A patient with scapular Ewing sarcoma; 5-year follow-up after extracorporeal irradiation and re-implantation of the scapula, a case report

Daniel Hoornenborg a,*, Ewout S. Veltman a, Foppe Oldenburger b, Jos A.M. Bramer a, Gerard R. Schaap a

a Department of Orthopaedic Surgery, Academic Medical Center Amsterdam, Meibergdreef 9, 1100 DD Amsterdam, the Netherlands
b Department of Radiotherapy, Academic Medical Center Amsterdam, Meibergdreef 9, 1100 DD Amsterdam, the Netherlands

1. Case

In September 2007 a 9½-year-old boy was diagnosed with a Ewing sarcoma (EWS-FL1 translocation positive) of the right scapula. MRI showed a tumor of 8.0 × 2.9 × 6.2 cm located in the right scapula with dorsal displacement of the infraspinatus and teres minor muscles (Fig. 1). The tumor extended in to the glenoid, but the shoulder joint was not contaminated. Staging, including bone scan (Fig. 2) and CT-imaging, did not show metastatic lesions. The child was treated with chemotherapy according to the EURO-EWING 99 protocol [1]. After 6 VIDE courses soft tissue involvement was absent on MRI. Intraoperative extra-corporal irradiation (IEI) was chosen since functional results with scapula prostheses are poor [2]. After the resection of the scapula pathologic samples were taken from the specimen: vital tumor cells were seen on the surface of the dorsal part of the periost. The resected scapula was irradiated with a tumoricidal dose of 120 Gy by means of 6 MV photons (Fig. 3), creating dead autologue bone graft of the correct dimensions for re-implantation and reconstruction [3]. During irradiation the scapula was kept in a plastic bag filled with sterile fluid. After resection the scapula was re-implanted into the body. Tendons and muscles were re-inserted. The boy was treated with postoperative chemotherapy according to the earlier mentioned protocol.

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During follow-up regular MRI and radiographic imaging was performed to check for local or metastatic recurrence of the Ewing sarcoma.
Postoperative examination of shoulder function gives ante-flexion of over 160°, abduction of over 160° (Fig. 4), exorotation to 60°, endorotation to the left scapula as shown in Fig. 4. Sensibility and strength are intact.

At five year follow-up there has been subtotal resorption of the scapula (Fig. 5), leaving the glenohumeral joint intact. Remaining shoulder function gives anteflexion of over 120°, abduction of 100°, exorotation to 60°, endorotation to the left scapula as shown in Fig. 6. Sensibility and strength are intact.

2. Discussion

Malignant tumor of the scapula is a rare condition. Several surgical treatment types are available, such as total scapulectomy, constrained prosthesis and scapular allograft. All of these surgical treatment options result in very little or no shoulder function [2,4–7].

Resection, extracorporeal irradiation and re-implantation (IEI) is described in literature as a treatment option for bony malignancy [3,8–15]. It has been used in incidental cases, both in Ewing Sarcoma and other bone tumors [3,9,11–15]. As far as we know IEI had never been described in a child with a scapular Ewing sarcoma.

Types of cancer affecting the scapula described in literature are chondrosarcoma, synovial sarcoma, Ewing sarcoma (ES) and metastasis [4–7,16,17]. Modern treatment of ES of bone in children and adults consists of chemotherapy and local treatment by surgery or radiotherapy or a combination of both. If possible,
surgery with appropriate margins alone is the preferred local treatment.

Resection, IIEI and re-implantation of the scapula combined with chemotherapy has proven to be a successful treatment option in this young patient with malignancy of the scapula and without metastatic disease. This case shows that shoulder function can remain largely intact at 5 year follow-up because of preserved glenohumeral joint function.

Contributions of authors

D. Hoornenborg and E. S. Veltman were responsible for drafting the manuscript and reviewing the literature. F. Oldenburger was responsible for the radiotherapy. G. R. Schaap treated the patient as orthopaedic surgeon. G. R. Schaap and J. A. M. Bramer were responsible for the final review of the manuscript.

References

(1) Ladenstein R, Potschger U, Le Deley MC, Whelan J, Paulussen M, Oberlin O, et al. Primary disseminated multifocal Ewing sarcoma: results of the Euro-EWING 99 trial. Journal of Clinical Oncology 2010;28(20):3284–91.
(2) Tang X, Guo W, Yang R, Ji T, Sun X. Reconstruction with constrained prosthesis after total scapulectomy. Journal of Shoulder and Elbow Surgery 2011;20(7):1163–9.
(3) Davidson AW, McCarthy SW, Stalley PD. En-bloc resection, extra-corporeal irradiation, and re-implantation in limb salvage for bony malignancies. Journal of Bone & Joint Surgery (British Volume) 2005;87(6):851–7.
(4) Griffin AM, Shaheen M, Bell RS, Wunder JS, Ferguson PC. Oncologic and functional outcome of scapular chondrosarcoma. Annals of Surgical Oncology 2008;15(8):2250–6.
(5) Hayashi K, Karita M, Yamamoto N, Shira T, Nishida H, Takeuchi A, et al. Functional outcomes after total scapulectomy for malignant bone or soft tissue tumors in the shoulder girdle. International Journal of Clinical Oncology 2011;16(3):568–73.
(6) Mayil VN, Mohanlal P, Bose JC, Gangadharan R, Karthisundar V. The functional and oncological results after scapulectomy for scapular tumours: 2–16 year results. International Orthopaedics 2007;31(6):831–6.
(7) Nakamura S, Kusuzaki K, Murata H, Takeshita H, Hirata M, Hashiguchi S, et al. Clinical outcome of total scapulectomy in 10 patients with primary malignant bone and soft-tissue tumors. Journal of Surgical Oncology 1999;72(3):130–5.
(8) Arpornchayanan O, Leerapun T, Pruksakorn D, Panichkul P. Result of extra-corporeal irradiation and re-implantation for malignant bone tumors: a review of 30 patients. Asia-Pacific Journal of Clinical Oncology 2012.
(9) Kim JD, Lee GW, Chung SH. A reconstruction with extra-corporeal irradiated autograft in osteosarcoma around the knee. Journal of Surgical Oncology 2011;104(2):187–91.
(10) Poffyn B, Sys G, Mulliez A, Van MG, Van HL, Forsyth R, et al. Extra-corporeal irradiated autografts for the treatment of bone tumours: tips and tricks. International Orthopaedics 2011;35(6):889–95.
(11) Puri A, Gulia A, Jambhekar N, Laskar S. The outcome of the treatment of diaphyseal primary bone sarcoma by resection, irradiation and re-implantation of the host bone: extra-corporeal irradiation as an option for reconstruction in diaphyseal bone sarcomas. Journal of Bone & Joint Surgery (British Volume) 2012;94(7):982–8.
(12) Anacak Y, Sabah D, Demirci S, Kamer S. Intraoperative extra-corporeal irradiation and re-implantation of involved bone for the treatment of musculoskeletal tumors. Journal of Experimental & Clinical Cancer Research 2007;26(4):571–4.
(13) Krieg AH, Davidson AW, Stalley PD. Intercalary femoral reconstruction with extra-corporeal irradiated autogenous bone graft in limb-salvage surgery. The Journal of Bone & Joint Surgery (British Volume) 2007;89(3):366–71.
(14) Krieg AH, Mani M, Speth BM, Stalley PD. Extra-corporeal irradiation for pelvic reconstruction in Ewing’s sarcoma. The Journal of Bone & Joint Surgery (British Volume) 2009;91(3):395–400.
(15) Takahashi S, Okuda S, Sasai K, Kotoura Y. An bloc resection, extra-corporeal irradiation, and re-implantation of an entire tibia. Journal of Orthopaedic Science 2006;11(3):298–302.
(16) Uematsu A, Brower TD, Winter Jr. WG. Scapulectomy for the treatment of malignant tumors of the scapula. Southern Medical Journal 1979;72(1) 4–7.
(17) Zhang K, Duan H, Xiang Z, Tu C. Surgical technique and clinical results for scapular allograft reconstruction following resection of scapular tumors. Journal of Experimental & Clinical Cancer Research 2009;28:45.