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Outcomes of surgical treatment alone in elder patient with classic-type epithelioid sarcoma. Case report

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Abstract

Epithelioid sarcoma (ES) is an extremely rare malignant soft tissue tumor that has a known propensity for local recurrence, regional lymph node involvement, and distant metastases. It is a slow-growing tumor occurring mainly in young adult males, with a predilection for distal extremities, particularly in the hand (the fingers) and foot. Its clinical and histological characteristics resemble those of various benign and malignant conditions and its differential diagnosis from other forms of cancer is required through various immunohistochemical stains. Although a multidisciplinary approach is essential, surgical resection is the mainstay treatment of ES, eventually combined with neoadjuvant or adjuvant radiotherapy or chemotherapy.

Here, we describe a relatively rare presentation of classic-type ES in the elder patient. We are reporting the application of surgical treatment alone with excellent both functional and cosmetic results.

Keywords: Classic-type epithelioid sarcoma; Surgical treatment; Rotation flap

1. Introduction

Epithelioid sarcoma (ES) is an extremely rare (1%) malignant soft tissue tumor that has a known propensity for local recurrence, regional lymph node involvement, and distant metastases. It is a slow-growing tumor occurring mainly in young adult males in the distal extremities (classic-type ES) (Chase and Enzinger, 1985).

Recently, the proximal variant of ES has been reported to be a more aggressive subtype (Asano et al., 2015).

The treatment of choice is wide resection and chemoradiotherapy (Miettinen et al., 1999). Nevertheless, this type of sarcoma has a poor clinical outcome and a high rate of local recurrence.

Few reports have been published about ES in elderly patients. Here, we present a case of indolent classic distal-type ES in a 76-year-old male, treated with complete surgical non-mutilating resection alone resulting in optimal function preservation of the upper limb with emphasis on interdisciplinary approach.

2. Case report

A 76-year-old male came to our department with asymptomatic swelling on the ulnar side of dorsal surface

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of the left hand since 10 years. The swelling was nodular with ill-defined margins, 3 × 2 cm, firm with central ulceration and yellowish crust (Fig. 1A). There was neither regional lymphadenopathy nor alterations of routine hematological and biochemical parameters.

A biopsy, previously performed elsewhere, revealed proliferation of epithelioid cells with clear or weakly basophilic sometimes vacuolated cytoplasm. The specimen was immunohistochemically positive for Actin ML, CDM 5.2, CD34 and ERG and completely negative for Melan A, S100 and CD 31. The diagnosis of a cutaneous epithelioid vascular proliferation has been suggested.

We performed radical tumor excision, resulting in a 5.0 cm wide defect over the dorsum of the left hand (Fig. 1B). We decided to use a rotation flap to close the surgical wound preserving the underlying extensor tendons, vessels and nerves as much as possible. The flap was sutured in place with 4/0 polyamide suture (Fig. 1C).

To facilitate the sliding of the flap and avoid a "dog-ear" effect, two small triangles of skin were excised on the distal side of the secondary defect. The sutures were removed 15 days later, and the 1-month follow-up visit revealed both esthetically and functionally excellent results (Fig. 1D).

The pathological diagnosis was ES formed by a relatively monomorphic population of epithelioid cells with moderate cytologic atypia and rare mitotic figures, separated by abundant collagen stroma and ulcerated epidermis (Fig. 2A, a). The immunohistochemical study, in addition to the positivity for Actin ML, CAM 5.2, CD34 and ERG reported in the previous biopsy and confirmed in the current one, results were positive for epithelial membrane antigen (EMA), AE1-AE3 e D2-40 with loss of INI-1 in most part of the cells (Fig. 2B–D). Multiple sections was taken for margin assessment. The surgical margins were microscopically negative. TC total body was negative at base-line and at 6 months after tumor excision.

The patient maintains regular follow-up visits at 3, 6 and 9 months with no recurrence of the tumor.

3. Discussion

ES, first described by Enzinger in 1970, is a rare malignant soft tissue tumor that generally appears in fascial planes, aponeuroses, and tendon sheaths of the extremities, particularly in the hand and foot (Chase and Enzinger, 1985). Its prevalence is high among young adults (20–40 years), but it rarely was found in children and older people (Casanova et al., 2006). Its clinical characteristics are similar to many other pathological conditions, including chronic granulomatous inflammation, nodular fasciitis, and synovial sarcoma (Miettinen et al., 1999).

Classic-type ES has some typical features, such as location at superficial distal sites an indolent growth rate, a tendency toward locoregional recurrence, with multiple or confluent nodules and plaques along an extremity

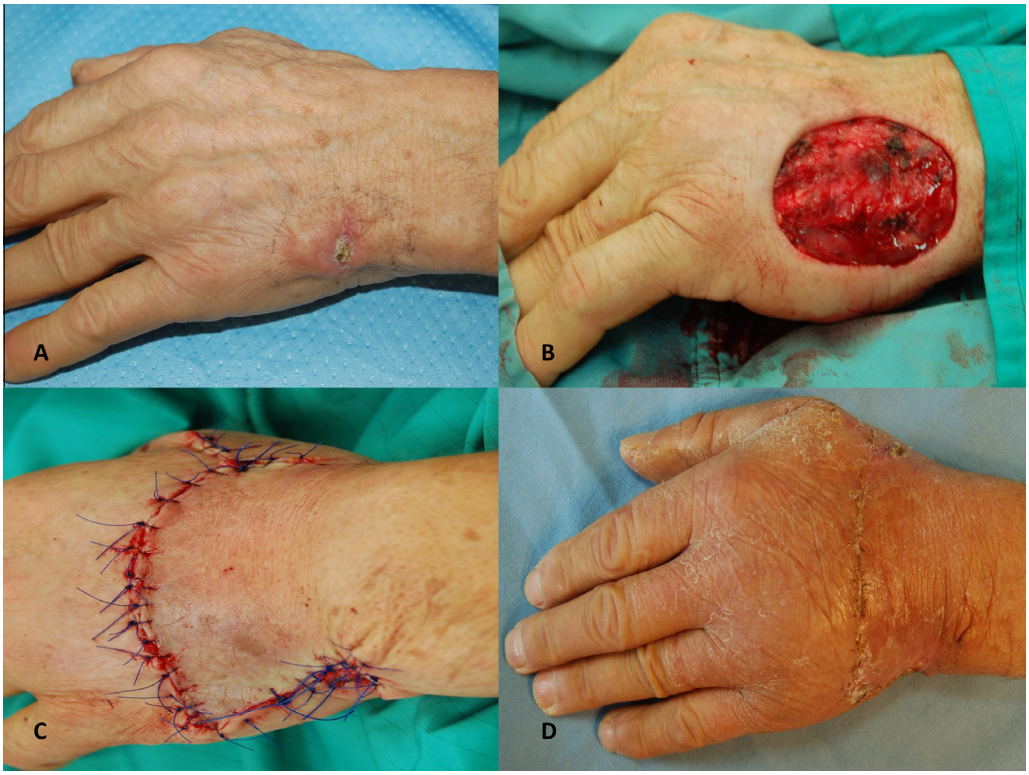

Figure 1. Clinical appearance of the epithelioid sarcoma located on the back of the left hand over the fifth metacarpal (A). Round defect following the excision of the tumor (B). The rotation flap delineated with a semicircular curve and raised while preserving the dorsal nerves and veins and sutured in position (C). The end result of the flap, 15 days after the removal of sutures (D).

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