Epithelioid finger-sarcoma in an 11 year old girl – a case report

Epitheloides Sarkom am Finger eines 11-jährigen Mädchen – ein Fallbericht

Abstract

Malignant tumors of pediatric hand are very rare. This case report describes an epithelioid sarcoma at the finger of an 11 year old girl and discusses general treatment options in this rare patient population.

Zusammenfassung

Maligne Tumore an Kinderhänden sind sehr selten. Dieser Fallbericht beschreibt ein epitheloides Sarkom am Finger einer 11-jährigen Patientin und diskutiert die Behandlungsoptionen bei diesem seltenen Patientengut.

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Introduction

Malignant tumors of pediatric hand are very rare and therefore difficult to handle. The following case report describes an epitheloid sarcoma at the finger of an 11 year old girl, which was underestimated elsewhere as a ganglia.

Case presentation

An 11 year old girl was admitted to our hospital. Her parents described a tumor formation of approximately 1 cm in terms of a nodule at the end phalanx of the left third finger, which grew within 4 weeks. The preoperative x-ray showed no pathologic findings, therefore the initial clinical diagnoses focused on a ganglion and an ambulant resection was performed by a non-hand surgeon outside the hospital. Unfortunately, postoperative histology revealed the presence of a sarcoma, which was not completely removed. Reference pathology confirmed the rare diagnosis of an epithelioid sarcoma. The girl was then admitted to our department for further therapy. The initial examination showed a bland scar, with no functional deficits (Figure 1a). A new x-ray revealed no further findings and staging of the patient was negative. We performed a further wide resection of the former tumor-bed with temporary wound closure (Figure 1b). Postoperative histology showed no residual tumor cells within the resection, therefore wound closure was achieved by a split-skin graft, which healed primary (Figure 1c). No functional impairments remained. The girl underwent periodical follow-up care in terms of clinical and radiological (MRT) controls, which were guided by a pediatric oncologist. Chemotherapy or radiotherapy has not been applied. Until now, almost 1.5 years later, there is no evidence for recurrence.

Discussion

Epithelioid sarcomas, a subcategory of non-rhabdomyosarcomas, are very rare, high-grade tumors that constitute less than 1% of all soft tissue sarcomas (which represent itself only about 8% of all malignant tumors) [1], [2]. Only few cases of epithelioid sarcomas are described in literature [2], [3], [4]. It shows preference for distal parts of the extremities, particularly the hand and differ from benign lesions (e.g. ganglion) in ways of that are often non apparent until there is recurrence. A frequent property of these tumors is the loss or inactivation of the tumor suppressor gene SMARCB1/INI1 [5], which was unfortunately not investigated in the present case. The indication for nodal biopsy is controversial: epithelioid sarcoma would appear to have a greater tendency than others to spread to regional lymph nodes, and biopsy may be indicated in such cases, though pediatric cases seem to have a weaker propensity for nodal spread than in adults [3].

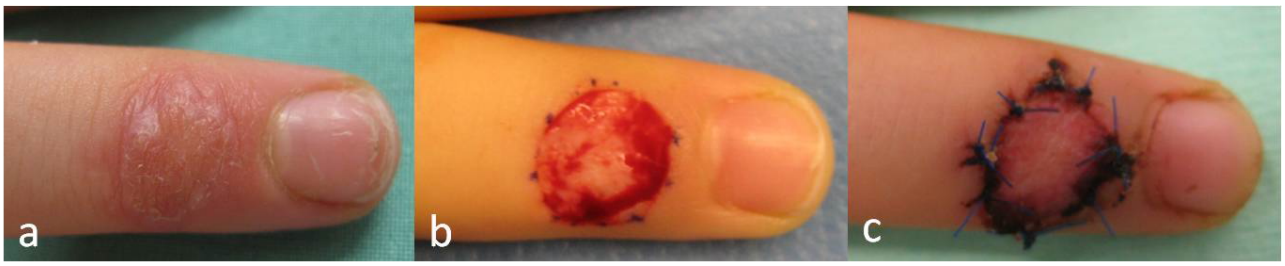

Figure 1: (a) Clinical findings on admittance, (b) surgical excision, (c) wound closure with split skin graft

Poor data records do not allow a reliable prognostic conclusion, but small tumors (<5 cm) without metastasis might have a beneficial prognosis with R0-resection [2]. But even an R0-resection still has a high risk of relapse due to tumor growth along the tendon sheets in up to 85% [2] – therefore close ontological screenings are inalienable. However, surgical resection clearly remains the mainstay of treatment, with surgeons striving for tumor free margins [2], [4]. Since epithelioid sarcomas prefer the dorsal/distal parts of the upper extremity, and fingers in particular, amputation of the affected side should be considered as an option, especially after local recurrence. Due to the small study population, the role of adjuvant therapies remains uncertain and should be discussed individually in an interdisciplinary tumor board [1].

Conclusion

Epithelioid sarcomas are in general very rare malignant tumors, but they show preference for distal parts of the upper extremity. Since they have an indolent and sometimes slow growth rate, they are often confounded with benign lesions. Therefore, every unclear tumor of pediatric hands should be admitted to a specialized center with hand surgical and oncological experience; detailed preoperative diagnosis in terms of x-ray, MRT, ultrasound and possibly biopsy should be performed in advance. Good coordination between hand-surgeon, pediatric oncologist, pathologist, and radiotherapist is mandatory to plan a successful multimodality treatment for these patients.

Notes

Conflicts of interest

None declared.

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