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SCLEROSING EPITHELIOID FIBROSARCOMA; A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT... Sclerosing epithelioid fibrosarcoma (SEF) is a rare subtype of Fibrosarcoma and was first reported in 1995. This tumour is rarely encountered and can easily be mistaken by clinicians for other diagnoses. This paper reports one case of SEF and review related literature. **Objectives:** To determine the clinical pathologic characteristics and discuss differential diagnosis of sclerosing epithelioid fibrosarcoma (SEF). **Methods:** One case of SEF was studied by clinical analysis, light microscopy and the review of the literature. **Results:** The patient was an adult and had a tumor located in the subcutaneous tissue of the left leg, which was painless and grew slowly. Macroscopically, it was described as nodular and non-encapsulated. The cut section was gray-white, firm to elastic in consistency. The microscopic examination showed that the round to ovoid epithelioid cells with clear or eosinophilic cytoplasm arranged in cords, nests, sheets or alveolar pattern. The stroma showed a dense hyalinized collagenous background. **Conclusions:** Sclerosing epithelioid fibrosarcoma is a low-grade variant of fibrosarcoma, histologically, it should be differentiated from a variety of tumors which have the epithelioid appearance and sclerosing stroma.

Key words: Sclerosing epithelioid fibrosarcoma; Clinicopathological Features; Differential diagnosis; Light microscopy

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INTRODUCTION

Sclerosing epithelioid fibrosarcoma (SEF) is a rare subtype of fibrosarcoma. Meis-Kindblom and other authors first reported it in 1995¹. Histological features of this tumor are the epithelial cells distributed among a large number of hyaline collagen fibers. This tumour is rarely encountered and can easily be mistaken by clinicians for other diagnosis. This paper reports one case of SEF and review related literature. Clinical pathological features, differential diagnosis and biological behavior will be discussed.

CASE REPORT

A 54-year-old female presented with painless mass on the left leg for about 1 year.

She was admitted to the hospital for treatment in March, 2014. Physical examination showed normal temperature, negative cardiopulmonary symptoms, soft and non tender abdomen and there was no hepatosplenomegaly. The rest of the medical history of the patient was

uneventful. During surgery the tumor was situated in the subcutaneous tissue and measured 1.5cm×1cm×1cm in size and the margins of the tumor were clearly defined.

The specimen was fixed in 10% neutral buffered formalin and paraffin embedded. Microtomy was done and 4um sections obtained, mounted on glass slide and followed by hematoxylin-eosin staining thereafter and examined under a microscope.

Macroscopically it was a nodular mass with no capsule, the size was about 1.5cm × 1cm × 1cm, attached to a little skin, and the area of the skin was 2cm×1.5cm. The cut section was gray-white, firm to elastic in consistency with no significant necrosis and calcification.

Microscopically the tumor was located in the subcutaneous tissue with clear Margin (Figure 1). The tumor cells were uniform, small to medium size, round to ovoid epithelioid cells, arranged

in cords or nests pattern, (Figure 2) and part of the area showed alveolar pattern (Figure 3), distributed in a large number of hyaline collagen fibers (Figure 4). High-powered magnification showed clear cytoplasm, and part of the tumor cells with eosinophilic cytoplasm (Figure 5). The characteristic of the nucleus was uniform nuclear chromatin and inconspicuous nuclear atypia. The nucleolus was obvious and centered, and the mitosis was easily to seen (Figure 6).

The Pathological Anatomical diagnosis was Left leg; Sclerosing epithelioid fibrosarcoma.

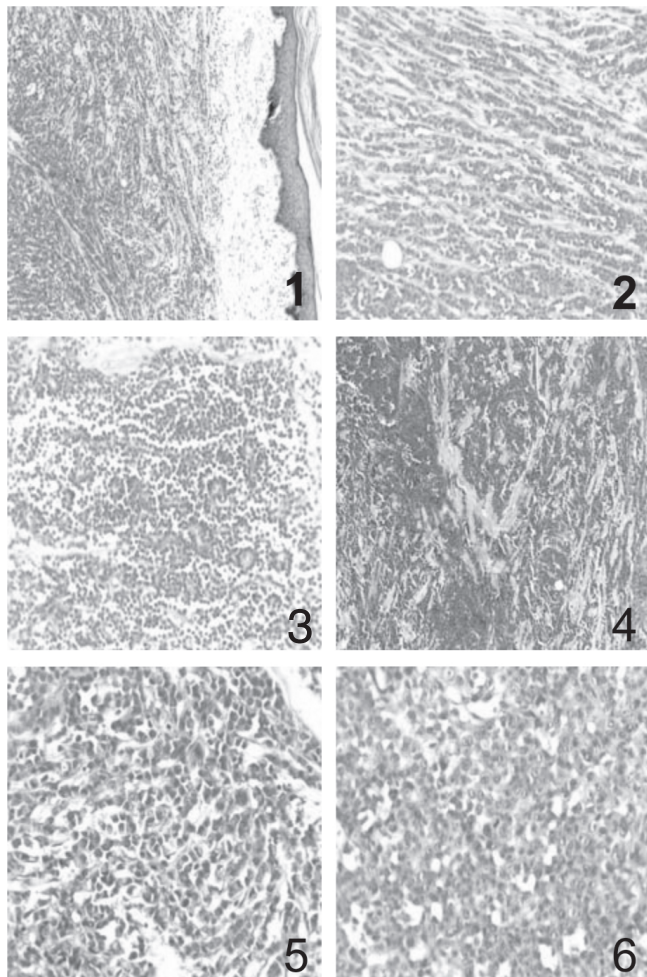


Figure-1: The tumor was located in the subcutaneous tissue with clear Margin.

Figure-2: The tumor cells arranged in cords or nests Pattern.

Figure-3: Part of the area show alveolar pattern

Figure-4: A large number of hyaline collagen

fibers in the stroma.

Figure-5: Eosinophilic cytoplasm.

Figure-6: Obvious and centered nucleolus, mitosis was seen

DISCUSSION

Clinical features

SEF often occurs in adults. The age of the patient varies from 14 to 87 years old, and the median age is 45 years². There isn't a significant gender difference in disease incidence³. The tumor is often located in the deep muscle tissue of the lower extremities, but in this case it was on superficial site. Other sites are trunk, upper limbs, head and neck, etc. Rare areas include the abdominal cavity, pelvic cavity, intracranial, penis, ilium and sacrum³. Clinically, most patients show partial slowly enlarging mass, from several months to several years, and in some patients it is associated with pain.

Imaging Features

CT or MRI scan often shows low signal intensity, which implies that the tumor contains more collagen components, however, the cell component is relatively small⁴.

Pathological features

Grossly the mass is nodular or lobulated with no capsule, measuring 2 to 22cm diameter. The cut section is usually grey-white, sometimes with myxoid or cystic degeneration, however, calcification and necrosis are rare. Microscopy shows that the tumor cells are small to medium size, round or oval, clear cytoplasm, like epithelial cells. Nuclear chromatin is uniform, and nuclear atypia is not obvious. Mitosis is rare. The tumor cells are arranged in cords, nests or alveolar pattern. The collagen fibers significantly show hyaline degeneration. There are cartilage, bone and nerve sheath differentiation in a few cases^{5,6}. The histologic features of this case was in accordance with most of the above features, in addition, the cytoplasm of some cells was eosinophilic, the nucleolus was prominent and mitosis was easily seen (8 / 10HPF).

Ultrastructure and genetic characteristics

Electron microscopy shows that the rough endoplasmic reticulum and golgi complexes contain collagen secretory granules and a large number of intermediate filaments in the tumor cells. No basement membrane surrounding the cell implies the differentiation of fibroblasts⁷. The cytogenetic changes of chromosome in SEF include the rearrangement of 10p11 and the amplification of 12q13 and 12q15, and it also involves HMGIC gene⁷. The latest research shows that some of the SEF patients have the FUS gene rearrangement which occurs in the low-grade malignant fibrous myxoid sarcoma. This feature suggests that they may be linked closely; thus, some scholars consider that part of the SEF may be a variant of low-grade malignant fibrous myxoid sarcoma, not a unique fibrosarcoma variant^{8,9,10}.

Differential Diagnosis

(1) Metastatic carcinoma: The invasive lobular carcinoma of the mammary gland or signet ring cell carcinoma of the digestive tract, but the patient had no history of cancer.

(2) Solitary fibrous tumor (SFT): This tumor also contains large amounts of rope-like collagen fibers, but a variety of histological structure can be seen, including patternless pattern, storiform and hemangiopericytoma-like structure. Besides, the shape of collagen in the stroma is varied, in addition to the rope-like, the asbestos-like or irregularly shaped collagen stroma can also be seen. Additionally, the vessel wall in the tumor is often accompanied by hyaline degeneration.

(3) Sclerosing rhabdomyosarcoma: This tumor consists of more primitive differentiation of small round cells, spindle cells or polygonal cells. The nucleus is hyperchromatic, and the nuclear shape is irregular. Spider web-like or ribbon rhabdomyoblasts which contain stripes are visible in some cases.

(4) Synovial sarcoma: This tumor often occurs near the large joints, mostly accompanied by spindle cells and epithelioid cells two-way differentiation. Even a one-way differentiation,

there is no hyalinization of collagen fibers in the stroma.

(5) Epithelioid leiomyosarcoma: The cytoplasm of tumor cells are rich, lightly stained and transparent, and part of them are dark eosinophilic. The tumor cells are more pleomorphic. But this case hasn't obvious cell atypia.

(6) Clear cell sarcoma of soft tissue: The tumor cells are rich and mostly arranged in nests pattern. It is separated by fibrous connective tissue between nests, but no obvious sclerosing stroma.

Biological behavior and treatment

Compared to the classic fibrosarcoma, most scholars think that sclerosing epithelioid fibrosarcoma belongs to a low grade malignant fibrosarcoma. Meis-Kindblom et al¹ reported that the recurrence rate was 57%, the recurrence time was 2.3-11 years after operation, and the transfer rate was 43%, the transfer time is 4.7-14 years after surgery, the mortality was 25%, the time of death is 3.1 to 13.7 years after surgery. But Antonescu⁷ reported that its clinical manifestations were similar to the moderate malignant sarcoma, and the case which was accompanied by nerve infiltration with worse prognosis. SEF should be locally excised, if necessary and supplemented by postoperative radiotherapy. Close follow-up should be done in order to detect the possibility of local recurrence and distant metastasis.

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“The future belongs to those
who prepare for it today.”

Malcolm X

