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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

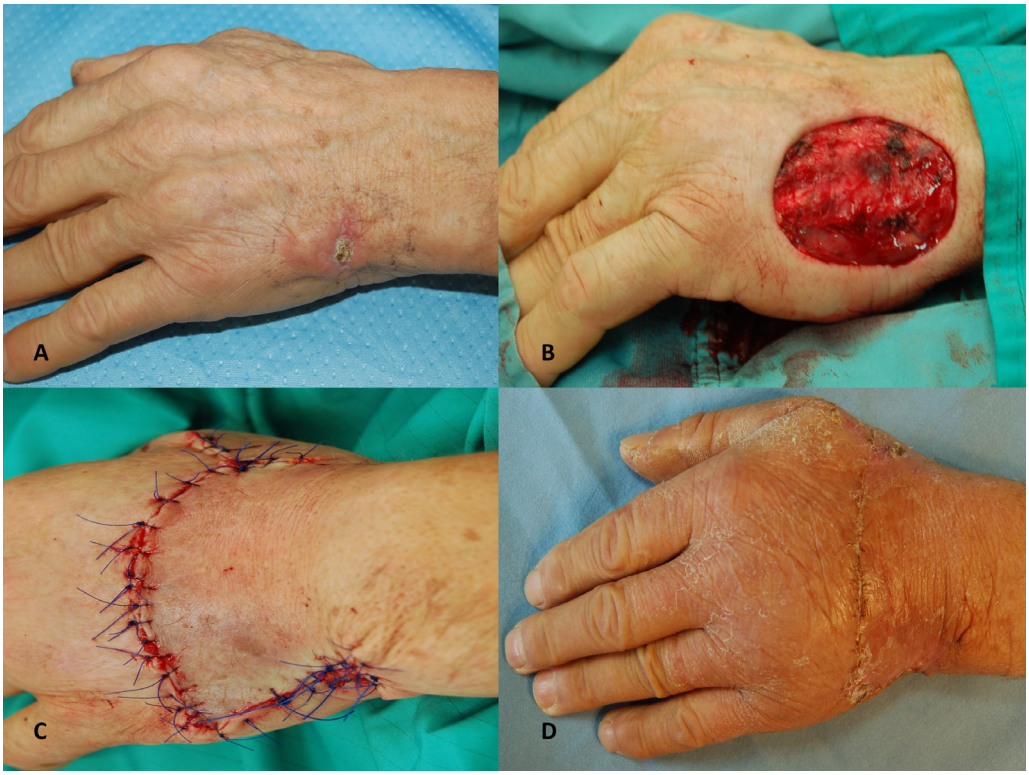

Figure 1. Clinical appearance of the epithelioid sarcoma located on the back of the left hand over the fifth metacarpal (A). Round defect following the excision of the tumor (B). The rotation flap delineated with a semicircular curve and raised while preserving the dorsal nerves and veins and sutured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

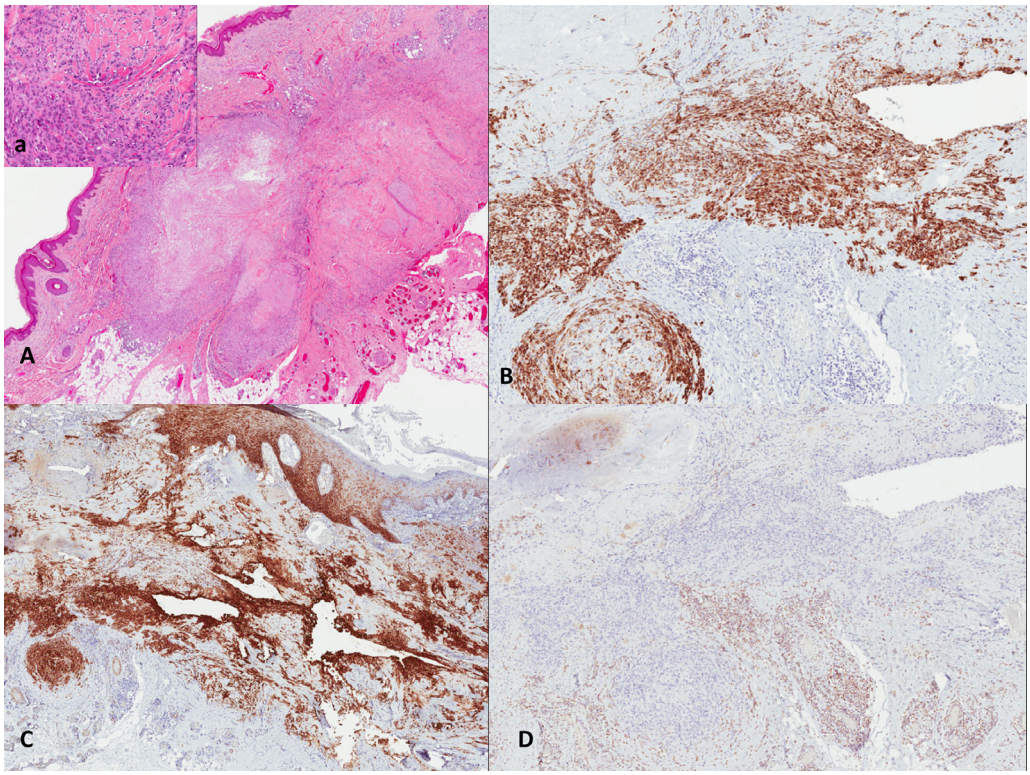

Figure 2. Histopathologic examination revealed polygonal epithelioid cells arranged in nodular aggregates with moderate cytologic atypia and rare mitotic figures, separated by abundant collagen stroma and ulcerated epidermis (A) (H&E, ×20). Inside box (a) showing tumor with large round or polygonal epithelioid cells in a pseudogranulomatous pattern, prominent nucleoli, and abundant eosinophilic cytoplasm (H&E, ×200). The result of the immunohistochemical study: Tumor cells showing immunoreactivity for pan-keratin AE1-AE3 (B), epithelial membrane antigen (EMA) (C), and negative for integrase interactor 1 (INI1) (D) (IHC, ×100).

(in-transit-type metastases). Disease recurrence can occur many years after the first diagnosis (Chase and Enzinger, 1985).

The proximal-type ES is associated with a higher tumor stage, presence of rhabdoid cells, higher tumor grade, and vascular invasion (Asano et al., 2015).

The extent of primary surgery, tumor size and site, TNM status, had the most critical impact on prognosis (Asano et al., 2015).

ES arising in the extremities was found to have a more favorable prognosis than ES of the trunk, head and neck region. Tumor size appeared to be less important in prognosis, instead the presence of deep-seated occurrence, proximal location, local recurrence, mitosis, necrosis, hemorrhage and vascular invasion, lymph node and distant metastases are correlated with unfavorable survival rates. Sentinel lymph node biopsy (SNB) and regional lymphadenectomy aren't useful on the primary state.

ES has been reported to have a high rate of distant metastasis (40–57%) in the lymph nodes, lung, liver, scalp and bones (Asano et al., 2015).

The Overall Survival (OS) rates at 5 year are 32–78% (distal-type 53–100% and proximal-type 33–79.5%) (Asano et al., 2015; Casanova et al., 2006).

Surgical resection is the mainstay treatment of ES, and histologically disease-free margins are the most important prognostic factors for recurrence (Chase and Enzinger, 1985; Casanova et al., 2006).

Because distal sites often are affected, amputation should be considered as an option, especially after the first local disease relapsing, but modern limb salvage techniques combined with adjuvant radiotherapy or chemotherapy are now the standard treatment options for extremity ES and contribute to improved functional outcomes (Miettinen et al., 1999; Hurren and Cormack, 2000).

Adjuvant therapies are recommended according to the high risk of local and distant disease recurrence, as suggested by the presence of residual disease after initial surgery, proximal-type ES, high tumor grade and stagy, presence of vascular and lymph node invasion (Asano et al., 2015).

The case reported herein is a relatively rare presentation of ES in the elder patient. Keeping count of the surface peripheral location, indolent course of the disease, moderate cytologic atypia and low mitotic activity of the tumor, the absence of lymph node and visceral metastases we preferred the surgical strategy alone after a face-to-face confrontation with pathologist, oncologist and radiotherapist. Patient was informed about the diagnosis, prognosis and the potential benefits and harms of treatment options. We are reporting the application of the rotation flap principle to reconstruct a large defect on the dorsum of

the hand (Hurren and Cormack, 2000). The flap offers repair with local skin of similar color, texture and thickness. The end results were excellent both functionally and cosmetically with no recurrence of the tumor at 9 months.

4. Conclusion

Even if the mainstay of therapy is surgery, interdisciplinary approach involving dermatologists, pathologists, radiologists, surgeons, radiation therapists, and medical oncologists is mandatory in cases of ES.

Although the prognosis of ES is very poor, a patient-specific approach to decide therapy planning and performance is necessary in order to ensure the best possible functionality. The close clinical and instrumental follow-up is absolutely necessary for early detection of disease recurrence.

Competing interests

The authors declare that they have no competing interests.

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