Acta orthop. scand. 48, 241–244, 1977

CLEAR-CELL SARCOMA OF TENDONS AND APONEUROSES

Report of Two Cases

KARI ALITALO, PEKKA PAAVOLAINEN, KAARLE FRANSSILA & VEIJO RITSILÄ

Division of Plastic Surgery, Helsinki University Central Hospital, and Department of Pathology, University of Helsinki, Finland.

Two cases of clear-cell sarcoma of tendons and aponeuroses are reported. The first patient was a 19-year-old man with a tumor in the medial collateral ligament of the knee. The tumor was enucleated, and after postoperative radiotherapy a wide local excision was done. The patient was symptomless 4½ years after the operation. The other patient was a man aged 68 with a tumor between the medial malleolus and the Achilles tendon. He also received postoperative radiotherapy after enucleation. Three months later a metastasis was noticed in the ipsilateral para-iliac nodes. The patient died 16 months after the operation. The histology and clinical course of the two cases are analyzed. Some integrative remarks based on earlier reports of 33 cases and the present two cases are presented.

Key words: clear-cell sarcoma; malignant tumor of tendons and aponeuroses; soft somatic tumor of the extremities

Accepted 21.i.77

CASE REPORTS

Case 1: A 19-year-old man was admitted to hospital because of a tumor of the left knee. He gave a history of two episodes of mild trauma to the knee during his military service, 6 and 2 months prior to admission. After the second injury, the knee was painful during exercise, and the patient discovered a small nodule on the medial side of the knee.

On examination, both the function and X-rays of the knee were normal. The only finding was a small tender nodule under the skin on the medial side of the knee, which was thought to be a cyst of the medial meniscus. At operation in March 1972, the medial meniscus appeared normal, but a nodule about 1 cm in diameter was found in the medial collateral ligament. This light-brown tumor was easily enucleated. Microscopy revealed a clear-cell sarcoma of tendons and aponeuroses.

The operated region was treated by tele-cobalt therapy (3116 R over a period of 24 days), followed by a wide local excision of the medial joint capsule. During this operation, no signs of tumor were noted. After 2 weeks, another dose of 3136 R for 3 weeks was given to the same area.

A radio-ulcer developed on the medial aspect of the knee. The ulcer was closed with an abdominal tubed pedicle flap carried via the wrist and the right knee.

When last examined in November 1976, 4 years and 7 months after the operation, no signs of metastases or local recurrences were observed.

Case 2: The patient was a man aged 68 with a history of relapsing erysipelas on both legs. One month before surgery he observed a firm, movable nodule on the dorsal aspect of his right ankle. During the following weeks the tumor grew in size, and the patient reported slight pain when walking. There was no history of trauma to the ankle.

On examination, a firm, ovoid, movable mass, 4 cm in diameter, was palpated beneath the skin

of the medial calcaneal region. No destruction of bone could be demonstrated radiologically, and the function of the ankle joint was normal. Clinically the tumor was regarded as a fibroma.

An extirpation was done in June 1974. The solid, well-demarcated tumor was situated between the medial malleolus and the Achilles tendon, with no invasion of surrounding tissue. It was surrounded by dense fibrous tissue. The lesion probably originated from the Achilles tendon sheath. The tumor was light-brown colored, and the cut surface gave the impression of a benign fibroma or leiomyoma. However, the histological diagnosis was clear-cell sarcoma of tendons and aponeuroses.

Postoperative tele-cobalt therapy up to a tumor dose of 6000 R for 7 weeks was given. Three months later lymphography demonstrated metastases in the right para-iliac nodes. This was verified histologically, and after extirpation the regional nodes were treated by a fotone dose of 5000 R for 2 months. Thereafter, the patient was symptomless for 9 months. He died in November 1975, 17 months after the diagnosis. At autopsy, bronchopneumonia and metastases in lungs and adrenals were found.

HISTOLOGICAL FINDINGS

The staining methods used were haematoxylin and eosin, Weigert-van Gieson, Alcian blue, Periodic acid-Schiff reaction (PAS) with and without diastase, Gömöri's reticulum stain, Gömöri's method for iron and Fontana method for melanin.

The microscopic examinations revealed the tumors to be non-encapsulated, but their exact relation to the surrounding tissues could not be evaluated because both tumors had been removed by enucleation. The tumors were composed of solid nests and fascicles of cells surrounded by connective tissue septa. The septa were mostly thin and delicate but sometimes the cell groups were separated by thick and dense fibrous areas (Figure 1). Reticulum fibers were seen around the cell nests, but they were scarce or absent between individual tumor cells (Figures 2 and 3). The tumor cells were either round or spindle-shaped. The nuclei were large, pale-staining and round with prominent basophilic nucleoli. In case 1, some giant cells with multiple uniform nuclei were observed. Mitotic figures were few, about 1–4 per 10 high-power fields. The cytoplasma was abundant and pale-staining, usually clear and often filled with tiny vacuoles (Figure 4). In case 2, small amounts of diastase-sensitive PAS-negative material was found in the cytoplasm of some tumor cells. No intra-or extracellular Alcian blue-positive mate-

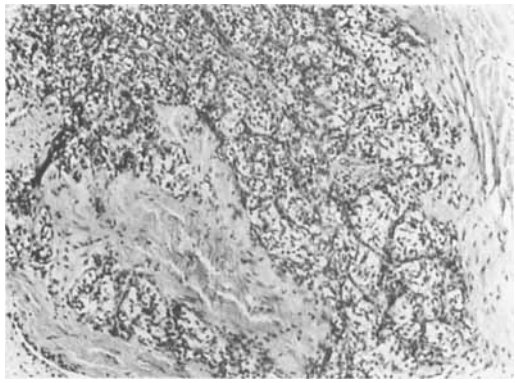

Figure 1. Clear-cell sarcoma. Case 1. The tumor is composed of solid nests of cells surrounded by delicate connective tissue septa. Also dense fibrous areas are seen between the nests. (H and E, × 50).

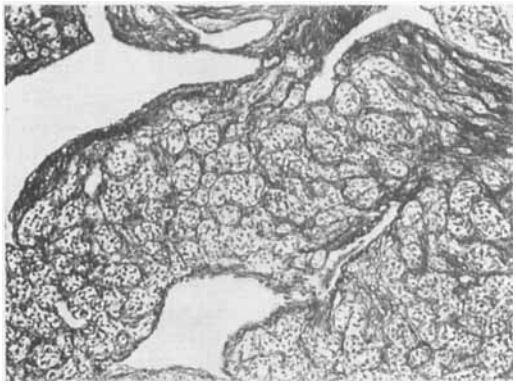

Figure 2. Clear-cell sarcoma. Case 1. Reticulum fibers surround cell nests (Gömöri's reticulum stain, × 50).

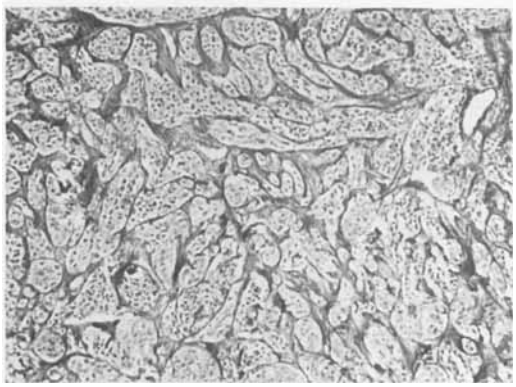

Figure 3. Clear-cell sarcoma. Case 2. Both thin reticulum fibers and thicker collagenous septae surround cell nests. (Gömöri's reticulum stain, × 50).

rial was observed. In both cases, both intra- and extracellular iron was present, but no Fontana-positive material was found. The metastatic tumor in case 2 did not differ histologically from the primary tumor.

DISCUSSION

The two tumors presented here fulfil the histologic criteria of the clear-cell sarcoma of tendons and aponeuroses presented by Enzinger in 1965. The most important of these criteria are the typical reticulum pattern, the clear cytoplasm of the tumor cells and the large pale nuclei with a prominent nucleolus. Histologically it may be difficult to differentiate this tumor from synovial sarcoma, alveolar soft part sarcoma, chemodectoma, malignant melanoma, or metastasis from a clear-cell carcinoma. The tumors presented here did not have the pseudoglandular pattern of synovial sarcoma. Nor did the cytoplasm of the tumor cells reveal PAS-positive, diastase-resistant granules typical of alveolar soft part sarcoma. In chemodectoma, the septa surrounding the cell groups contain blood vessels, but these were not seen in the present cases. Fontana-positive pigment, which is usually present in malignant melanoma, was not found in these two tumors. Nor was any other tumor found in the skin or elsewhere which could have transferred metastases. Consequently the histological diagnosis made seems to be correct.

MacKenzie (1974) has described one and Bearman et al. (1975) two clear-cell tumors originating from tendon which contained melanin pigment. Electron-microscopy showed melanosomes in the cases described by Bearman et al. The tumors thus represented melanomas with an apparent soft tissue origin. No melanin has been mentioned in the other reported cases. In a case report by Hoffman & Carter (1973), the primary tumor showed the features of clear-cell sarcoma but in a recurrent tumor and in metastases 3 years later melanin pigment was seen. It is possible that some of the cases reported represent amelanotic melanomas.

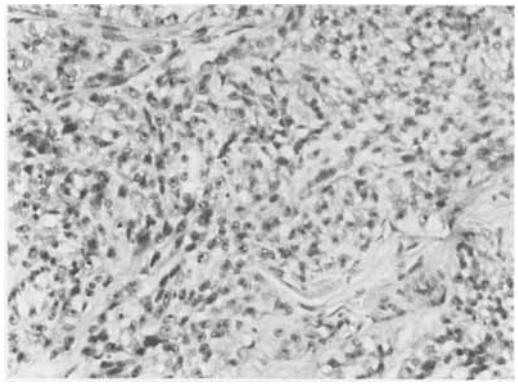

Figure 4. Clear-cell sarcoma. Case 2. The nuclei are large and pale with prominent nucleoli. The cytoplasm is clear and abundant and filled with vacuoles of varying size. (H and E, × 150).

Although it is possible that clear-cell sarcoma of tendons and aponeuroses is not histogenetically a homogenous group it seems to be a rather homogenous group clinically. Therefore, some integrative remarks based on earlier reports (Bennett 1947, Guichard et al. 1968, Rousselot et al. 1968, Kubo 1969, Vuillard 1969, Dutra 1970, Jung et al. 1970) and the present two cases are presented here (a total of 35 cases; cases with melanin pigment were not included).

The median age of the patients at diagnosis was 32 years (range 12–68 years). Males and females were equally represented (17 males, 18 females).

At the time of the first operation, all tumors were solitary. The diameter of the tumor varied from 1 to 6 cm. The topographic distribution of the primary tumor is shown in Table 1.

Metastases were observed in 18 of the 30 cases with a follow-up of at least 1 year. They were located most frequently in regional lymph nodes and lungs; also metastases in bone, liver, heart and adrenals were observed. The median interval between the first operation and

the occurrence of the first metastasis was 4 years (range 1 month to 30 years).

Clear-cell sarcoma seems to be especially prone to recur; in the 30 cases with a follow-up of at least 1 year recurrences were observed in 20 cases. This may be partly explained by the rather conservative surgical treatment due to the benign, well-demarcated appearance of the tumor at operation.

Table 1. Anatomical distribution of the 35 clear-cell sarcomas reported.

Location	No. of patients
Lower extremity:	
Planta pedis	6
Achilles tendon	4
Knee	5
Ankle	4
Patellar tendon	3
Heel	1
Buttock	1
Toe	2
Upper extremity:	
Finger	5
Palm	1
Forearm	2
Triceps	1
Total	35

The mortality rate in clear-cell sarcoma is high; 18 out of the 30 patients with a follow-up of at least 1 year have died. The actuarial 5-year survival rate was about 50 per cent. Although the mortality rate is high the growth of the clear-cell sarcoma seems to be rather slow. The duration of symptoms prior to the treatment was less than 1 year in 13 cases, 1 to 4 years in 11 cases and more than 5 years in 8 cases (maximum 19 years); no data were available for three patients. Three deaths from cancer occurred more than 10 years after treatment (11, 12 and 36 years later).

Thirty-two of the cases were primarily treated by local excision. For 25 patients this was the only treatment during the first stage. This treatment was followed by radical operations in 15 cases (extensive local excisions, 9 amputations). Only 10 patients received radiotherapy. The different forms of therapy did not seem to differ in therapeutic value. However, if local excision is not adequate, recurrence is the rule. No significant differences appear between patients treated early or those treated after several recurrences.

ACKNOWLEDGMENTS

We would like to thank docent Börje Sundell, M.D., for his help in the preparation of this paper.

REFERENCES

Bearman, R. M., Noe, J. & Kempson, R. L. (1975) Clear-cell sarcoma with melanin pigment. *Cancer* **36**, 977–984.

Bennett, G. A. (1947) Malignant neoplasms originating in synovial tissues (synoviomata). *J. Bone Jt Surg.* **20**, 259–291.

Dutra, F. R. (1970) Clear-cell sarcoma of tendons and aponeuroses. *Cancer* **25**, 942–946.

Enzinger, F. M. (1965) Clear-cell sarcoma of tendons and aponeuroses. *Cancer* **18**, 1163–1174.

Guichard, A., Feroldi, J. & Lamy, M. (1968) Le sarcoma alveolaire à cellules claires des tendons. *Ann. Anat. Path.* **13**, 11–20.

Hoffman, G. J. & Carter, D. (1973) Clear-cell sarcoma of tendons and aponeuroses with melanin. *Arch. Path.* **95**, 22–25.

Jung, A., Kehr, P. & Vuillard, N. (1970) Le sarcoma à cellules claires des tendons et des aponévroses. *Rev. Chir. Orthop.* **56**, 553–562.

Kubo, T. (1969) Clear-cell sarcoma of patellar tendon studied by electron microscopy. *Cancer* **24**, 948–953.

MacKenzie, D. H. (1974) Clear cell sarcoma of tendon and aponeuroses with melanin production. *J. Path.* **114**, 231–232.

Rousselot, P., Weill-Bousson, M., Lefakis, P. & Wetter, J.-M. (1968) Le sarcome à cellules claires des tendons et aponévroses. *Ann. Anat. Path.* **13**, 325–334.

Vuillard, N. (1969) Contribution à l'étude des tumeurs malignes des gaines tendineuses: le sarcome à cellules claires. Thesis, Strasbourg.

Correspondence to: Veijo A. Ritsilä, M.D., Rantapolku 2 d F, SF-02170 Espoo 17, Finland.