patient was dead seven months after local excision and block dissection of regional lymph nodes, while the other had a history of the growth of eight years prior to local excision and is well four years later.

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THIS TUMOR IS RARE AND THOROUGH STUDIES in the past[1–9] have revealed certain distinctive features for accurate typing of these tumors. It has close association with tendons and aponeuroses and the presence of melanin[1,2,5,8] in tumor cells in very occasional cases is puzzling. So far two typical cases have been encountered among Chinese and both contain melanin pigmented cells.

CASE REPORTS

Case 1

A 25-year-old Chinese male presented with a slowly enlarging mass in lateral aspect of right popliteal fossa for one year. The mass was firm, smooth, tender and measured 4 cm in diameter. It was attached to underlying muscle but not to the overlying skin. Two discrete, hard, mobile masses were also present in the right groin each measuring 2 cm across. At operation a fleshy mass 4 cm across was found in the musculotendinous junction of the biceps femoris within the muscle sheath. There was infiltration of adjacent muscle and tendon. The tumor was completely excised. Six weeks later, block dissection of the right inguinal lymph nodes was carried out.

The excised mass was firm, nodular, weighing 55 g. It was partly covered on one side by muscle. Cut surfaces showed mainly whitish tissue in which a broad tendinous band traversed the mass with fine branching fibrous septa. The right inguinal lymph nodes were enlarged, matted together and weighed 110 g. They were infiltrated by firm greyish tissue with foci of haemorrhage and necrosis.

Three months after the lymph node excision, a recurrent growth appeared in that area and a biopsy was taken. Following this a course of radiotherapy was given. The patient died of recurrent growths in the primary site and in the right groin seven months later and no autopsy was performed.

Case 2

A 17-year-old Chinese female complained of a swelling in the medial aspect of her left heel for 8 years. During the last six months, the mass gradually enlarged, painful and bluish in colour. On physical examination, a soft mass 2.5 cm across was found attached to her tendocalcaneus. The mass was excised completely. It measured 2.3 × 2 × 1.4 cm, firm and covered by a thin layer of fibrous tissue. She remains well up to now, i.e., 4 years after excision.

HISTOPATHOLOGIC FINDINGS

The primary growths in both cases show areas of compact nests and fascicles of fusiform cells separated by thin fibrous septa (Fig. 1). This pattern predominates in the tumor of case 2. Their nuclei are large, oval or slightly elongated with fine chromatin and distinct nucleoli. Mitotic figures are very scanty in Case 2 but in Case 1 they average 2 per high power field. In addition Case 1 reveals sheets and masses of polygonal tumor cells (Fig. 2) with nuclei that are large, pleomorphic and hyperchromatic or vesicular with prominent nucleoli. The cytoplasm is moderate in amount and ranges from eosinophilic to clear. The latter contains glycogen. In scattered areas of the tumor, few multinucleated giant cells are seen with nuclei arranged in a complete ring around the periphery (Fig. 3).

Melanin pigmented tumor cells are plentiful

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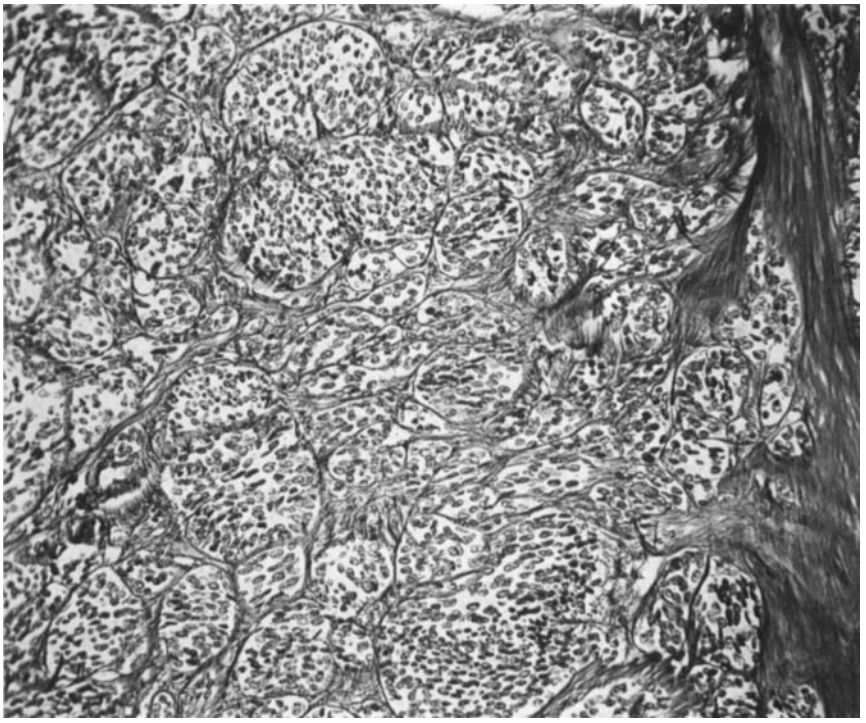

FIG. 1. Case 1: Compact fascicles of tumor cells separated by collagenous strands (modified Gordon and Sweet's method for reticulin ×150).

in Case 2 but scanty in Case 1 (Fig. 4). Most of the melanin pigment is seen in the fusiform cells. The pigment reacts positively to Masson Fontana and Schmorl's stains. This reaction is abolished after preliminary bleaching with potassium permanganate and oxalic acid.

Lymph node metastases of Case 1 show sheets and masses of tumor cells varying from irregular

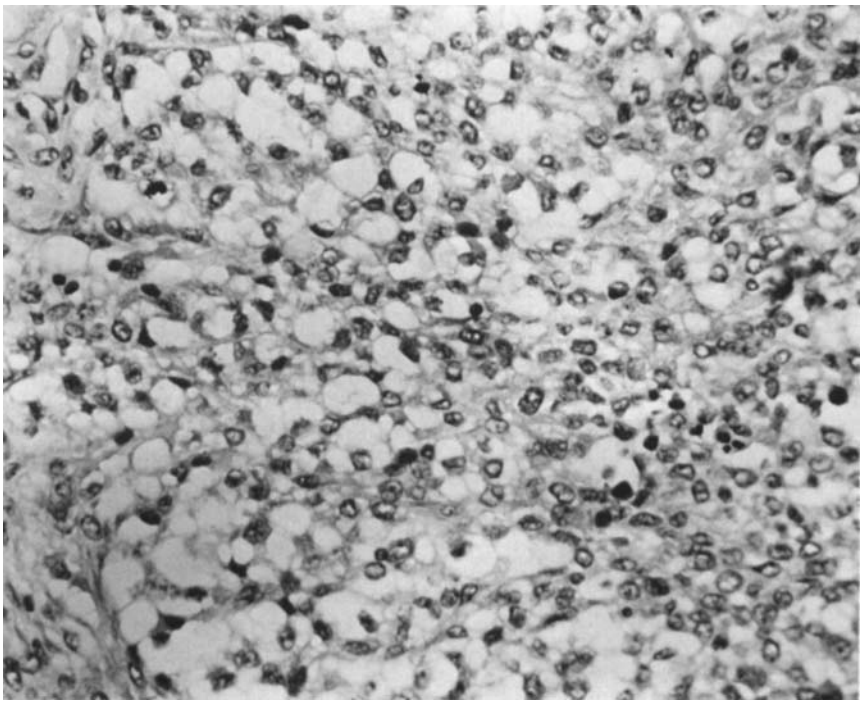

FIG. 2. Case 1: Sheet of tumor cells with eosinophilic or clear cytoplasm (H&E, ×300).

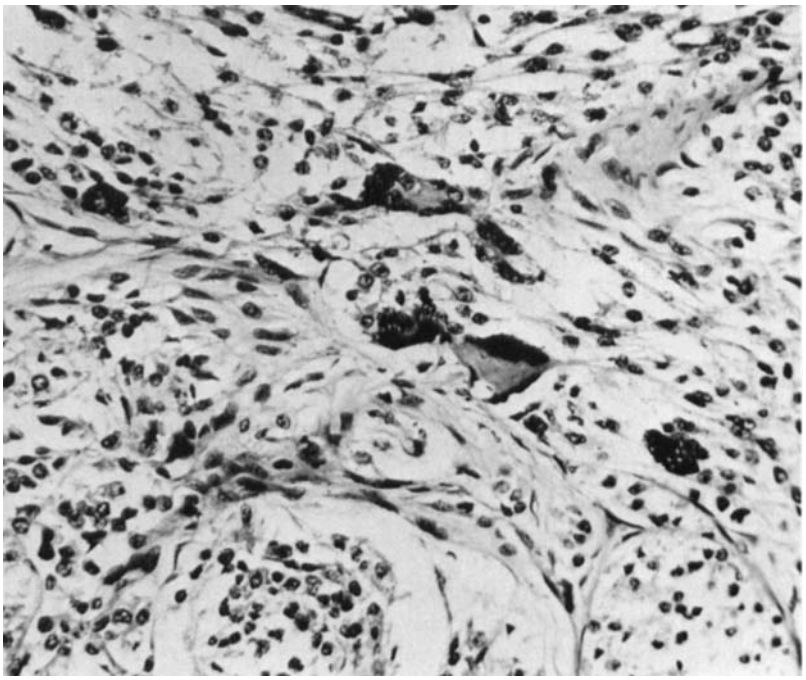

FIG. 3. Case 1: Giant cells with peripheral nuclei (H & E, ×300).

polygonal to slightly elongated in shape. Their nuclei are pleomorphic, with coarse chromatin pattern and some with distinct nucleoli. Serial sections of blocks from the lymph node metastases and special stains reveal no melanin in any tumor cells. Fine reticulin fibrils are present in between masses of metastatic tumor cells.

DISCUSSION

This group of malignant tumors is usually slow growing and of uncertain histogenesis. Neuroectodermal origin has been suggested. Electron microscopy demonstrates melanosomes in tumor cells indicating that the tumor is

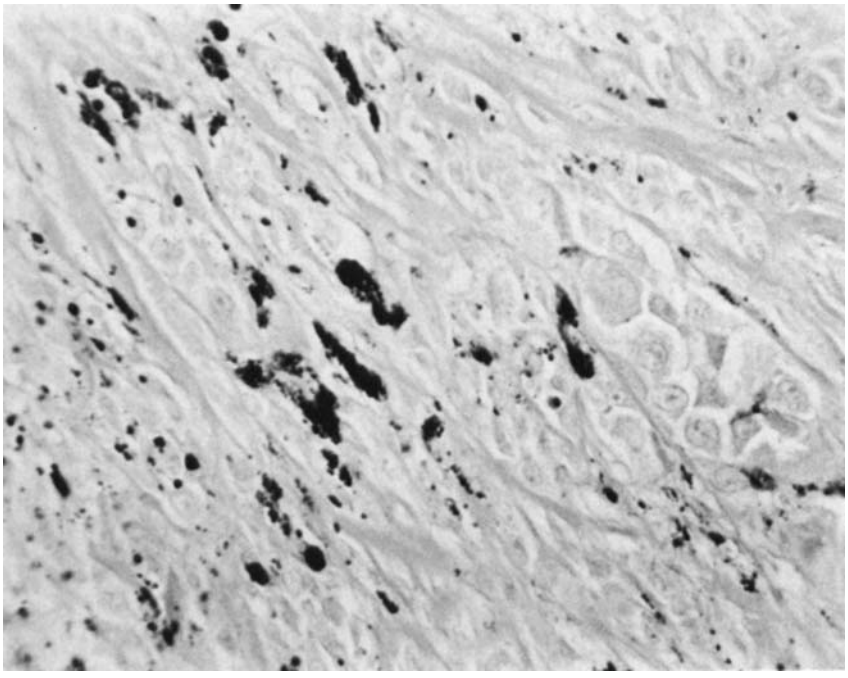

FIG. 4. Case 2: Abundant melanin pigment in tumor cells (Schmorl's stain ×500).

related to malignant melanoma.[1,5] Our Case 2 with many melanin pigmented cells in the growth favors this possibility. Other histological features, namely, areas of plump spindle cells arranged in nests and fascicles, sheets of polygonal cells with eosinophilic or clear cytoplasm are distinctive characters of clear cell sarcoma and important criteria for histological typing. Furthermore, these two tumors are closely associated with tendons.

Melanin pigment, if present, together with scattered multinucleated giant cells with nuclei arranged in a complete ring around the periphery is an additional feature helpful in confirming the diagnosis. Differentiation from a metastatic renal cell carcinoma and other clear cell tumors can be difficult if the growth shows predominantly clear cells with little or none of the characteristic cell types and pattern mentioned above.

It is significant that the lymph node metastases of Case 1 show predominantly sheets and masses of irregular polygonal or slightly elongated tumor cells without fascicular pattern or melanin pigmented cells which are present in the primary. Tumor cells in metastases appear less differentiated than the primary and this is also observed by other authors.[4,5,8] Thus, if only the metastases are biopsied, the histology will provide no clue as to the nature of the primary.

In Enzinger's series of 21 cases,[4] the youngest patient at the time of operation was a 13-year-old boy and the oldest was a 65-year-old woman. Most of his cases were young adults, more among females and usually in the lower extremities. The present two cases are young Chinese adults, one female and one male and both tumors are found in the lower extremities.

Case 1 died of recurrence seven months after the first operation and Case 2 remains well up to now, *i.e.*, four years after local excision. For recurrent growths Enzinger stated that in nearly all instances the growths reappeared within a year.[4] This is true of Case 1. For Case 2, taking into account the first appearance of the growth and the follow-up period, a total of 12 years have elapsed. There is still a remote possibility of metastasis as Enzinger had one case with metastases after 30-year interval.[4] This case, however, had repeated surgical procedures which up to now are lacking in our Case 2. Thus the two cases described above with diverse outcome represent two ends of a wide spectrum of clinical behavior of clear cell sarcoma.

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