Scientific Research Publishing

Open Journal of Orthopedics, 2022, 12, 335-344
https://www.scirp.org/journal/ojo
ISSN Online: 2164-3016
ISSN Print: 2164-3008

Osteofibrous Dysplasia-Like Adamantinoma of the Tibia Diagnosed at Age 6 Years Progressing into a Classic Adamantinoma over 39 Years, Reconstruction Performed by a Modified Capanna Technique: A Case Report

G. Ulrich Exner[1*], Pascal A. Schai[2], Tobias C. Bühler[3], Michael O. Kurrer[4], Natasha Forster[5]

[1]Orthopaedie Zentrum Zurich, Seestrasse, Zurich, Switzerland
[2]Luzerner Kantonsspital Wolhusen Spitalstrasse, Wolhusen, Switzerland
[3]Kantonspital Baden Im Ergel, Baden, Switzerland
[4]Pathologikum Zurich, Switzerland, Zurich, Switzerland
[5]Swissparc AG, Zurich, Switzerland
Email: *guexner@gmail.com, pascal.schai@luks.ch, tobias.buehler@ksb.ch, michael.kurrer@hin.ch, natasha.forster@swissparc.ch

How to cite this paper: Exner, G.U., Schai, P.A., Bühler, T.C., Kurrer, M.O. and Forster, N. (2022) Osteofibrous Dysplasia-Like Adamantinoma of the Tibia Diagnosed at Age 6 Years Progressing into a Classic Adamantinoma over 39 Years, Reconstruction Performed by a Modified Capanna Technique: A Case Report. *Open Journal of Orthopedics*, **12**, 335-344.
https://doi.org/10.4236/ojo.2022.127032

Received: June 1, 2022
Accepted: July 23, 2022
Published: July 26, 2022

Abstract

Background: It is generally accepted that there is a spectrum ranging from Osteofibrous Dysplasia (OFD) to Osteofibrous dysplasia-like Adamantinoma (OFD-AD) to "classic" Adamantinoma (AD). However, it is controversial whether OFD may progress into OFD-AD and AD or these pathologic changes reflect sampling issues. We herein report the fifth case of late development of AD from OFD-AD to underline the importance of long-term follow-up. **Case:** At 6 years bone biopsy of the tibia revealed the diagnosis of OFD-AD, confirmed by repeat biopsies until the age of 18 years. At age 45 years a progressive focal osteolysis was biopsied and showed histologically a classic AD. The affected tibial diaphysis was resected and pasteurized. Reconstruction was performed adapting the "Capanna technique" of inserting the ipsilateral mobilized fibula into the replanted autologous, pasteurized tibial segment as biologic stabilizer. At 1.5 years follow-up, the reconstruction has healed allowing full weight bearing.

Keywords

Osteofibrous Dysplasia-Like Adamantinoma, Tibia, Classic Adamantinoma, Capanna Technique, Pasteurized Autograft

1. Introduction

Osteofibrous dysplasia (OFD), also known as Kempson-Campanacci lesion, is a rare benign fibro-osseous lesion predominantly affecting the tibia [1]. OFD-like Adamantinoma (OFD-AD) differs from OFD by the inclusion of small clusters of epithelial cells spread throughout the lesion as opposed to single cells, whereas the classic Adamantinoma (AD) is a malignant biphasic tumor with nests or sheets of epithelial cells that are surrounded by spindle cells with an osteofibrous component. Furthermore OFD-AD shows keratin-expressing epithelial cells which are present as small scattered groups and are visible with hematoxylin and eosin staining; however they may be so sparse that they are overlooked unless they are rendered visible with immunohistochemical staining for keratin. Classic AD and OFD-AD share similarities in location and appearance, representing a continuum making their distinction even more complex [2]. There is some controversy distinguishing OFD-AD or "differentiated adamantinoma" from the classic form with regard to the amount of epithelial component that is compatible with the differentiated form and whether progression from differentiated to classic adamantinoma can occur [3].

Accordingly, there is no consensus regarding the best treatment for OFD. Campanacci and Laus [1] recommended delaying surgery as long as possible because of frequent spontaneous regression during childhood and intervention may even activate progression. Spontaneous regression was surprisingly even seen in a 38-year-old man with a rare variant of OFD with rhabdoid elements [4].

Based on the analysis of 16 patients, Lee *et al.* [5] recommended radical extraperiosteal excision in all cases of OFD. However, in a more recent study the same group stated "that it is safe to treat these patients in a conservative manner with annual surveillance" [6].

In a large single-institution study [7] no patient with OFD (n = 42 f/u 38 to 300 months, average 118 months), or OFD-AD (n = 10 f/u 36 to 316 months, average 97 months) progressed to AD. Resection with clear margins was deemed necessary only for patients with classic AD. In a large multicenter study [8] OFD-AD (n = 117) showed aggressive behavior without metastatic potential; only one (histology proven) of the OFD-AD patients developed classic AD (histology proven) within the OFD-AD over a period of 29 years (no f/u reported). In this study AD (n = 141) showed full malignant potential with local recurrence (32%), metastases (18%) and fatal outcome (11%).

Three more patients have been described with transition from OFD-AD to AD, two by Hazelbag [9], one by Hatori [10].

We describe an additional case of OFD-AD of the tibia documented by repeated biopsies at ages 6, 8, 18, and 20 years, which developed into a classic AD within the OFD-AD at the age between 41 to 45 years. The case is of interest as it underlines the importance of "open-end" follow-up (f/u) for patients with the OFD-AD, and highlights the surgical techniques applied.

2. Case Report

The female patient was born in 1975. The first biopsy of the tibia in 1981 revealed an OFD with elements suggesting OFD-AD. Repeat biopsies in 1983, 1984 and 1993 again showed OFD-AD and wide resection was recommended. At this time symptoms were limited to leg length discrepancy, requiring a shoelift on the opposite to compensate the 4 cm difference. Wedge osteotomy for correction of the procurvatum and shortening through the proximal part of the lesion (Figure 1) in 1995 healed uneventfully; the resected wedge was diagnosed again as OFD-AD. Postoperative imaging (MRI, X-Ray) remained unchanged until 2016 (Figure 2), the signal alterations even appeared to get smaller between 2005 and 2016. MRI in 2020 showed an enlarging osteolytic lesion within the preexisting diaphyseal changes with cortical erosion. FDG-PET-CT in 2020 (Figure 3) showed 5 metabolically active foci, the largest with a diameter of 17 mm, however without evidence of metastases; unfortunately earlier PET-CTs were not performed. CT-guided biopsy of the most active lesion was diagnosed again as OFD-AD and curetted. The curetted material however was found to contain a typical AD without dedifferentiation (Figure 4).

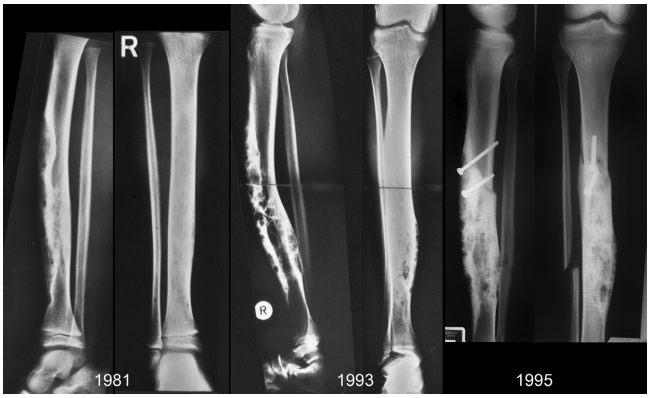

Figure 1. X-rays between 1981 (age 6 years) and 1993 show the increasing deformity of the tibia. X-rays in 1995 show the correction 6 weeks after wedge osteotomy.

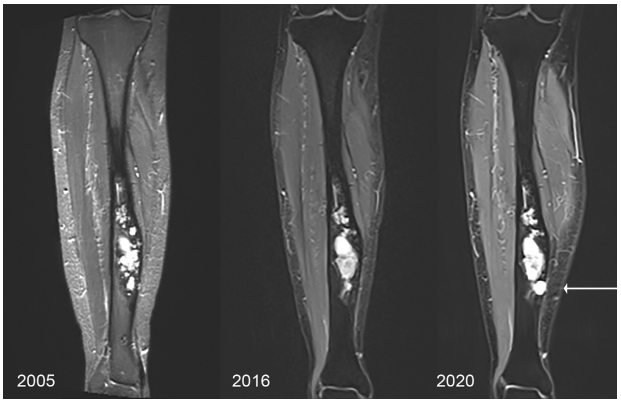

Figure 2. MRI T1 sequences show no significant change between 2005 and 2016. In 2020 in the distal region of the main lesion shows focal enlargement (arrow) and erosion of the cortex, which pathologically was diagnosed as AD in the curettage.

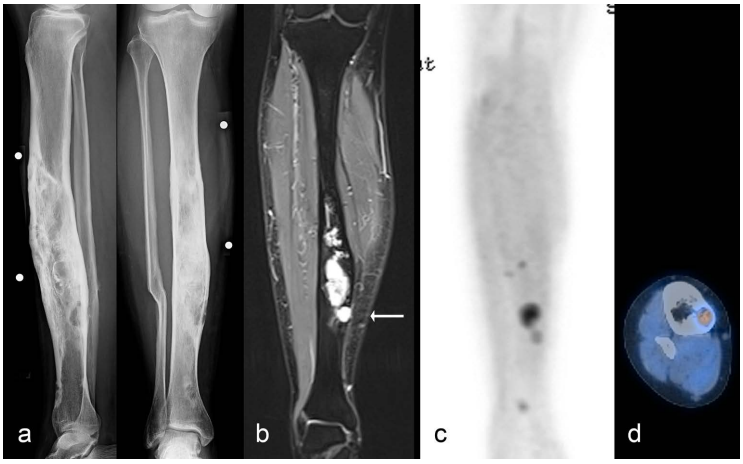

Figure 3. Synopsis of X-Ray (a), MRI (b) and FDG-PET-CT ((c) and (d)) in 2020 shows the metabolically active osteolysis (arrow) distal to the main changes, which was curetted and pathologically demonstrated to be Adamantinoma.

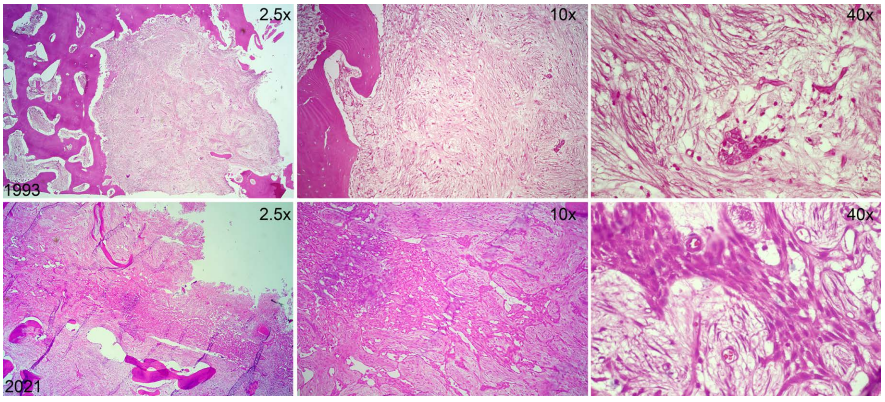

Figure 4. Biopsy taken 1993 (top row) shows fibrous tissue with epithelial islands of OFD-AD, in the bottom row curettage exhibits features of AD with prominent epithelial islands in fibrous background. Magnifications as indicated in the slides top right corner.

An extralesional resection of the involved 21 cm long diaphyseal tibial segment was performed; no elements of osteofibrous dysplasia or Adamantinoma were found in the pathologic examination of the periosteum and sections from the ends of the resected bone segment, indicating tumour free resection margins. The resected specimen was debrided and reamed (**Figure 5**). A slot to accommodate the transposed fibula was made. The tibial segment was pasteurized in 10% saline at 65˚C for 25 minutes according to Ji *et al.* [11] and replanted. For biologic reconstruction the ipsilateral fibula was transposed as a vascularized bone flap, retaining its proximal and distal vascular pedicle and inserted into the prepared slot in the pasteurized tibial autograft.

Skin slough developed distally and to avoid impending exposure of the reconstruction, radical soft tissue debridement was performed and the defect covered with a free microvascular gracilis muscle flap and split skin graft. Distally the fibula and pasteurized tibia fused primarily (**Figure 6**). Proximally autologous trabecular iliac bone graft at 9 months after the initial procedure led to fusion of

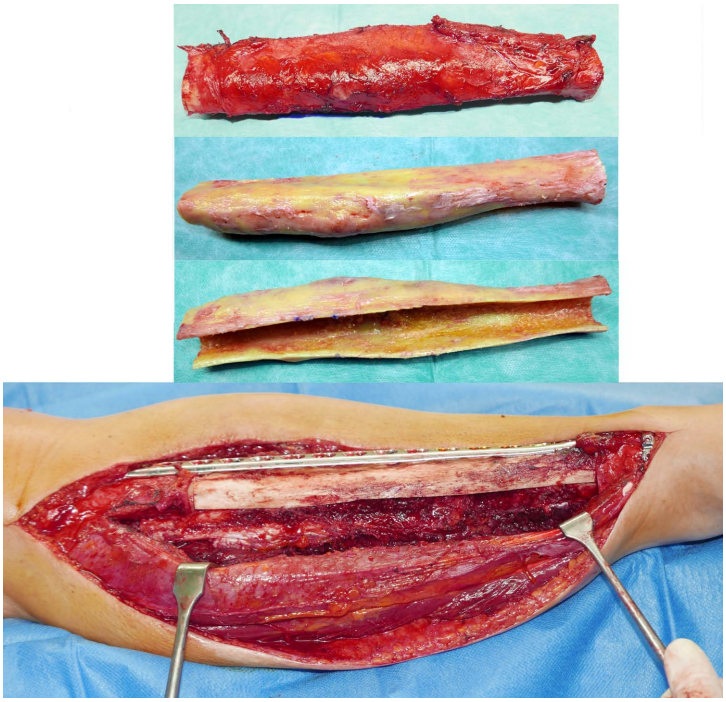

Figure 5. Intraoperative documentation. The resected diaphysis (top) was cleaned from surrounding soft tissue and bone marrow and then pasteurized. The slot is cut to allow inserting the pedicled fibula without compromising the vascular pedicles. Reconstruction with the replanted bone and plate stabilization before wound closure (bottom).

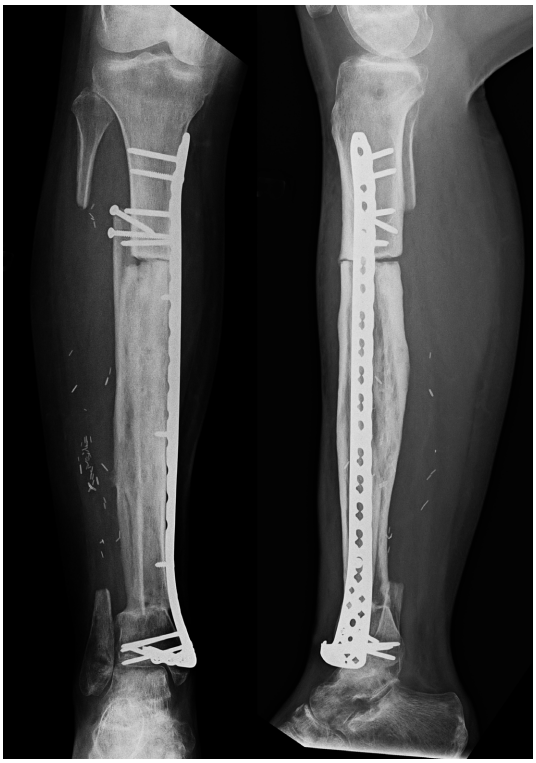

Figure 6. X-Rays at 7 months postoperative showing fusion of the fibula and replanted tibia distally; a gap between the proximal tibia segments remains and no fusion of the fibula proximally has developed.

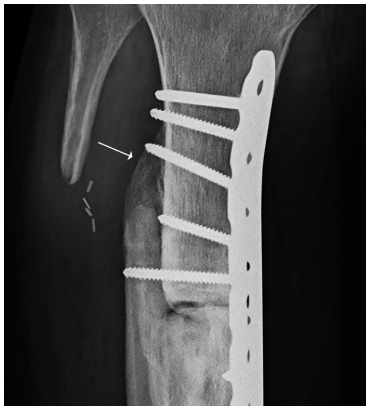

Figure 7. X-Rays at 5 months after trabecular bone plasty and reosteosynthesis 14 months after the initial surgery show the proximal fusion of the fibula to the tibia (arrow).

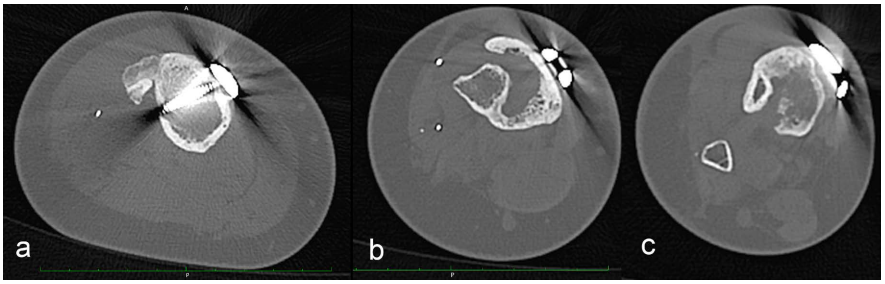

Figure 8. CT images show fusion of the fibula to the proximal tibia (a), fusion of the fibula to the pasteurized tibia segment in the middle third (b) and distally (c).

the proximal fibular stump to the proximal tibia (**Figure 7** and **Figure 8**).

At 15 months after resection the patient is fully weight bearing. In a PET-CT there are neither active foci nor metastases.

3. Discussion

This case adds to the four cases of the rare progression of osteofibrous dysplasia-like adamantinoma to classic adamantinoma hitherto reported [8] [9] [10]. The terminology "Osteofibrous dysplasia of the tibia and fibula" often is referred to as "Campanacci syndrome". It frequently shows spontaneous regression and surgery should be delayed as any intervention may even activate progression. Morphologic features (particularly the shared presence of cytokeratin-positive cells) suggest the association with adamantinoma [12] [13] [14]. The terms differentiated Adamantinoma and OFD-AD are used synonymously [2]. "Classic" AD is a slowly developing malignant tumor with usually late appearance of metastases, whereas the dedifferentiated adamantinoma is a high grade malignant tumor with loss of epithelial differentiation and gain of sarcomatoid change [2].

Therefore patients affected by pathologic features in this spectrum present diagnostic and therapeutic challenges.

Our patient chose a biologic reconstruction. The procedure was planned in analogy to Capanna *et al.* [15], Lu *et al.* [16], Weichmann *et al.* [17] and Momeni

et al. [18]. As an appropriate allograft was not available, the decision was made to replant the resected tibia after intraoperative external pasteurization for autologous biologic stabilization. Other options for tumor inactivation would have been external beam irradiation [19] or freezing in liquid nitrogen [20].

Before resection of the tibia diaphysis an external fixator was mounted to stabilize the leg during preparation of the fibula. The tibia was resected extraperiosteally at the predetermined level including the most distal changes (X-Ray, MRI, PET).

On a separate table the 21 cm long resected tibia segment was reamed, slotted and freed from all soft tissue. All removed tissue was examined pathologically. The removed periosteum was free of tumor, while the bone showed the OFD-AD and some islands of AD.

The prepared tibia (**Figure 5**) was then pasteurized at 65˚C for 30 minutes in 10% saline [11]. However, other authors used normal saline at 65˚ for 30 to 60 minutes [21] [22].

The vascularized fibula was mobilized in a classic fibula-pro-tibia technique. 6 cm of the distal fibula were spared to avoid ankle instability. In order to fit the fibula into the distal remaining tibia metaphysis and to attach the fibula to the retained proximal tibia diaphysis the resected/pasteurized tibia segment was shortened by 2 cm. The fibula could comfortably be inserted into the slot of the tibia. Osteosynthesis was performed using a standard LCP anteromedial tibia plate Depuy Synthes®. The patient developed a distal skin slough, which needed debridement and the soft tissue defect was successfully reconstructed with a free microvascular M. gracilis flap. The clinical situation at 12 months postoperative is presented in **Figure 9**.

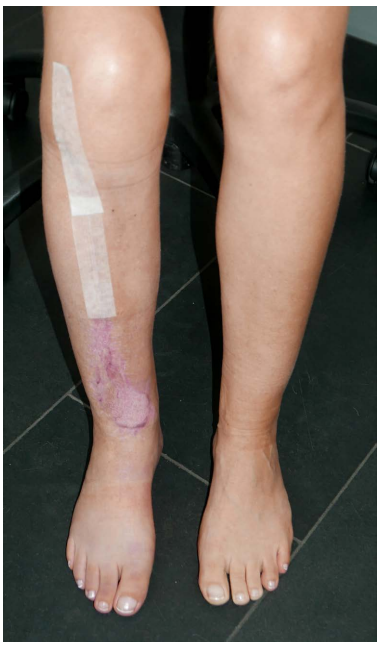

Figure 9. Clinical situation at 12 months after resection. Distally the scars of the free microvascular M. gracilis covered with split skin can be recognized. Proximally the patient still applied strips for scar cosmesis.

4. Conclusion

Besides the pathologic "diagnostic label" we are confronted with the need to assess and understand the biology in cases that fall within the spectrum including OFD, OFD-AD and AD. This case is of special interest because of the long-term f/u of an OFD-AD over 36 years and late transition into AD. The correct balance between careful observation and resection appears mandatory. We suggest to follow patients with OFD-AD indefinitely with imaging at long intervals and as soon as the patient notices any change of symptoms.

Ethical Approval

The study was approved by the institutional review boards.

Informed Consent

The patient has been informed and agreed to submission of her case for publication. All details and radiographic images have been deidentified to protect patient confidentiality.

Authors' Contribution

All authors have been involved in the treatment of the patient in a team approach. They contributed equally to the evaluation and interpretation of images and pathologic findings and finalizing the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Campanacci, M. and Laus, M. (1981) Osteofibrous Dysplasia of the Tibia and Fibula. *The Journal of Bone and Joint Surgery. American Volume*, **63**, 367-375. https://doi.org/10.2106/00004623-198163030-00007

[2] Nielsen, G.P. and Hogendorn, P.C.W. (2020) Osteofibrous Dysplasia, Adamantinoma of Long Bones. Editorial Board. Soft Tissue and Bone Tumors. WHO Classification of Tumors Series, 5th Edition, Vol. 3, International Agency for Research on Cancer, Lyon, 460-466. https://publications.iarc.fr/588

[3] Most, M.J., Sim, F.H. and Inwards, C.Y. (2010) Osteofibrous Dysplasia and Adamantinoma. *Journal of the American Academy of Orthopaedic Surgeons*, **18**, 359-366. https://doi.org/10.5435/00124635-201006000-00008

[4] Exner, G.U., von Hochstetter, A.R. and Pfirrmann, C.W.A. (2018) Osteofibrous Dysplasia with Rhabdoid Elements in a 38-Year-Old Man with Spontaneous Regression over Five Years. *JBJS Case Connector*, **8**, e51. https://doi.org/10.2106/JBJS.CC.17.00294

[5] Lee, R.S., Weitzel, S., Eastwood, D., Monsell, F., Pringle, J., Cannon, S.R. and Briggs, T.W.R. (2006) Osteofibrous Dysplasia of the Tibia. Is There a Need for a Radical Approach? *The Journal of Bone and Joint Surgery. British Volume*, **88**, 658-664. https://doi.org/10.1302/0301-620X.88B5.17358

[6] Dala-Ali, B., Donnan, L., Masterton, G., Briggs, L., Kauiers, C., O'Sullivan, M., Calder, P. and Eastwood, D.M. (2022) Osteofibrous Dysplasia of the Tibia. The Importance of Deformity in Surveillance. *The Bone & Joint Journal*, **104**-**B**, 302-308. https://doi.org/10.1302/0301-620X.104B2.BJJ-2021-0815.R1

[7] Scholfield, D.W., Sadozai, Z., Ghali, C., Sumathi, V., Douis, H., Gaston, L., Grimer, R.J. and Jeys, L. (2017) Does Osteofibrous Dysplasia Progress into Adamantinoma and How Should They Be Treated? *The Bone & Joint Journal*, **99**-**B**, 409-416. https://doi.org/10.1302/0301-620X.99B3.38050

[8] Schutgens, E.M., Pizzi, P., Baumhoer, D., Pollock, R., Bové, J.V.M.G., Hogendorn, P.C.W., Dijkstra, P.D.S., Rueten-Budde, A.J., Jutte, P.C., Traub, F., Leithner, A., Tunn, P.U., Funovics, P., Sys, G., San-Julian, M., Schaap, G.R., *et al.* (2020) Surgical Outcome and Oncological Survival of Osteofibrous Dysplasia-Like and Classic Adamantinoma. An International Multicenter Study of 318 Cases. *The Journal of Bone and Joint Surgery. American Volume*, **102**, 1703-1713. https://doi.org/10.2106/JBJS.19.01056

[9] Hazelbag, H.M., Timiniau, A.M., Fleuren, G.J. and Hogendorn, P.C.W. (1994) Adamantinoma of the Long Bones. A Clinicopathological Study of Thirty-Two Patients with Emphasis on Histopathological Subtype. Precursor Lesion, and Biological Behavior. *The Journal of Bone and Joint Surgery A*, **76**, 1482-1499. https://doi.org/10.2106/00004623-199410000-00008

[10] Hatori, M., Watanabe, M., Hosaka, M., Sasano, H., Narita, M. and Kokubun, S. (2006) A Classic Adamantinoma Arising from Osteofibrous Dysplasia-Like Adamantinoma in the Lower Leg. A Case Report and Review of the Literature. *The Tohoku Journal of Experimental Medicine*, **209**, 53-59. https://doi.org/10.1620/tjem.209.53

[11] Ji, T., Li, Y., Xing, Z., Tang, X., Yang, R. and Guo, W. (2021) Assessment of the Viability and Union Feature of Diaphysis Reconstruction Using Pasteurized Tumor Bone and Intramedullary Free Fibular after Tumor Resection. *Journal of Pediatric Orthopaedics*, **41**, e833-e840. https://doi.org/10.1097/BPO.0000000000001936

[12] Sweet, D.E., Vinh, T.N. and Devaney, K. (1992) Cortical Osteofibrous Dysplasia of Long Bone and Its Relation to Adamantinoma. A Clinicopathologic Study of 30 Cases. *The American Journal of Surgical Pathology*, **16**, 282-290. https://doi.org/10.1097/00000478-199203000-00009

[13] Cerniak, B., Rojas-Corona, R.R. and Dorfman, H.D. (1989) Morphologic Diversity of Long Bone Adamantinoma. The Concept of Differentiated (Regressing) Adamantinoma and Its Relationship to Osteofibrous Dysplasia. *Cancer*, **64**, 19-34. https://doi.org/10.1002/1097-0142(19891201)64:11<2319::AID-CNCR2820641123>3.0.CO;2-0

[14] Kahn, L.B. (2003) Adamantinoma, Osteofibrous Dysplasia and Differentiated Adamantinoma. *Skeletal Radiology*, **32**, 245-258. https://doi.org/10.1007/s00256-003-0624-2

[15] Capanna, R., Campanacci, D.A., Belot, N., Beltrami, G., Manfrini, I.M. and Ceruso, M. (2007) A New Reconstructive Technique for Intercalary Defects of Long Bones: The Association of Massive Allografts with Vascularized Fibular Autograft. Long-Term Results and Comparison with Alternative Techniques. *Orthopedic Clinics of North America*, **38**, 51-60. https://doi.org/10.1016/j.ocl.2006.10.008

[16] Lu, Y., Zhu, H., Huang, M., Zhang, C., Chen, G., Ji, C., Wang, Z. and Li, J. (2020) Is Frozen Tumour-Bearing Autograft with Concurrent Vascularized Fibula an Alternative to the Capanna Technique for the Intercalary Reconstruction after Resection of Osteosarcoma in the Lower Limb? *The Bone & Joint Journal*, **102**-**B**, 646-652.

https://doi.org/10.1302/0301-620X.102B5.BJJ-2019-1380.R1

[17] Weichmann, K.E., Dec, W., Morris, C.D., Mehrara, B.J. and Disa, J.J. (2015) Lower Extremity Osseous Oncologic Reconstruction with Composite Microsurgical Free Fibula inside Massive Bony Allograft. *Plastic and Reconstructive Surgery*, **136**, 396-403. https://doi.org/10.1097/PRS.0000000000001463

[18] Momeni, A., Weber, K.L. and Kovach, S.J. (2018) A Modification of an Established Method of Intercalary Extremity Bone Defect Reconstruction. The "Hemi-Capanna" Technique. *Annals of Plastic Surgery*, **81**, 240-243. https://doi.org/10.1097/SAP.0000000000001554

[19] Klein, A., Bakhshai, Y., Roeder, F., Birkenmaier, C., Baur-Melnyk, A. and Dürr, H.R. (2021) Technique and Results after Immediate Orthotopic Replantation of Extracorporeally Irradiated Tumor Bone Autografts with and without Fibular Augmentation in Extremity Tumors. *BMC Musculoskeletal Disorders*, **22**, Article No. 750. https://doi.org/10.1186/s12891-021-04629-3

[20] Yamamoto, N., Hayashi, K. and Tsuchiya, H. (2019) Progress in Biological Reconstruction and Enhanced Bone Revitalization for Bone Defects. *Journal of Orthopaedic Science*, **24**, 387-392. https://doi.org/10.1016/j.jos.2019.01.015

[21] Singh, V.A., Saad, M. and Pailoor, J. (2010) Which Is the Best Method of Sterilization of Tumour Bone for Replantation? A Biomechanical and Histopathological Study. *BioMedical Engineering OnLine*, **9**, 48-62. https://doi.org/10.1186/1475-925X-9-48

[22] Liu, T., Zhang, Q., Guo, X., Zhang, X., Li, Z. and Li, X. (2014) Treatment and Outcome of Malignant Bone Tumors of the Proximal Humerus: Biologic versus Endoprosthetic Reconstruction. *BMC Musculoskeletal Disorders*, **15**, Article No. 69. https://doi.org/10.1186/1471-2474-15-69