

Tran's Arterial Embolization of an Unusual Pleomorphic Rhabdomyosarcoma in Adulthood for Better Preoperative Conditioning

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ABSTRACT

Rhabdomyosarcomas, being the most common sarcomas of childhood, have an extremely poor prognosis, and pleomorphic rhabdomyosarcomas occur very rarely in adults. Despite advanced diagnostics, it is not uncommon for such tumours to be misinterpreted.

The 73-year-old patient was diagnosed with a swelling on the inner side of the left thigh 3 years previously after a minor trauma, which was interpreted as a small well-revascularised haemangioma in original CT angiography. While this mass showed little change over the last 2.5 years, a sudden progression of growth occurred 6 months ago. With the working diagnosis of a haemangioma, differential diagnosis of a schwannoma, the vessels supplying the tumour from profunda femoral artery were visualised preoperatively in a selective angiography and embolised. Shortly before the operation, duplex sonography revealed a renewed hypervascularisation of the tumour. The supplied vessels from the superficial femoral artery and from the main left profunda trunc were embolised with Embosphere Microspheres 500-700 (Merit). Complete devascularisation of the tumour was achieved, so that the tumour could be resected very effectively in accordance with the compartment. The histological work-up of the tumour resulted in the diagnosis of a pleomorphic rhabdomyosarcoma grade II with tumor-free margins.

Preoperative embolisation could be an effective method for preoperative conditioning of such rhabdomyosarcoma. Using this example, this study discusses the current diagnostic and therapeutic options for adult rhabdomyosarcomas.

Keywords

Embolization, Rhabdomyosarcoma, Adulthood.

Introduction

Soft tissue sarcomas are malignant tumors of diverse histologic origin with varying malignancy [1,2]. The mechanisms of origin are not well understood [3-6]. 40% of all sarcomas are characterized by a specific translocation or mutation (7), 60% of soft tissue sarcomas show no characteristic translocations [7,8], of these 20% represent undifferentiated sarcomas [7,8]. Immunohistochemical and molecular techniques for correct diagnosis are essential [9]. Genetically unstable forms include

leiomyosarcomas, pleomorphic rhabdomyosarcoma, liposarcoma, the undifferentiated pleomorphic sarcomas and, specifically, malignant fibrous histiocytoma (MFH) [8]. Despite tremendous advances in diagnostics, histologic typing of soft tissue tumors is, as a whole, often difficult [1,6,7,10].

Rhabdomyosarcoma, a sarcoma with differentiation originating from skeletal muscle cells occurring preferentially in childhood and adolescence, is a highly malignant soft tissue tumor [11].

RMS (ERMS) and alveolar RMS (ARMS) rhabdomyosarcomas, being the most common sarcomas of childhood, have an extremely

poor prognosis, and pleomorphic rhabdomyosarcomas occur very rarely in adults [1,12-14].

In the present case, the occurrence, diagnosis, and treatment of pleomorphic rhabdomyosarcoma in a 76-year-old female patient in the form of a soft tissue tumor with aspects of arteriobenous malformation are presented.

Case Report

The tumour, which had been detected in the patient already 3 years earlier, showed a very slow size progression with a blood supply from the profunda femoral artery. The CT angiography showed a 2.5 cm large mass with strong contrast in the marginal area and irregular internal structures in the medial area of the thigh musculature classified it as a haemangioma (Figure 1). In the CT angiography of the left leg performed one year later, even a slight increase in size of the intramuscular mass to approx. 34.6 mm x 41.9 mm x 26.7 mm was found, which was now supplied by both feeding branches from the superficial femoral artery and profunda femoral artery (Figure 2). With very early contrast in the arterial phase and a clear homogeneous increase in the KM accumulation in the late phase, this was interpreted as an intramuscular haemangioma. In a recent MRI, the now 47mm x 46mm x 41mm mass was smoothly circumscribed and surrounded by the hypointense capsule. It showed predominantly moderate hyperintensity with single clear hyperintense areas on the T2 image, hypointense on the T1 image, and relatively strong and homogeneous KM enhancements after KM administration. No evidence of cortical infiltration or compartmental overshoot was noted. Thus, this mass was interpreted as a schwannoma with cystic degeneration and necrosis (Figure 3).

In an interdisciplinary conference, it was decided to perform intra-arterial embolisation if the tumour was very vascularised.

The selective DSA showed an arterial supply to the medial part of the tumour (2/3 of the tumour volume), mainly via a fine proximal and distal branch from the superficial femoral artery, and a further arterial supply to the tumour (approx. 1/3 of the tumour volume), mainly to the lateral proximal part via a fine branch from the main left profunda trunk.

After super selective probing of a more distal branch from the left superficial femoral artery, a micro catheter was advanced via a 0.014' wire to the lateral tumour portion. Due to early venous filling indicating fistula, embolisation was performed with Gelfoam and venous outflow was compressed with a blood pressure cuff during the intervention. Subsequent angiographic control showed complete occlusion of the feeding tumour artery in the lateral tumour portion. Subsequently, super selective probing was performed, first of the more proximal tumour artery on the left and later of the more distal one. Both tumour embolisations were performed with embospheres (500-700 microm) until stasis was achieved. The final angiographic control showed extensive tumour devascularisation. (Figures 4 a,b,c).

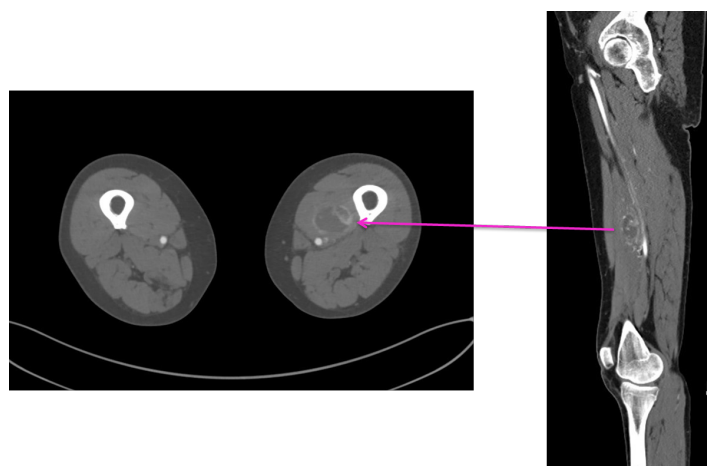


Figure 1: The CT angiography in 3 mm slides (10/2018): a 2.5 cm large mass with strong contrast in the marginal area and irregular internal structures in the medial area of the thigh musculature classified it as a haemangioma.

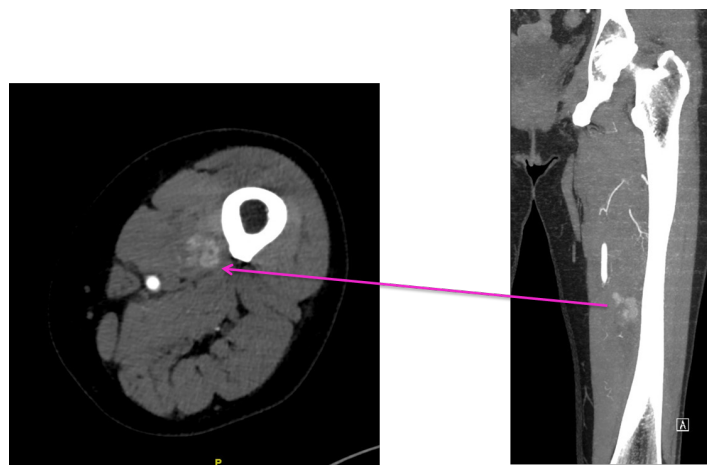


Figure 2: CT angiography in 3 mm slides (7/2019): intramuscular mass to approx. 34.6 mm x 41.9 mm x 26.7 mm supplied by both feeding branches from the superficial femoral artery and profunda femoral artery.

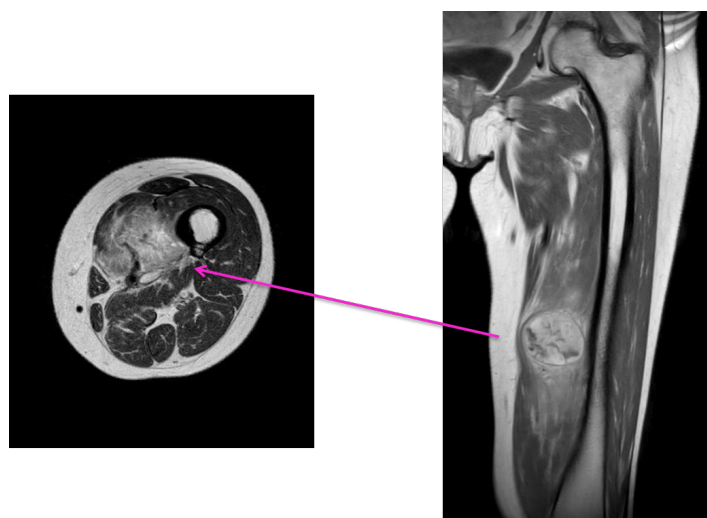


Figure 3: MRI with contrast agent (11/2020): a 47mm x 46mm x 41mm mass smoothly circumscribed and surrounded by the hypointense capsule with very early contrast in the arterial phase and a clear homogeneous increase in the contrast agent accumulation in the late phase.

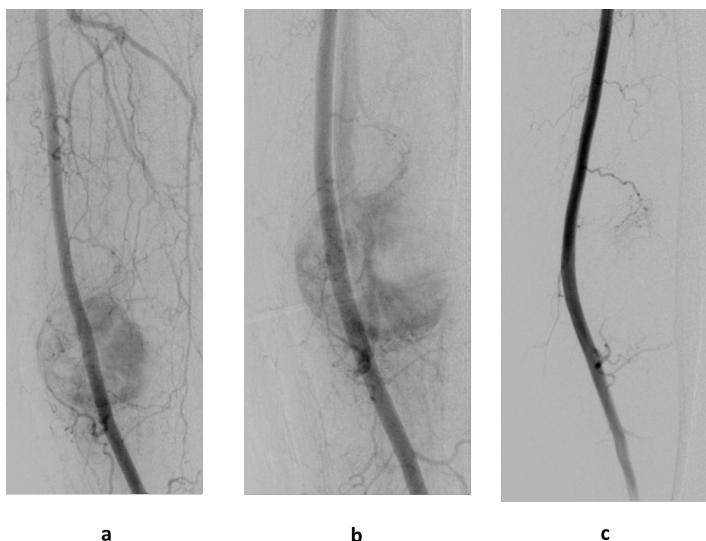


Figure 4 (a-c): The selective angiography (12/2020): an arterial supply to the medial part of the tumour (2/3 of the tumour volume) via a fine proximal and distal branch from the superficial femoral artery, and a further arterial supply to the tumour (approx. 1/3 of the tumour volume) mainly to the lateral proximal part via a fine branch from the main left profunda trunk (4a).

After superselective probing of a more distal branch very early venous filling of the superficial femoral vein (4b).

Complete devascularization of the tumour through embolisation of fine proximal and distal branch from the superficial femoral artery with Gelfoam and embolisation of more proximal and distal branches from main left profunda trunk (4c).

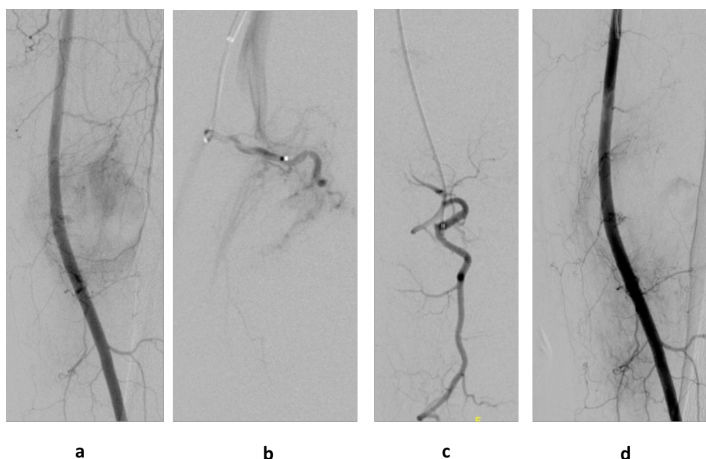


Figure 5 (a-d): The selective angiography (2/2021): Selective angiography via the superficial femoral artery with visualization of the tumour-supplying vessels from the superficial and profunda femoral arteries (5a).

Superselective probing of the proximal feeding tumour artery with and embolisation with Embosphere Microspheres 500- 700 (Merit) (5b).

Superselective probing of the distal feeding tumour artery with and embolization with Embosphere Microspheres 500- 700 (Merit) (5c).

The subsequent control with extensive tumour devascularization (5d).

However, the preoperative duplex control showed a pronounced

residual perfusion, especially of the lateral part, so that embolisation was performed again.

Selective DSA via the femoral superficial artery also confirmed significant CM accumulation of the tumour. After super selective probing of the more proximal feeding tumour artery with a 0.014' wire and a 2.4F micro catheter (Merit Maestro micro catheter), embolisation was then performed with Embosphere Microspheres 500- 700 (Merit). The subsequent control angiography showed extensive tumour devascularisation (Figures 5 a,b,c,d).

Subsequently, the tumour was resected in a compartmentalized manner. Intraoperatively, the tumour including the tumour capsule could very easily be dissected and resected from the musculature and nerve-vascular sheath. The histological work-up of the tumour resulted in the diagnosis of a pleomorphic rhabdomyosarcoma grade II. All resection margins were tumor-free.

Discussion

The most common adult sarcomas are liposarcomas, leiomyosarcomas and undifferentiated sarcomas. Osteosarcomas, Ewing sarcomas and chondrosarcomas are the most common sarcomas originating from bone tissue [15-19]. The extremities are the most frequently affected with 43%, followed by the viscera (19%), retroperitoneum (15%), trunk (10%) and head and neck (9%) [20]. With an incidence of <0.2% of all malignant tumours, soft tissue sarcomas are therefore a rare tumourity [21].

The importance of early diagnosis for the success of therapy has been emphasised in several studies [20-25]. MRI is recommended as the gold standard method [24]. In contrast, functional imaging is used more to assess the success of therapy [25]. Scintigraphy with 201Thallium (201TI) and positron emission tomography (PET) with 18F-FDG and 11C-methionine were able to contribute to the detection of a recurrence with a specificity of 97% [26-29]. Thus, it was even possible to derive a direct correlation between the decrease in 18F-FDG uptake and the overall prognosis as well as the risk of recurrence on the one hand, and with the tumour grade and proliferation rate on the other [30,31].

As our case shows, both CT and MRI can be misinterpreted, especially in well-vascularized rhabdomyosarcomas. In view of our working diagnosis of a well-vascularized schwannoma, we dispensed with a trial biopsy. These are recommended for histological confirmation in the form of a multiple core-needle biopsy in the case of deep, or an excision biopsy in the case of superficial tumours, which has prognostic significance [32,33].

The 5-year survival rate depending on the UICC/AJCC stage is approx. 85-96% in stage I, 72-78% in stage II, 50% in stage III and approx. 10% in stage IV [34].

Tumour grading, size and histological subtype are the most important prognostic factors for disease-specific survival (DSS) in patients with trunk and limb STS. Some studies have even found

better long-term outcomes with deep tumour location, positive margins and lower limb localization [19,35-37]. While the risk of local recurrence is not a major problem even in high-grade tumours, which account for almost 75 % of trunk and limb sarcomas, with compartment resection or amputation, the prognosis of these patients is mainly determined by the distant metastasis [38].

The treatment of sarcomas poses a special challenge at a multidisciplinary therapy centre. The therapy options consist of a combination of resection, chemotherapy and radiotherapy. The therapy approaches differ depending on the individual risk assessment based on histology, degree of malignancy, localization of the tumour and age of the patient. The use of new drugs and new radiotherapy techniques, such as particle therapy, aim not only to improve tumour control but also to reduce the therapy-related late effects.

Treatment options for WTS include various combinations of extensive resection, pre- or postoperative radiation and adjuvant or neoadjuvant chemotherapy. All other therapy options such as regional hyperthermia or isolated hyperthermic limb perfusion with tumour necrosis factor- and melphalan have more of an experimental character [39].

Regardless of histological type, surgical resection with negative microscopic margins (R0 resection) is the goal of curative sarcoma therapy, which should be limb-sparing, function-preserving with tumour-free margins [40,41]. The biological behaviour of the tumour usually determines the short-term outcome, and the quality of surgical resection determines late survival [42-45].

The concept of limb preservation in soft tissue sarcomas of the extremities is underlined by results of several studies in which almost equivalent survival rates were achieved after limb-preserving surgery with radiation [46-48].

Preoperative radiotherapy leads to a better surgical outcome and thus to better long-term functional results and thus a better quality of life, despite increased postoperative wound healing disturbances [49-55]. However, measurable effects on quality of life were not observed even with moderate side effects of radiation such as loss of strength, oedema and limb range of motion [56]. Postoperative radiotherapy also significantly reduced the rate of local recurrence after 10 years in patients with high-grade tumors [39]. However, this effect could not be demonstrated in low-grade tumors, so that postoperative radiation therapy of this tumor type does not improve the prognosis [57].

Against this background, National Comprehensive Cancer Network (NCCN) recommends either pre- or postoperative radiation only for soft tissue sarcomas of the extremities in stages II, IIIA and IIIB. For stage IA and IB soft tissue sarcomas with proven tumour-free resection margins, surgery alone is usually sufficient [58]. While neoadjuvant therapy with or without radiation is recommended for better respectability, especially in advanced, non-respectable or metastatic WTS soft tissue sarcomas, the use of adjuvant therapy in high-risk situations is controversial [59-61]. Accordingly, neoadjuvant chemotherapy is recommended for

patients with high-grade tumours of >10 cm [62,63].

A new feature of this concept is the pre-operative embolisation of the feeding vessels, which not only led to a reduction in the size of the tumor. Rather, it made the compartment resection of the tumour much easier. Such procedures have been used sporadically as preoperative conditioning of well-vascularized soft tissue tumours, but are not considered evidence-based due to their small-group nature [64,65].

There is extensive experience in the treatment of AV fistulas. For embolization of multiple AV fistulas, transvenous embolization with a precipitating mixture consisting of ethylene-vinyl alcohol copolymer and the organic solvent dimethylsulphoxide (DMSO) with tantalum powder admixtures for X-ray visualization is usually recommended to reduce the risk of embolism [66,67].

Conclusions

It might be worth considering using this method for preoperative conditioning of well-vascularized sarcomas of the extremities due to the very poor prognosis of soft tissue sarcomas. Further prospective studies will be necessary to evaluate this procedure.

Ethics approval and consent to participate

This article does not contain any studies with human participants performed by any of the authors. This case report is a general therapy without an experimental approach.

Consent for publication

The patient's declaration of consent for the publication of her anonymised images (CT angiography, MRI angiography) is available as a copy.

Availability of data and material

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

Authors contributions

Sheila Fatehpur and Masoud Mirzaie described the course of the disease. Johann Philipp Addicks and Van Khiem Tran described the interventional parts. Raphaela Verheggen contributed to the intraoperative and postoperative course.

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Apart from the authors mentioned above, there are no other persons who have contributed to this article.

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