Case report

Limits and chances in an unfortunate course of recurrent orbital rhabdomyosarcoma

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Abstract

Background*:* Orbital rhabdomyosarcoma (RMS) in childhood has an excellent survival rate after chemotherapy and radiation, and mutilating surgery can often be avoided. *Case report:* As a rarity we present an unfortunate disease course in a child suffering from orbital embryonal RMS which did not enduringly respond to multimodal therapy including local excision and exenteration orbitae. After short intervals and despite tumor-free margins, orbital RMS recurred twice and led to an extended exenteration orbitae including the bony margins. Because of the lack of standards for adjuvant therapy in cases of recurrences after exenteration orbitae, therapy had to be restricted to a wait- and- see strategy as the only chance of tumor control.

Conclusion*:* Although survival rates of orbital RMS are high, the possibility of recurrence should not be underestimated. In cases of refractory RMS, new concepts are needed to offer further chances for survival.

Key words: Orbital rhabdomyosarcoma, recurrence, orbital exenteration

Introduction

Rhabdomyosarcoma (RMS) is the most common malignant soft tissue tumour in children and is reported to account for 4 % of all orbital masses. The Intergroup Rhabdomyosarcoma Studies (IRSs) established that the ideal management of this disease is multimodality treatment, using a combination of surgery, chemotherapy, and radiotherapy (Maurer et al 1993). Ten-year event-free survival and overall survival are described as 77 % and 87 % of patients, respectively (Oberlin et al 2001). Against this background, we present an unfortunate disease course in a child suffering from orbital embryonal RMS which did not respond to multimodal therapy. We review our observations and the need for further concepts to offer better chances for survival in such refractory cases.

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Case report

At first presentation, a 7-year-old boy showed a rapid onset of outward proptosis of the globe (Fig. 1). MRI displayed a 15 x 10 x 11 mm tumor in the left orbita (Fig. 2). Biopsy of the tumor offered the histological diagnosis of an embryonal RMS (Fig. 3). Staging showed no evidence of bony infiltration or metastatic disease. The child received chemotherapy consisting of three cycles of vincristine, actinomycin D, and ifosfamide (VAI).

An evaluation with MRI after the third cycle showed no response but tumor progression to a 22 x 10 x 13 mm tumor mass. Excision of the tumor via a transconjunctival approach showed histologically tumor-free margins. Chemotherapy was continued with a fourth cycle of VAI and three cycles of VA. Additionally, the child was admitted to radiotherapy and received hyperfractionated radiation up to a total dose of 41.4 Gy. The control MRI showed complete remission after radiotherapy. Four months later, a rapidly progressive unilateral exophthalmos occurred on the left side again. MRI now showed a tumor mass lateral and inferior in the former tumor bed with a dimension of 24 x 10 x 10 mm. Furthermore, no bony infiltration could be detected. The pseudoencapsulated tumor was resected in total within the exenteration orbitae and a 15 mm^2 bony window from the lateral inferior aspect (Fig. 4). Negative margins with also no evidence of bony infiltration were histologically verified. Chemotherapy was carried on with enteral application of etoposide and idarubicin. After five months of follow-up, a clinical suspected nodule of 5 x 5 mm was again found at the infraorbital bony margin. The excision of the tumor with the underlying bone revealed the second recurrence of RMS with infiltration of the bony margins. In a last step, the infraorbital rim and two-thirds of the orbital floor were resected to guarantee tumor-free margins (Fig. 5). Histological evaluation confirmed complete resection. MRI total body scan showed no signs of metastatic disease. After 18 months of follow-up, no further recurrence of the disease was detected.

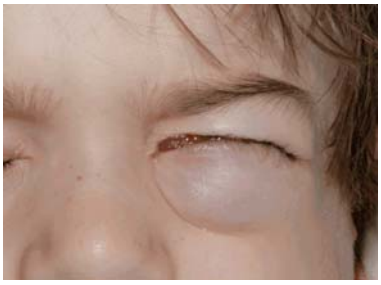

Fig. 1
A 7-year-old boy with rapidly progressive unilateral exophthalmos with conjunctivitis and chemosis on the left side at first presentation

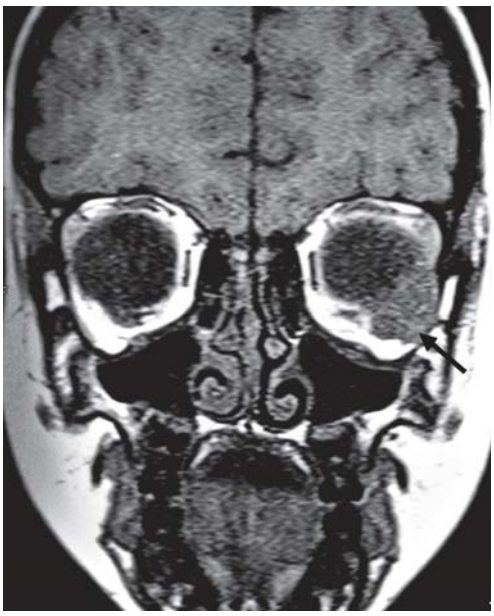

Fig. 2
T1-weighted coronal MRI depicting the inferiorly and laterally located RMS. The tumor signal is hypointense to intraocular muscles (arrow)

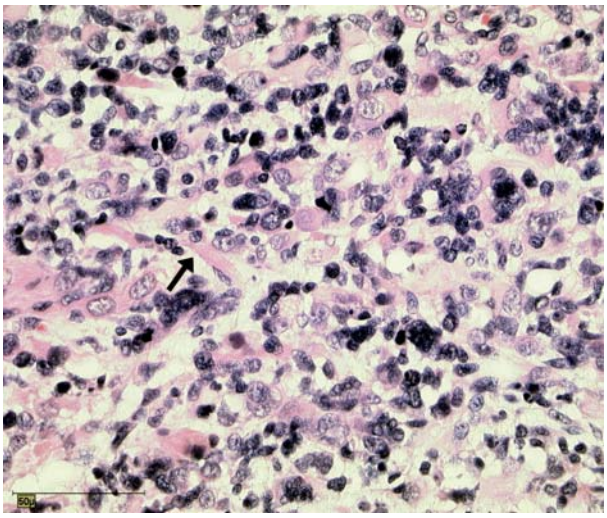

Fig. 3
Embryonal RMS composed of spindle-shaped cells (rhabdomyoblasts) characterized by bright eosinophilic cytoplasm. In addition, eosinophilic skeletal muscle residuals (arrow) can also be detected in the stroma (hematoxylin and eosin staining, magnification 40x).

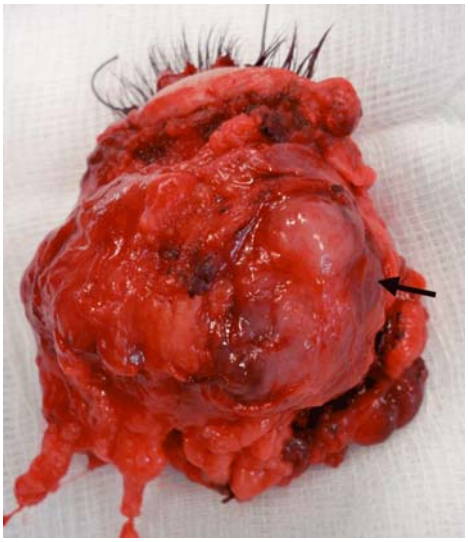

Fig. 4
Inferior posterior view of specimen of exenteration orbitae following the first recurrence of RMS with pseudoencapsulated tumor (arrow)

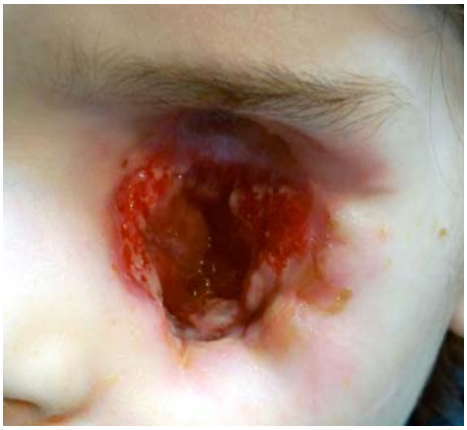

Fig. 5
Clinical situation with expanded resection defect after sacrificing the infraorbital bony margin following the second recurrence of RMS

Discussion

The remarkable unfortunate sequelae in the presented case were that the primary tumor did not respond to chemotherapy and was recurrent despite histological tumor-free margins and radiation. It was also remarkable that the second recurrence of RMS involved bone, despite bony infiltration having been ruled out in the first recurrence. Additionally, exenteration orbitae including resection of the first recurrence within the intact pseudo-capsule of the RMS revealed histologically tumor-free margins but did not prevent recurrence. This uncommon course obviously suggests that a type of embryonal RMS exists that seems to be refractory against chemotherapy and radiation and obviously spreads RMS cell clusters beyond common surgical resection margins. However, there are still no references in literature available which could enlighten this hypothesis. Therefore, new studies are warranted, which could contribute more to the molecular genetical aspects of the possibility of therapeutic refractory RMS. However, we learned that despite exenteration orbitae, recurrence of RMS is possible and that obviously, in some cases, it is necessary to sacrifice the complete bony margin of the orbit next to the RMS.

Another problem we had to face was that after exenteration orbitae no standardized further adjuvant therapy could be administered due to the lack of standard protocols for such cases. The application of etoposide and idarubicin showed no effect for the prevention of a possible recurrence. As a result, with the second recurrence of RMS, we were forced to fall back to the surgical- and wait-and-see strategies for tumor control, which were the standards of care for RMS during the first half of the 20th century (Jones et al 1965). However, the exenteration procedure for such cases of refractory RMS was found to be of value in prolonging survival with a 5-year survival rate of 71 % (Mannor et al 1997). But, on the other hand, for patients with recurrence of RMS after exenteration orbitae , a 5-year survival is hard to reach and further standard adjuvant concepts are needed beyond histologically controlled surgery and wait-and-see strategies.

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